Typical skeletal location and differential diagnosis of bone tumors.

Poster No.:	C-2418
Congress:	ECR 2015
Type:	Educational Exhibit
Authors:	M. Barros[1], L. A. Ferreira[1], Y. Costa[2], P. J. V. Coelho[1], F. Caseiro Alves[1]; [1]Coimbra/PT, [2]Faro/PT
Keywords:	Musculoskeletal bone, Bones, Oncology, Conventional radiography, CT, MR, Education, Education and training, Epidemiology
DOI:	10.1594/ecr2015/C-2418

Learning objectives

- Illustrate imaging findings of benign and malignant bone tumors and their typical distribution in the skeleton;
- Review of the prevalence and age group of the more frequent bone tumors;
- Non-tumor bone lesions will also be mentioned, which to some extent, can mimic and confound the diagnosis of a true bone tumor.

Background

Bone tumors have a nonspecific clinical presentation and are often incidental findings in imaging studies namely in conventional radiology. This imaging method provides very important information about the location and aggressive characteristics of a bone tumor.

The following diagram illustrates the main aggressive characteristics of a bone lesion (Fig. 1 on page 7).

To adequately characterize a bone lesion one must take into account several factors such as: whether the osseous lesion is lytic or sclerotic, well-defined or ill-defined margins, patient age, bone localization (epiphysis, metaphysis or diaphysis), single/multiple, matrix calcifications, zone of transition between the lesion and the adjacent normal bone, type of bone destruction and periosteal reaction, as well as associated soft tissue mass. It is also important to consider that certain osseous tumors show predilection towards different skeletal segments which helps in the differential diagnosis.

AGE

To evaluate a bone lesion it is of extreme importance to know the patient's age since certain tumors have a predilection for specific age groups.

The following figure illustrates bone lesions distribution according with age group (Fig. 2 on page 8).

BONE LOCATION

The location in the bone of an osseous lesion is fundamental since each region of bone has biologic properties that predispose it to different tumors. This characteristic is very helpful because it provides a diagnostic clue based solely on this trait.

The location in bone is described in two axes: longitudinal and axial.

Longitudinal location refers to the **epiphysis**, **metaphysis** and **diaphysis** of bone.

Epiphysis:

The epiphysis develops separately from the growth plate as a secondary ossification center, as such, tumors found on the epiphysis are unique. Other secondary ossification centers, such as the apophyses and sesamoids, should be considered as epiphyseal locations in terms of the tumor differential diagnosis. Typical tumors at this site include chondroblastoma, giant cell tumor (with contiguous involvement of the metaphysis), subchondral cyst, and infection.

Metaphysis:

The metaphysis is the most metabolically active portion of bone which makes it a common location for several bone lesions. Typical tumors at this site include nonossifying fibroma, aneurysmal bone cyst and giant cell tumor (while centered in the metaphysis, often extends into the epiphysis).

Diaphysis:

Tumors with a predisposition to the diaphysis include fibrous dysplasia, langerhans cell histiocytosis, osteoid osteoma, adamantinoma, Ewing sarcoma, lymphoma and multiple myeloma. Osteofibrous dysplasia is exclusively seen in the diaphysis.

Differentiating between a diaphyseal and a metaphyseal location is not always possible. Many lesions can be located in both the metaphysis and diaphysis or move from the former to the latter during growth.

The axial location of a tumor is also predictive of the diagnosis. Axial location in bone is described as **central medullary**, **eccentric medullary**, **intracortical** and **juxtacortical/ surface** (Fig. 3 on page 9).

Central and eccentric lesions:

If a medullary lesion is located centrally or eccentrically, this can aid in distinguishing between bone lesions such as bone cyst versus aneurysmal bone cyst, enchondroma versus chondromyxoid fibroma, and fibrous dysplasia versus nonossifying fibroma. In contrast to enchondromas, chondromyxoid fibromas originate eccentrically or intracortically, often with thinning or expansion of the cortex. Chondrosarcoma is usually located centrally, as well as the round cell marrow tumors such as Ewing sarcoma, lymphoma, and myeloma. The original location of osteosarcoma and malignant fibrous histiocytoma is typically eccentric but their enlarged size can make this finding difficult to make.

Intracortical lesions:

The differential diagnosis of intracortical lesions is limited, one must consider osteoid osteoma, cortical desmoids, osteofibrous dysplasia and adamantinoma.

Juxtacortical tumors:

Tumors that arise from the surface of the bone include osteochondroma, periosteal chondroma, and surface variants of osteosarcoma and chondrosarcoma.

BONE MATRIX

The presence of calcifications in the interior of an osseous lesion is diagnostically very useful. There are two types of mineralization:

- **Osteoid matrix**: trabecular ossification pattern in bone forming lesions, with a cloud-like or cotton-like appearance, which can be observed in osseous tumors like osteosarcoma. The osteoid matrix can be neoplastic or reactive. Evaluation of the morphology can be helpful in distinguishing between these patterns (Fig. 4 on page 10).

- **Chondroid matrix:** it will present itself as popcorn-like calcifications, ring and arc mineralization and is found in cartilaginous tumors such as enchondroma and chondrosarcoma. Chondroid mineralization is seen in benign and malignant neoplasms. In the case of chondroblastoma and chondromyxoid fibroma, the mineralization can be subtle and difficult to see on radiographs. Contrariwise, the punctate appearance of osteoid mineralization in osteoblastoma and osteonecrosis can be mistaken for chondroid mineralization (Fig. 5 on page 11).

- **Nonmineralized matrix:** it can be fluid, fat, or soft tissue and is best differentiated by MRI.

ZONE OF TRANSITION AND TYPE OF BONE DESTRUCTION

When evaluating a lesion, it is important to assess the transition zone between the lesion and adjacent normal bone. This characteristic is highly predictive of the aggressiveness of the tumor and is fundamental in determining if it's a slow growing or fast growing lesion.

A narrow transition area with sclerotic borders is a sign of slow growing lesions in favor of a benign lesion. However, despite benign radiographic features, in patients over 40 years, metastasis or myeloma also have to be considered.

On the other hand, a wide zone of transition reflects more rapid growth and reveals aggressiveness of the lesion, which is associated with malignant bone tumors. However, this is not a rule and may be present for example in osteomyelitis or eosinophilic granuloma, which can exhibit aggressive growth pattern.

The type of bone lysis is usually linked to growth rate of the tumor and can be geographic, moth-eaten, or permeative (Fig. 6 on page 12).

Geographic Bone Lysis

Geographic lysis describes a single focus of bone destruction. It can have a narrow or wide zone of transition and is subdivided as follows:

Type 1a: well defined with sclerosis and implying a slow rate of growth, includes bone cysts, chondroblastoma, enchondroma, nonossifying fibroma and fibrous dysplasia (Fig. 7 on page 13 Fig. 7 on page 13).

Type 1b: well defined without sclerosis and implying a slow to intermediate rate of growth, includes giant cell tumor, bone cyst, chondroblastoma, chondromyxoid fibroma, enchondroma, fibrous dysplasia, myeloma, and metastatic carcinoma. (Fig. 8 on page 14).

Type 1c: ill-defined, implying an intermediate to fast rate of growth. Tumors with this margin type include chondrosarcoma, aneurysmal bone cyst, malignant fibrous histiocytoma, giant cell tumor, osteosarcoma, metastatic carcinoma and myeloma (Fig. 9 on page 15Fig. 9 on page 15 Fig. 9 on page 15).

Moth-eaten

This type of bone lysis represents multiple small destructive foci of tumor in which each area is large enough to be defined as a focal lesion (Fig. 10 on page 16).

Permeative

This type also represents multiple destructive foci of a bone lesion. However, in contrast to the moth-eaten pattern, each individual site of bone destruction is too small (<5mm) to have a perceptible margin. Tumors with moth-eaten and permeative lysis have typically a wide zone of transition due to rapid tumor growth and the differential diagnosis is identical (Fig. 11 on page 17).

Lesions that can produce moth-eaten or permeative lysis include round cell tumors (Ewing sarcoma, lymphoma), osteosarcoma, chondrosarcoma, Langerhans cell histiocytosis, acute osteomyelitis, metabolic disorders, malignant fibrous histiocytoma, metastatic carcinoma and multiple myeloma.

CORTICAL DESTRUCTION AND PERIOSTEAL REACTION

Periosteal reaction reflects the biologic activity of the bone tumor. A periosteal reaction is a non-specific response and two types of periosteal reaction can be distinguished: non-aggressive and aggressive type. The non-aggressive pattern is a compact periosteal reaction type and is seen in benign tumors and post-trauma. In this benign pattern that occurs in slow growth lesions, the periosteum has enough time to form new bone with normal appearing cortex.

The aggressive pattern can present as "sunburst", "hair-on-end", lamellated pattern or as Codman triangle. In this pattern the periosteum doesn't have enough time to consolidate. This pattern of periosteal reaction can also occur in benign lesions with a more aggressive behavior such as osteomyelitis.

SOFT TISSUE MASS

Usually the benign bone lesions don't present with an associated mass of soft tissue, when the latter is present, it is associated with aggressive bone lesions.

SKELETAL DISTRIBUTION

In some cases knowing the particular affected bone can aid in diagnosis. While the location of a tumor within a bone is more diagnostically useful, some regions of the skeleton are especially predisposed to particular tumors. The sacrum and the anterior and posterior vertebral elements each have their own distinctive range of tumors such as the chordoma which locates preferentially at the clivus, vertebral bodies, and sacrum and the osteoblastoma which tends to arise in the posterior elements of the vertebral column. Intraosseous lipoma tends to appear in the calcaneus, intertrochanteric and subtrochanteric regions of the femur, more rarely arising in different parts of the skeleton. The following figure illustrates typical skeletal distribution of some bone lesions (Fig. 12 on page 18).

MULTIPLE LESIONS

Multiple lesions often indicate metastatic disease or multiple myeloma. This is not always true, several other entities can present as polyostotic lesions, such as brown tumors, non-ossifying fibroma, fibrous dysplasia, multifocal osteomyelitis, multiple enchondromas and eosinophilic granuloma.

IMAGING MODALITIES

The use of conventional radiology in detecting and characterizing bone lesions is fundamental but there are other complementary imaging modalities namely CT and MRI. CT is important to evaluate matrix calcification and cortical integrity. MRI is the technique of choice for local tumor staging, allowing the determination of intramedullary extension and neuro-vascular involvement and providing vital information regarding lesion surgical ressecability.

Scintigraphy is helpful in documenting bone metastasis or lesion multiplicity.

Images for this section:

Features of non aggressive lesions	Features of aggressive lesions
Sclerotic borders	Without sclerotic borders
Defined margins	Poorly defined margins
Narrow zone of transition	Wide zone of transition
Solid periosteal reaction	Aggressive periosteal reaction: “sunburst”, lamellated or “onion-skin” pattern, Codman triangle.
No soft tissue mass	Soft tissue mass

Fig. 1: Features of non aggressive/agressive bone lesions.

Age Group	Bone Lesions
< 20 years	Langerhans cell histiocytosis, unicameral bone cyst, osteomyelitis, nonossifying fibroma, chondroblastoma, osteosarcoma, Ewing sarcoma, osteoid osteoma, aneurysmal bone cyst, fibrous cortical defect.
20–40 years	Giant cell tumor, enchondroma, osteoblastoma, osteoid osteoma, chondromyxoid fibroma, fibrous dysplasia.
> 40 years	Chondrosarcoma, metastatic carcinoma, multiple myeloma, malignant fibrous histiocytoma and fibrosarcoma of bone, lymphoma in bone, Paget disease.

Fig. 2: Bone lesions distribution based on age group.

Axial Location	Bone Lesions
Central	Enchondroma, fibrous dysplasia, unicameral bone cyst, lymphoma, Ewing Sarcoma.
Eccentric	Nonossifying fibroma, aneurysmal bone cyst, and giant cell tumor, osteosarcoma.
Intracortical	Osteoid osteoma, osteofibrous dysplasia, adamantinoma.
Juxtacortical/Surface	Osteochondroma, juxtacortical chondroma, osteosarcoma surface variants (periosteal and parosteal osteosarcoma)

Fig. 3: Axial location of bone tumors.

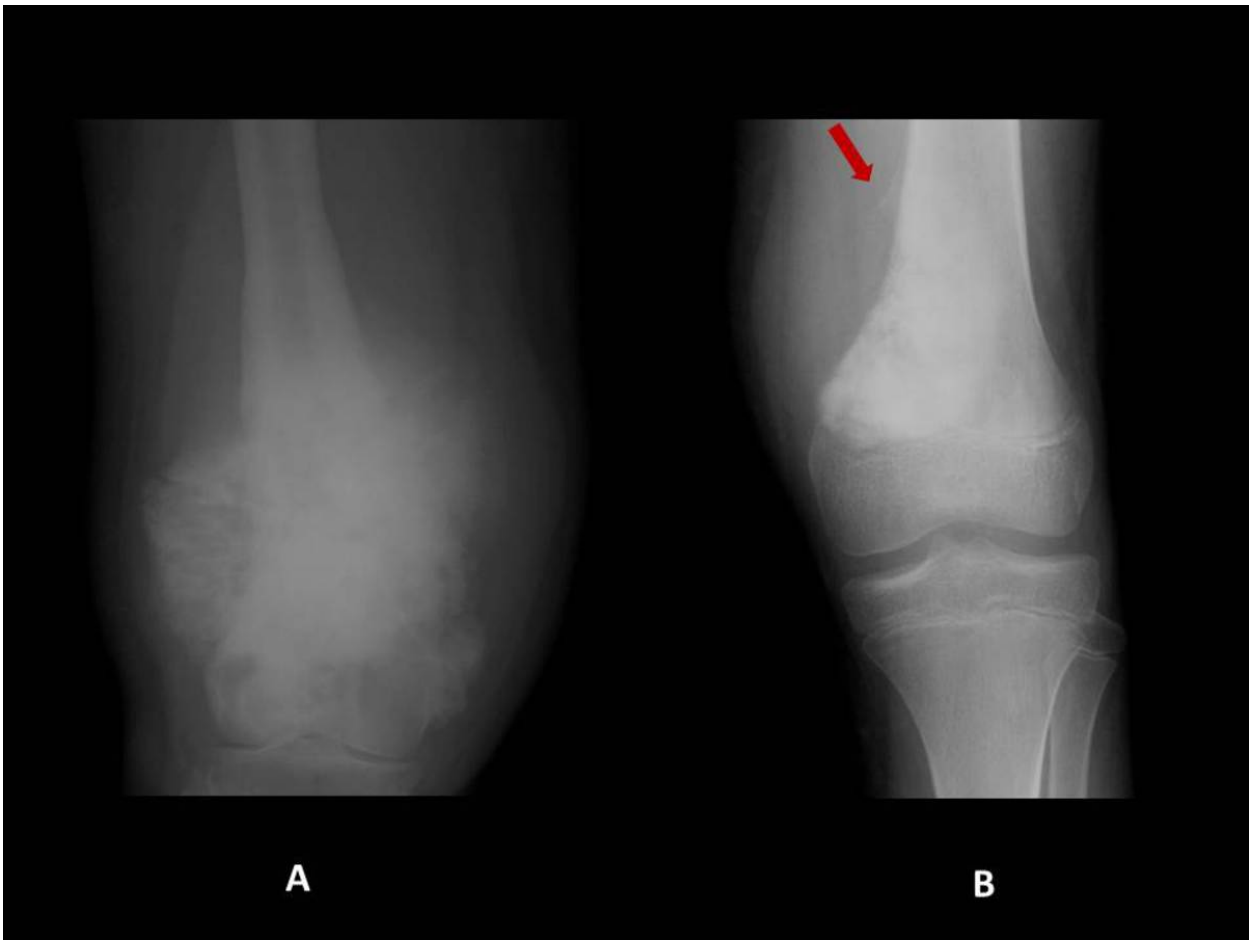


Fig. 4: Osteoid matrix - Cloud-like bone formation and sunburst periosteal reaction in two examples of osteosarcoma. (B) Note the Codman triangle (arrow).

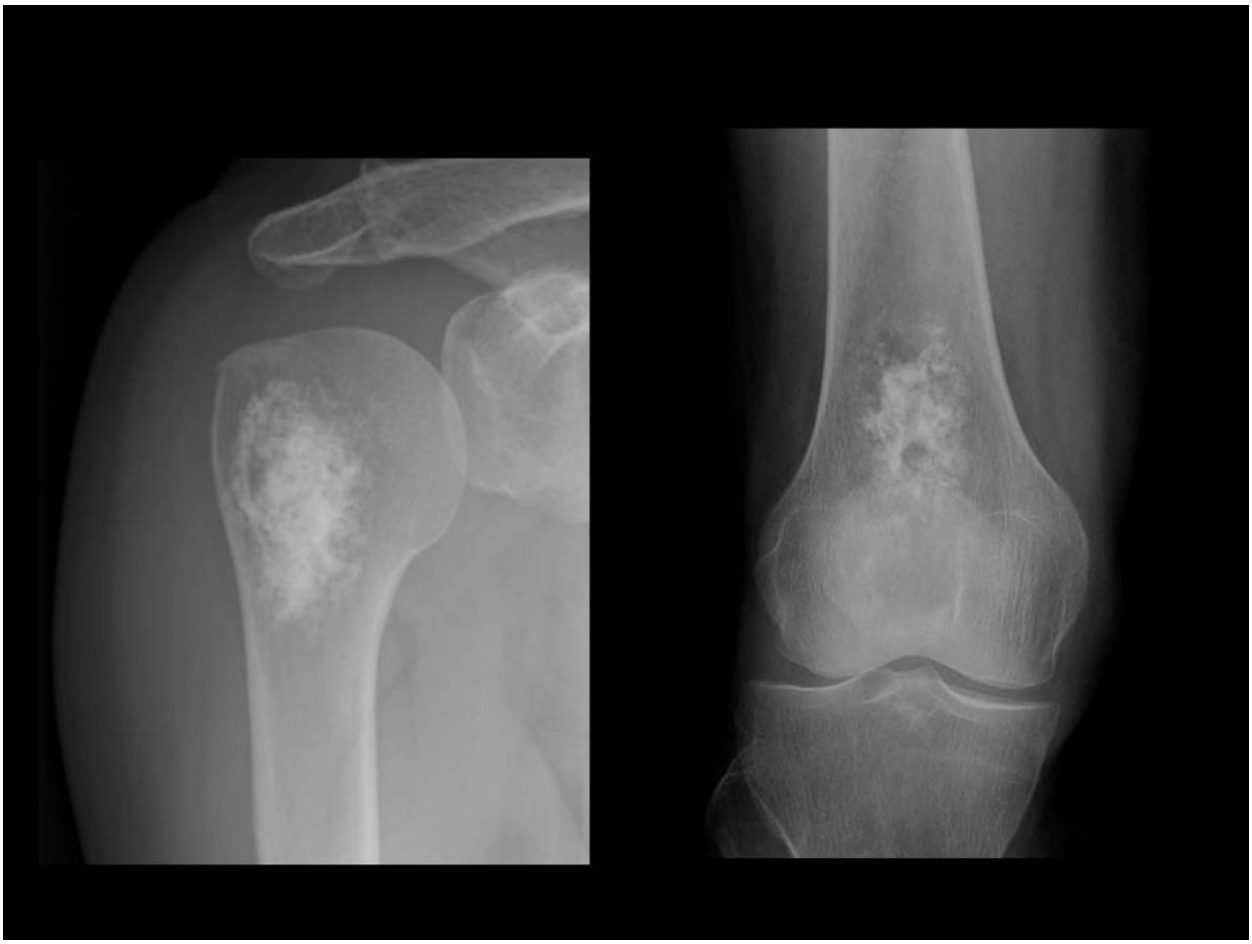
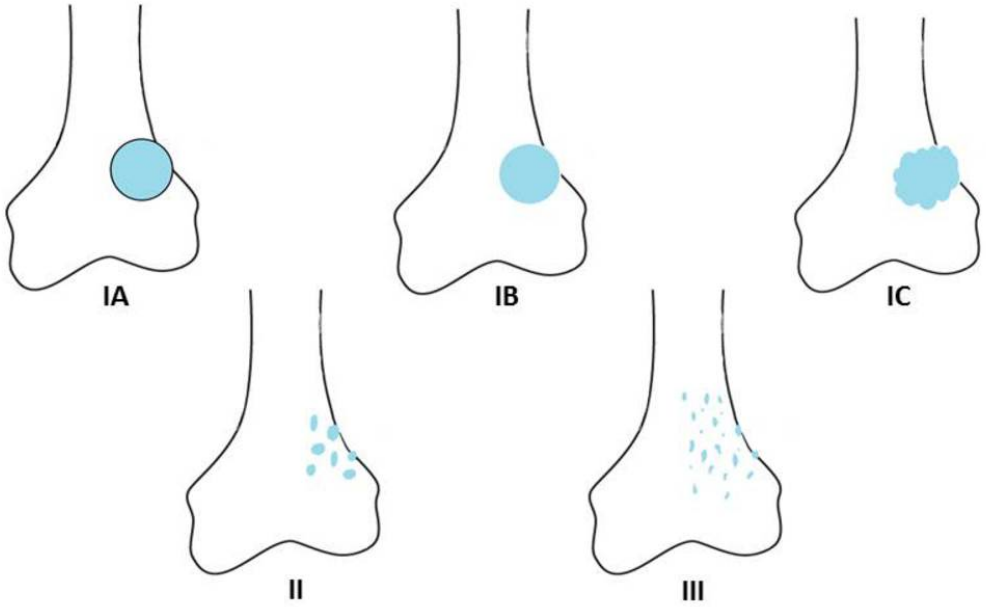


Fig. 5: Chondroid matrix - punctate, rings-and-arcs and popcorn mineralization in two examples of enchondroma.



Different types of bone destruction, as first described by Lodwick :

Type IA - Geographic with sclerotic margins;
Type IB - Geographic without sclerotic margins;
Type IC - Geographic with poorly defined margins;
Type II - Moth-eaten
Type III - Permeative (permeative growth throughout the Haversian channels).

Fig. 6: Types of bone destruction.

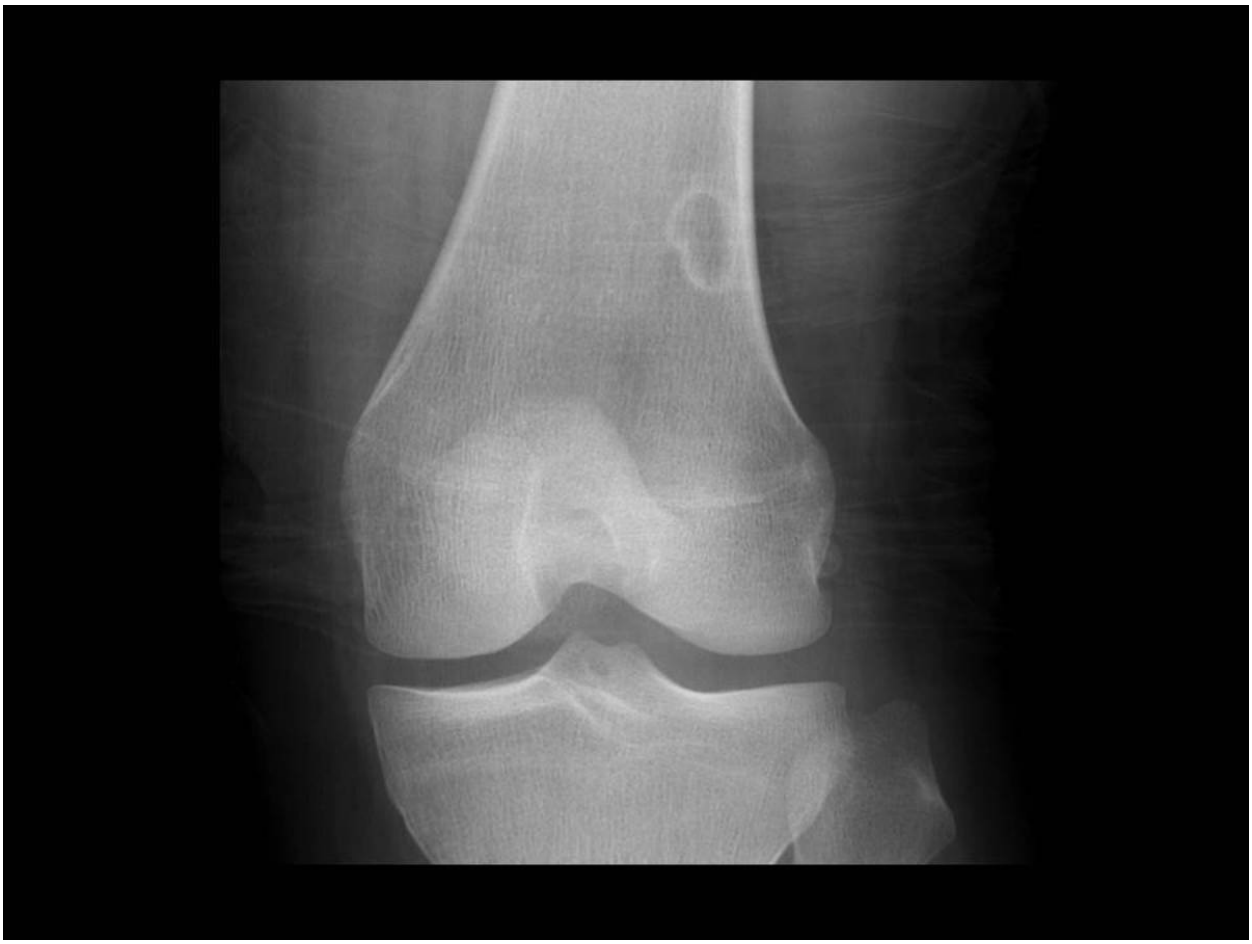


Fig. 7: Type 1a geographic lesion. Radiography shows a well-defined lucency with sclerotic rim in the femur - Non ossifying fibroma.

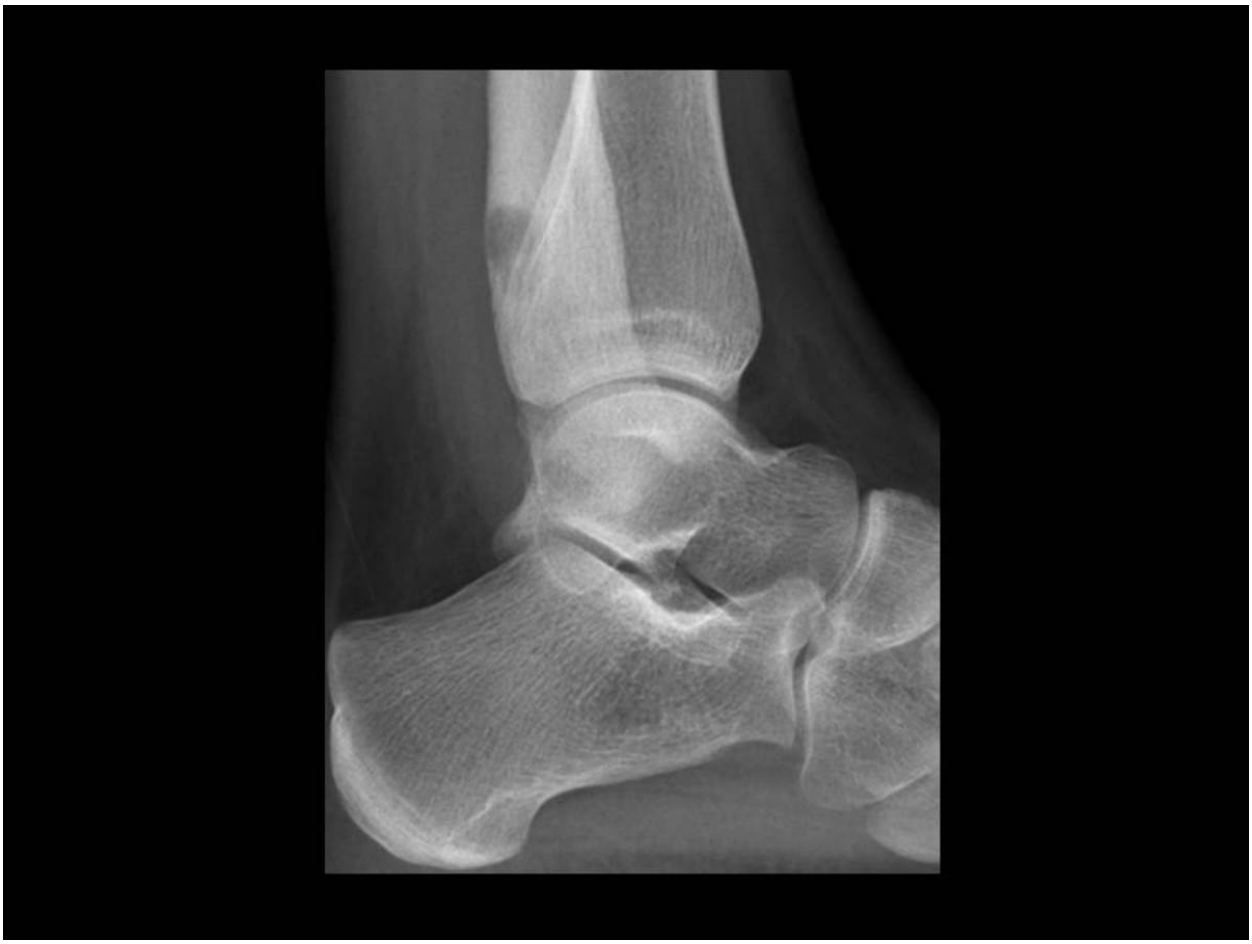


Fig. 8: Type 1b geographic lesion. Radiography shows well-defined geographic lesion without a sclerotic rim in a right fibula - Osteoid osteoma.

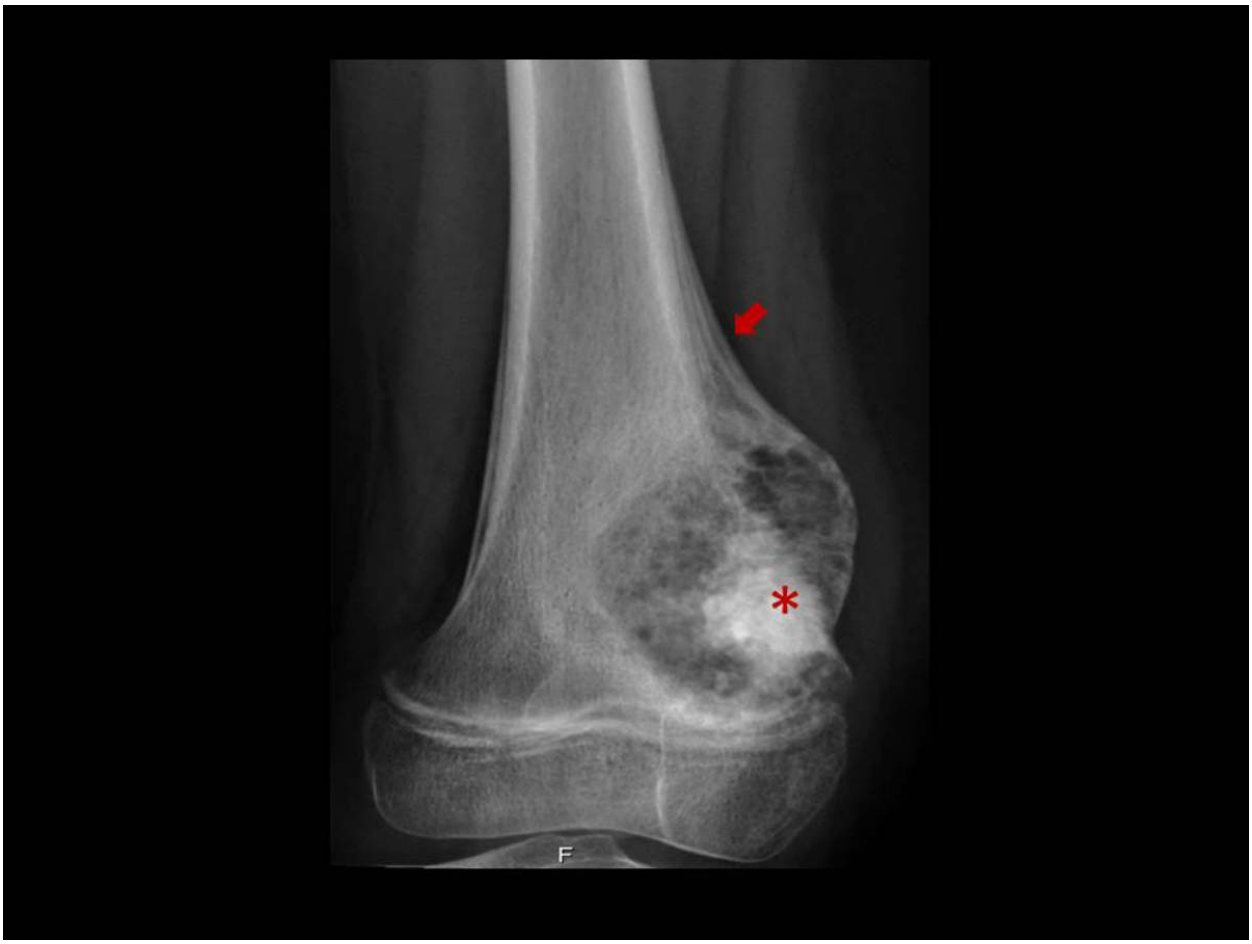


Fig. 9: Type 1c geographic lesion. Radiography shows ill-defined lesion in the femur - Osteossarcoma. Note the lamellated periosteal reaction (arrow) and tumor-induced new bone production (*)

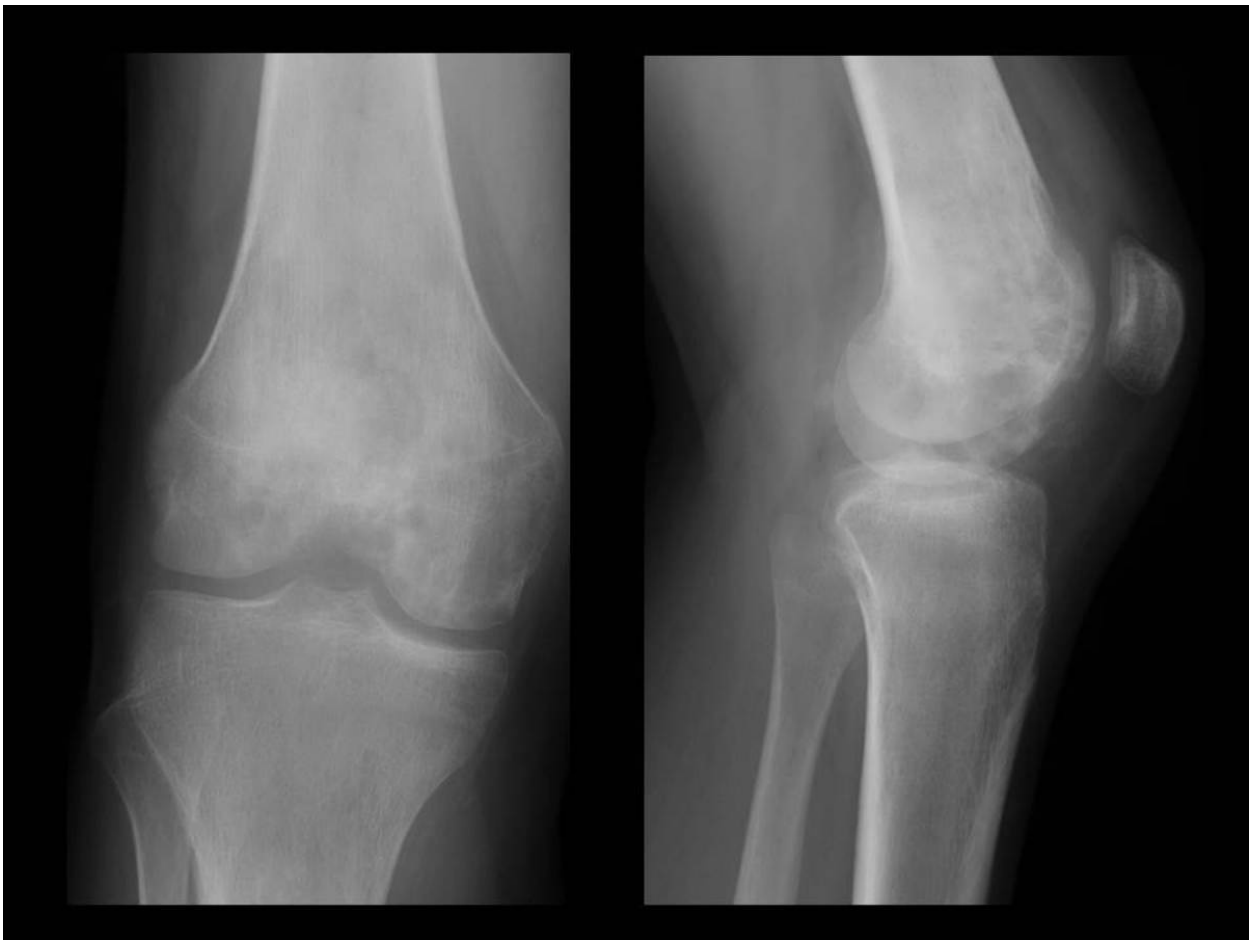


Fig. 10: Type II Moth-eaten lesion. Radiography shows multiple small destructive foci of a bone lesion with ill-defined margins and wide zone of transition - Osteossarcoma.

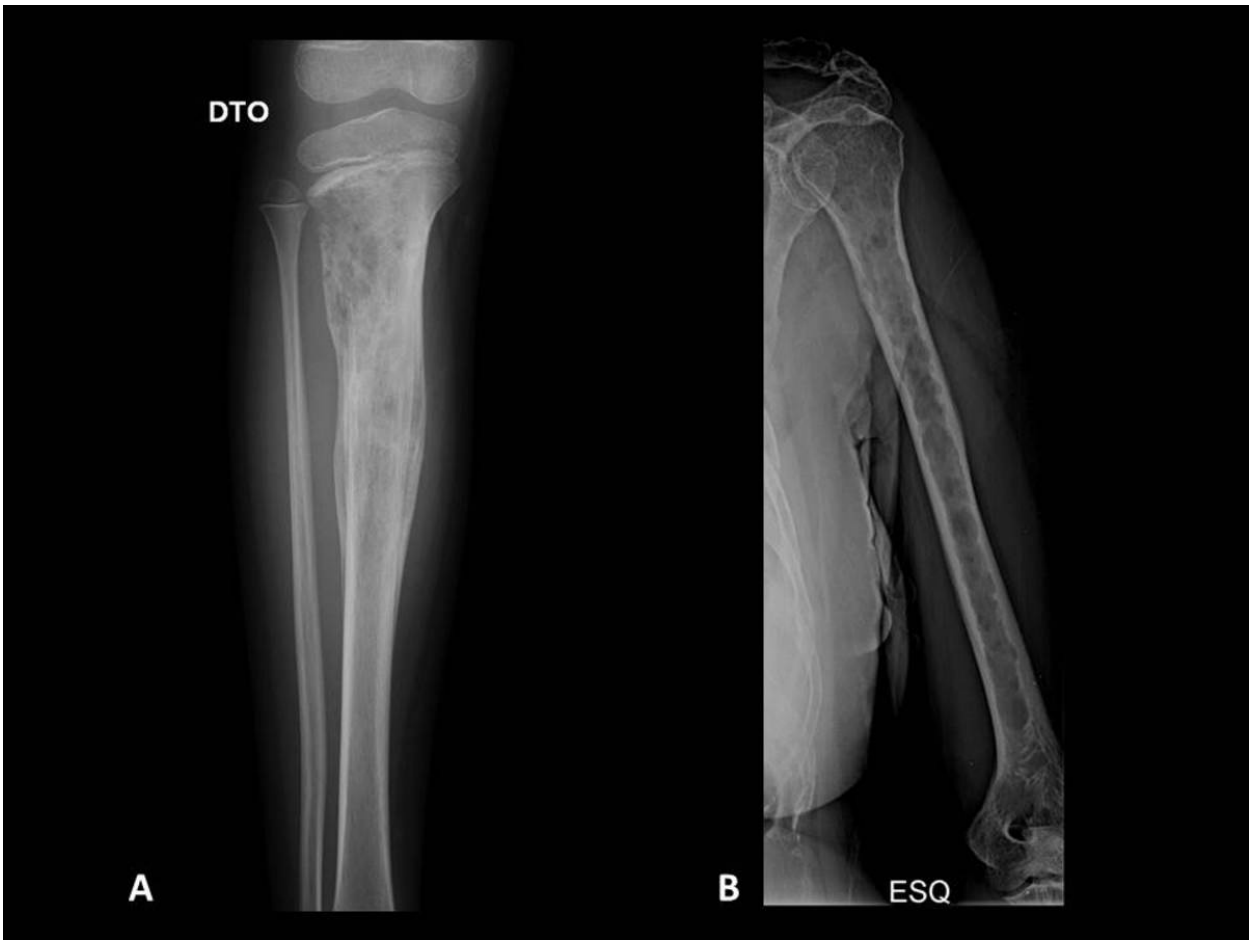


Fig. 11: A - Moth-eaten (type II) and Permeative (type III) lesion. Radiography shows multiple destructive foci of a bone lesion with poorly-defined borders and wide zone of transition. Some lesions are under 5 mm revealing the permeative character of the lesion. Note the lamellated pattern of periosteal reaction - Ewing´s sarcoma. B - Type III Permeative lesion. A diffuse permeative pattern is seen throught the entire humerus - Multiple myeloma. Note the cortical scalloping.

Tumors with distinctive skeletal distributions	
Adamantinoma	Anterior cortex of tibia
Osteofibrous dysplasia	Anterior cortex of tibia
Periosteal Desmoid	Posterior cortex of distal femur
Chordoma	Clivus, vertebral bodies, sacrum
Hemangioma	Vertebral bodies
Intraosseous lipoma	Calcaneus
Osteoblastoma	Posterior elements of spine
Aneurysmal bone cyst	Posterior elements of spine

Fig. 12: Typical skeletal distribution of bone tumors.

Findings and procedure details

A varying imaging approach is often required to better characterize the lesion and extent of the disease, nonetheless conventional radiography is essential and can provide just enough information to guide or even pinpoint the proper diagnosis.

Conventional radiography is an essential aid in the diagnosis of bone lesions, since it localizes the lesions and assesses their aggressive characteristics that suggest malignancy.

For the purpose of this educational exhibit, typical locations of osseous lesions will be approached along the skeleton and in the bone itself, as well as their prevalence according to the usual age group in which they usually occur.

CHONDROBLASTOMA

Chondroblastomas are rare benign cartilaginous neoplasms (1% of all primary bone tumors)characteristically presenting in the epiphyses of long bones and epiphyseal equivalentssuch as apophyses and sesamoids, in skeletally immature patients. Chondroblastomas occur predominantly in young patients (mean age of presentation is approximately 20 years old) and there is a male predilection.

The most common locations for this bone tumor are the proximal tibia, proximal humerus, distal femur and the apophysis of the greater trochanter. Chondroblastoma also occurs in talus and calcaneus, although less frequently (10%). Despite the lesion being typically seen in the epiphysis of growing bones, some cases have been reported after closure of the growth plate in the metaphyseal region (Fig. 13 on page 28).

Chondroblastoma typically appears as (Fig. 14 on page 29):

- Eccentrically lytic lesion located within the epiphysis;
- Well-circumscribed with sclerotic margin;
- Fine calcifications may be visible (40-60%)
- Solid periosteal reaction (seen in up to 50% of cases)
- Scalloping or expansion of cortical bone may also be present.

The diagnosis of chondroblastoma can usually be made by radiographic imaging features when the age of the patient is considered. CT is useful for defining the relationship to the growth plate and articular surface, for detection of matrix mineralization and assessment of the integrity of the cortex (Fig. 15 on page 30).

MRI is ideal for the evaluation of lesion extension and for demonstrating associated adjacent bone marrow and soft-tissue edema, which is seen in a large proportion of cases. Presence of bone marrow edema almost always accompanies chondroblastoma, but is rare in other chondroid tumors, like enchondroma or low-grade chondrosarcoma, as well as not being a usual feature of chondromyxoid fibromas or giant cell tumors. Fluid-fluid levels may occasionally be seen due to an associated aneurysmal bone cyst.

The differential is that of other lesions which have a predilection for the epiphysis or apophysis. The differential diagnosis includes enchondroma, aneurysmal bone cyst, clear cell chondrosarcoma, osteomyelitis with abscess and giant cell tumor.

Treatment typically consists of curettage and bone grafting, but radiofrequency ablation has also been used. However, due to their proximity to the articular surface and growth plate, complete excision is difficult. As a consequence, recurrence rates are relatively high.

In a few cases, malignant transformation has been seen with local invasion of soft tissue as well as pulmonary metastases.

INTRAOSSEOUS LIPOMA

Lipomas can be classified according to their location in the bone as intraosseous, cortical, or parosteal lesions. Intraosseous lipoma is considered a benign rare tumor (with an incidence of less than 0,1% of all primary bone tumors). However, with the increasing use of the imaging modalities, intraosseous lipomas are appearing in higher numbers as incidental findings. It is usually an asymptomatic lesion and most are found in the 4th and 5th decades of life with no gender predilection. Although intraosseous lipomas can be found essentially anywhere within the skeleton, the most common sites are in the calcaneus, intertrochanteric and subtrochanteric regions of the femur, followed by the ilium, proximal tibia, and sacrum. When located in long bones, they tend to be found in the metaphysis (Fig. 16 on page 31).

Intraosseous lipoma has a very characteristic radiographic appearance. It is a benign-appearing radiolucent lesion with well-defined margins, however, thinning and bulging of the cortex may be seen. Central calcifications are frequently present. CT may be helpful in the diagnosis, because the Hounsfield units are consistent with fat content, a finding supported by the MRI imaging. In most cases, the CT and MR imaging characteristics were diagnostic, making biopsy unnecessary (Fig. 17 on page 32).

Intraosseous lipomas may be treated conservatively. Symptomatic lesions with imminent fractures needed curettage and bone grafting. Recurrence after surgical therapy is very rare; however, there are rare case reports of malignant transformation.

OSTEOBLASTOMA

Osteoblastoma is a rare, benign, possibly locally aggressive and painful osteoid-producing tumor. It typically occurs in young patients, around the second decade of life. There is a recognized male predilection.

This bone tumor tends to occur more commonly in the spine usually in posterior elements than in the long bones (Fig. 18 on page 33 and Fig. 19 on page 34). In the long bones, it can be metaphyseal/diaphyseal and eccentric (Fig. 20 on page 35 and Fig. 21 on page 36).

Osteoblastoma is histologically similar to an osteoid osteoma except that it is much larger in size, typically larger than 2 cm. There is high associated vascularity, something which must be considered in a surgical approach.

Osteoblastoma can have a wide range of radiographic patterns:

- Predominantly lytic, with a rim of reactive sclerosis;
- Tends to be expansible, sometimes with cortical destruction;
- Internal matrix mineralization: appearance can mimic chondroid matrix.
- An associated soft tissue mass and secondary aneurysmal bone cyst may be present;

Treatment is through curettage or marginal excision with bone grafting.

CHORDOMA

Chordoma is a slow-growing malignancy arising from remnants of the notochord with a predilection for the sacrum and skull base (Fig. 22 on page 37).

Chordomas represent from 1% to 4% of all primary malignant bone tumours. These tumours occur almost exclusively in the midline of the axial skeleton (Fig. 23 on page 38 and Fig. 24 on page 39).

They arise between the fourth and seventh decades and affect men slightly more often than women. Chordomas often present late in their development, as the mass can be quite large before symptoms arise.

The radiographic appearance:

- Highly destructive lesion with irregular scalloped borders;
- Bone expansion;
- Calcifications in the matrix may occur as a result of tumor necrosis;
- Often large soft tissue mass.

Differential diagnosis: Chondrosarcoma.

Conventional radiography and tomography usually suffices to delineate the tumor but CT or MRI is required to demonstrate soft-tissue extension and invasion of the spinal canal.

Primary treatment is surgical resection and can be associated with a disabling loss of sacral nerve root function. Local recurrence is common due to the difficulty of obtaining negative margins. Metastasis are rare and usually a late event.

SOLITARY PLASMOCYTOMA

Solitary plasmocytoma is a unique mass of neoplastic monoclonal plasma cells which occurs most after 40 years of age (average age: 55 years).

The most common locations are the axial skeleton (spine, skull, ribs and pelvis) and in the diaphysis of long bones (Fig. 25 on page 40 and Fig. 26 on page 41).

Radiographic appearance:

- Geographically well circumscribed osteolytic lesion;
- Endosteal involvement;
- Often marked erosion, expansion, and cortical destruction;
- With advanced disease, destruction can appear moth-eaten or permeative;
- No periosteal reaction;
- No matrix mineralization.

Almost all patients receive chemotherapy with radiotherapy, although the majority of patients will relapse. Painful bone lesions with pathologic fractures can be surgically treated.

EWING SARCOMA

Ewing sarcoma arises from bone marrow and almost always presents with a large soft tissue mass. This tumor has a predilection for the metaphysis or diaphysis of the long

bones, as well as the flat bones such as the scapula, pelvis and the ribs (Fig. 27 on page 42). When it occurs primarily in the rib it can also be known as Askin tumor. Most occur before age 25 and have male predilection.

Clinically, it may present with local pain and systemic symptoms such as fever, fatigue and malaise, which may often mimic osteomyelitis.

Radiographic presentation (Fig. 28 on page 43 and Fig. 29 on page 44):

- Poorly defined osteolytic lesion;
- Permeative or moth-eaten type of bone destruction;
- Large soft tissue mass is almost always present;
- Lamellated ("onion-skin") periosteal reaction;
- No matrix mineralization;
- Intraosseous component can demonstrate subtle reactive sclerosis.

Conventional radiography and CT reveals the pattern of bone destruction and MRI is important for evaluate tumor extension. Radionuclide bone scan provides reliable information concerning the presence of skeletal metastases.

Differential diagnosis:

- Neuroblastoma
- Osteosarcoma
- Osteomyelitis
- Primary lymphoma

Ewing sarcoma is usually treated with chemotherapy, either alone or combined with radiation therapy, followed by surgical resection.

CHONDROMYXOID FIBROMA

Chondromyxoid fibroma is a rare benign cartilaginous lesion with a predilection for the metaphysis of long bones, 60% occurs around the knee joint, although less frequently, it may also found in the short tubular bones of the hands and feet. The chondromyxoid fibroma has an eccentric or cortical location in long bones and may be central when in short tubular or flat bones (Fig. 30 on page 45).

Typically occurs around the second and third decades of life and has no gender preference.

Radiographic appearance (Fig. 31 on page 46):

- Meta-diaphyses of long tubular bones;
- Eccentric well-defined lytic lesion surrounded by a sclerotic rim;
- Typically narrow zone of transition;
- Lobulated margins;
- Expanded and thinned cortex;
- Rare to find calcified tumor matrix within it (subtle chondroid mineralization may be visible on CT).

Differential diagnosis:

- Aneurysmal bone cyst;
- Non-ossifying fibroma;
- Fibrous dysplasia.

Chondromyxoid fibroma is usually treated with surgical excision and bone grafting.

CHONDROSARCOMA

Chondrosarcoma is a common malignant bone tumor (4% of all primary tumors) characterized by the formation of a cartilage matrix. There are several types of chondrossarcoma, each with typical clinical presentation, imagiologic and pathologic features. They can be subdivided regarding origin, being either primary or secondary and according to central or peripheral location.

When this bone tumor arises without a preexisting lesion it is called *primary chondrosarcoma*, on the other hand when it emerges on preexisting benign cartilaginous neoplasms such as enchondromatosis or multiple cartilaginous exostoses it is called *secondary chondrosarcoma*. Secondary chondrosarcomas arising from osteochondromas are typically low grade (Fig. 34 on page 49). At risk are the patients with Ollier disease and Maffucci syndrome.

In the case of primary chondrosarcoma, the histologic type most frequently found is conventional chondrosarcoma (medullary or central chondrosarcoma), with the histologic types clear cell chondrosarcoma, mesenchymal chondrosarcoma, myxoid chondrossarcoma and dedifferentiated chondrosarcoma less frequent.

Conventional Chondrossarcoma:

Also known as central or medullary chondrosarcoma, this tumor is seen twice as frequently in males than in females and more commonly in adults, usually in fourth to fifth decades of life. The most frequent locations are the pelvis and in metaphyseal region of long bones, particularly the femur and proximal humerus (Fig. 32 on page 47).

Most conventional chondrosarcomas are slow-growing tumors, only in rare cases do they metastize to distant areas.

Radiographic appearance (Fig. 33 on page 48):

- Expansive lesion in the medulla;
- Thickening of the cortex and endosteal scalloping;
- Chondroid mineralization, consisting of popcorn-like, annular or comma-shaped calcifications;
- A soft-tissue mass may be present.

In the early stage of development, chondrosarcoma can be indistinguishable from an enchondroma. The key feature in distinguishing low grade chondrosarcoma from enchondroma is the presence of endosteal scalloping. In most cases, conventional radiography is sufficient to make a diagnosis. CT and MRI help delineate the extent of bone destruction as well as marrow involvement and soft-tissue extension.

Prognosis is highly related to the grade of the tumor. Surgical wide excision is the principal treatment modality.

Clear Cell Chondrossarcoma:

Low-grade variant of chondrosarcoma (2% of all chondrosarcomas) has preference for epiphyseal region of long bones, and is found primarily in the proximal femur or humerus (90%).

It occurs at third and fourth decades of life and has a male preference. It may resemble chondroblastoma, but the latter occurs at younger age than clear cell chondrosarcoma.

Radiographic appearance:

- Lytic area occasionally containing calcifications and sclerotic border at epiphyseal region.

Mesenchymal Chondrossarcoma:

Mesenchymal chondrosarcoma is a very rare highly malignant lesion with a strong capacity to metastasize and tends to occur in the second or third decade of life.

Most occurs at craniofacial bones, ribs, iliac bone and vertebrae.

It presents radiographically with two different features, areas of permeative type of bone destruction and areas with typical calcifications of cartilaginous tumor.

Dedifferentiated Chondrossarcoma:

Dedifferentiated chondrosarcoma, the most aggressive type of all cartilage tumors, carries a very poor prognosis. This tumor arises from a preexisting benign chondral lesion or on a low-grade chondrosarcoma. It tends to occur in the fifth to ninth decades of life and the favored sites are the pelvis, femur and humerus.

It has two different components, one well-differentiated chondrogenic component and a high-grade non-cartilaginous component (malignant fibrous histiocytoma, fibrosarcoma or osteosarcoma).

Imagiologic appearance with presence of the two components (Fig. 35 on page 50):

- one component shows the characteristics of a well-differentiated chondrogenic tumor, including chondrogenic calcifications, the other component has an highly aggressive pattern, with aggressive bone destruction, ill-defined margins and a large soft tissue mass.

Juxtacortical chondrosarcoma:

Juxtacortical chondrosarcomas arise from the surface of bone and are typically low grade. The age of presentation is second to forth decade of life (younger than in conventional chondrosarcoma) and occurs more in the male gender. It has a slow and indolent growth and usually occurs in the femoral and humeral metaphysis. Occasionally, juxtacortical chondrosarcoma may be indistinguishable from periosteal osteosarcoma. One way to distinguish both is the existence of chondroid matrix in the former, but this distinction isn't always obvious.

Juxtacortical chondrosarcoma is differentiated from benign juxtacortical chondroma by its larger size, typically greater than 3cm, and its ability to invade marrow.

Juxtacortical chondrosarcoma has similar radiographic and pathologic features as central chondrosarcoma.

ANEURYSMAL BONE CYST

Aneurysmal bone cyst is a common bone lesion and most occur in patients younger than 20 years old. The cause of this lesion is unknown, but alterations related to vascular malformation are believed to play an important role. Aneurysmal bone cyst can develop de novo or it may be secondary to an underlying osseous neoplasm such as a chondroblastoma, osteoblastoma, giant cell tumor or chondrosarcoma.

It usually presents with slow growth but can occasionally show rapid growth and even pathologic fracture, which is the most frequent complication.

An aneurysmal bone cyst tends to occur in long bone metaphysis and eccentric position. It typically presents in the posterior elements of the vertebral column having a differential diagnosis with osteoblastoma. This tumor may sometimes be seen in the diaphysis of a long bone, as well as in flat bones such as the pelvis or scapula (Fig. 36 on page 51).

Radiologic appearance (Fig. 37 on page 52):

- Cystic cavities with cortical thinning and expansile remodelling;
- Eccentric, although very large lesions can appear central in location;
- Intact rim surrounding the lesion;
- Solid layer of periosteal response;
- Multiple internal septations;
- No mineralized matrix;
- Fluid-fluid levels.

Although conventional radiography is usually sufficient for evaluating the characteristics of the lesion, CT is particularly helpful in determining the integrity of the cortex and may also show internal ridges. The aneurysmal bone cyst contains a large amount of bloody fluid, which is why fluid-fluid levels can be seen on CT or MRI, representing the sedimentation of red blood cells and serum within the cystic cavities.

Differential diagnosis:

- Simple bone cyst;
- Chondromyxoid fibroma;
- Giant cell tumor;
- Chondromyxoid fibroma;
- Telangiectatic osteosarcoma.

The treatment for aneurysmal bone cyst consists of surgical removal and occasionally bone grafting. Recurrence of the lesion is frequent.

Images for this section:

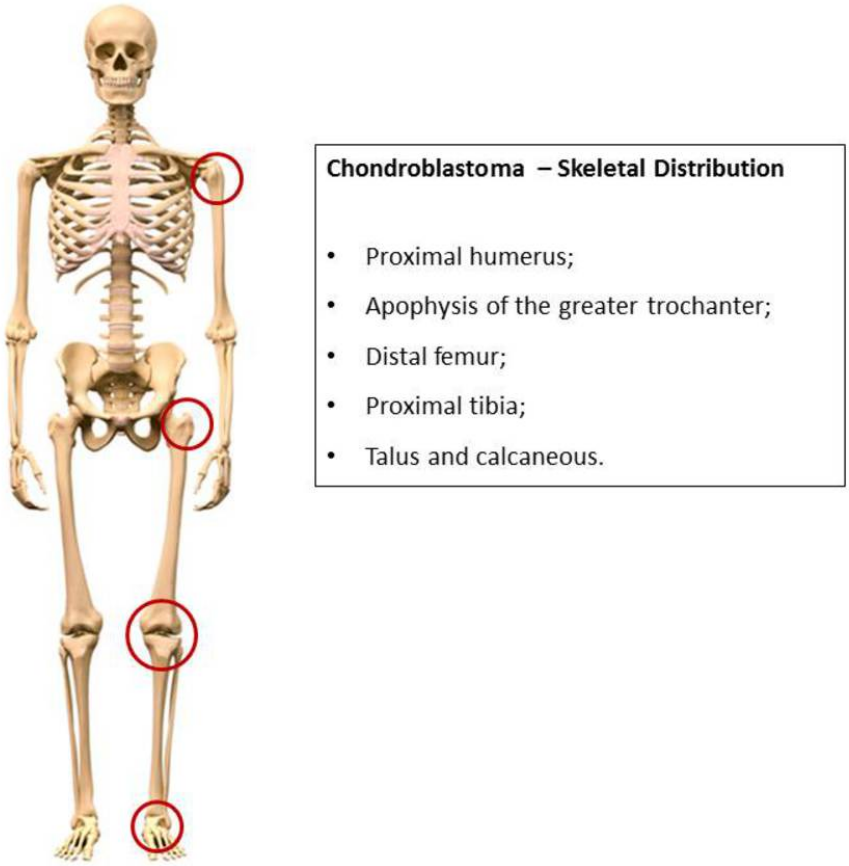


Fig. 13: Chondroblastoma - Skeletal Distribution.

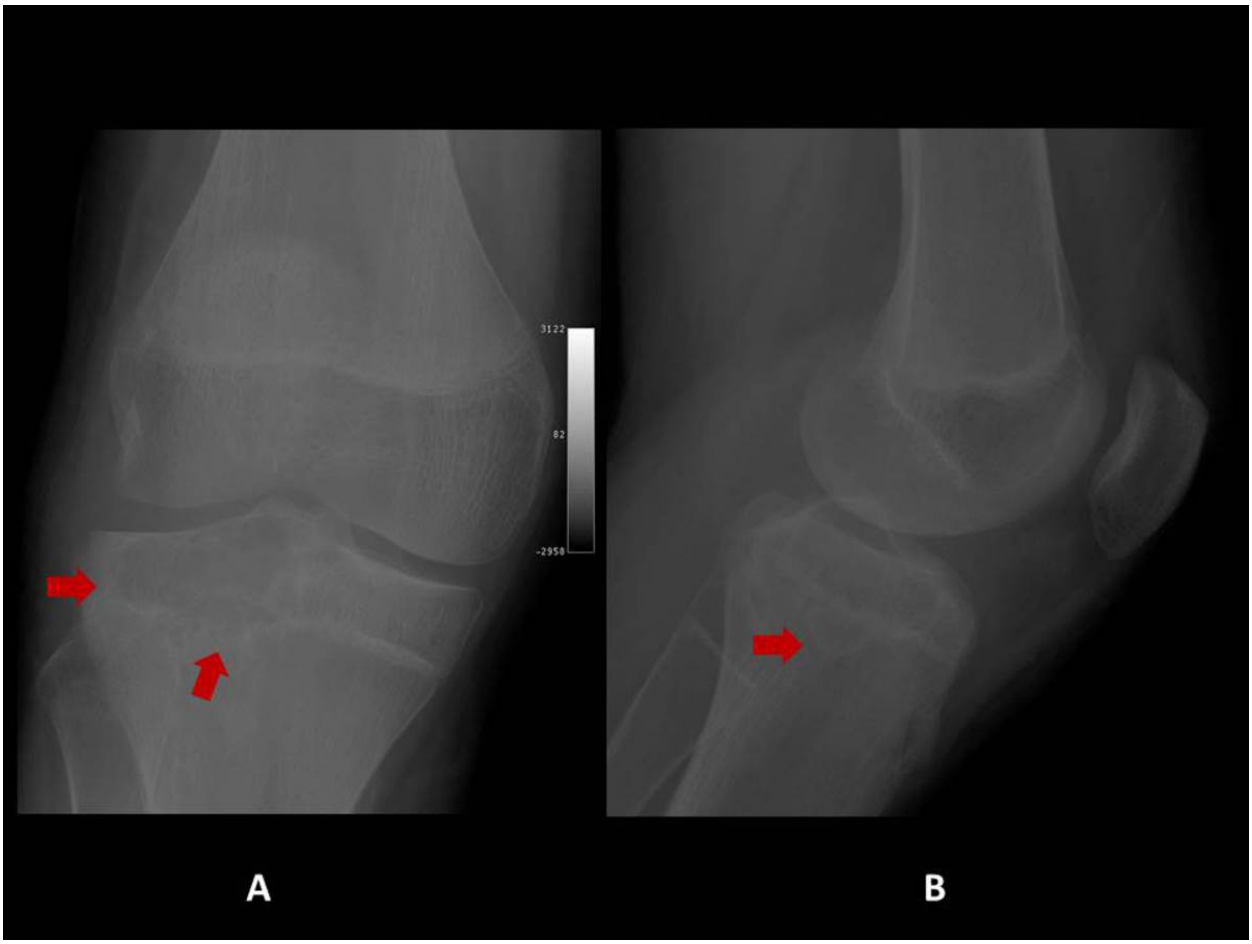


Fig. 14: Chondroblastoma. Anterior-posterior (A) and lateral (B) radiographs of the right knee reveal a well-demarcated lytic lesion in the proximal epiphysis of the right tibia surrounded by a thin sclerotic rim (arrows).

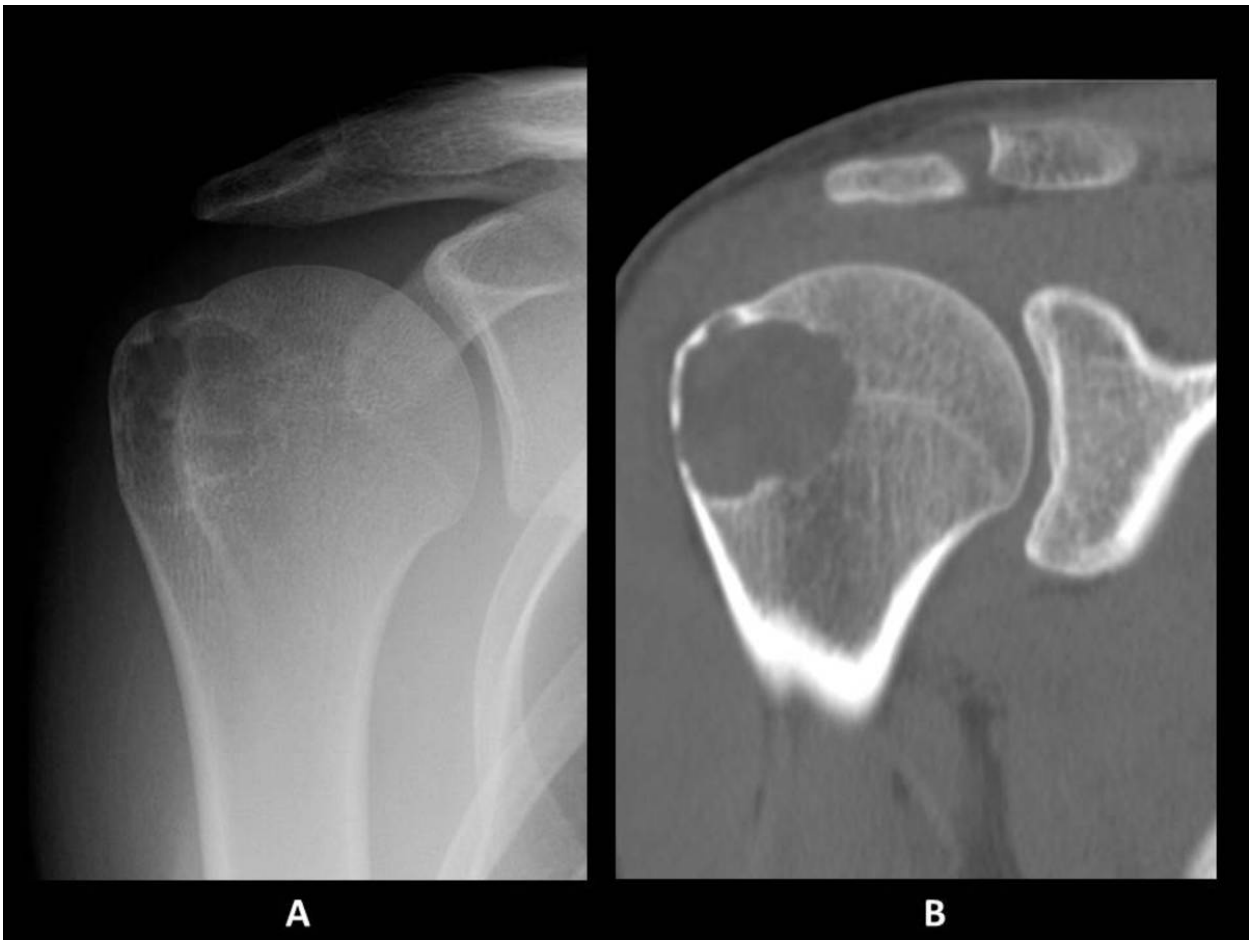


Fig. 15: Chondroblastoma. Anterior-posterior radiography (A) and coronal computed tomography reformation (B) of the proximal epiphysis of the right humerus reveal a geographic lytic lesion, with lobulated margins surrounded by a thin sclerotic rim. On CT it is not possible to observe calcification of the matrix, which may be absent in 40-60 % of cases.

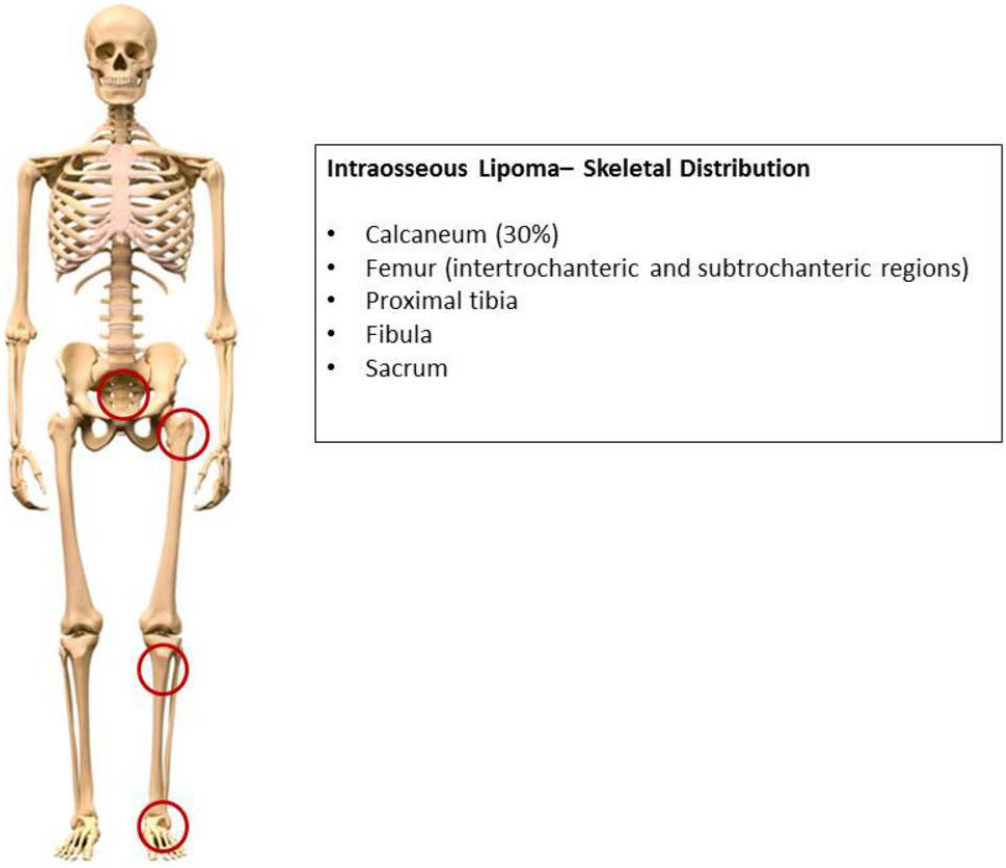


Fig. 16: Intraosseous Lipoma- Skeletal Distribution.

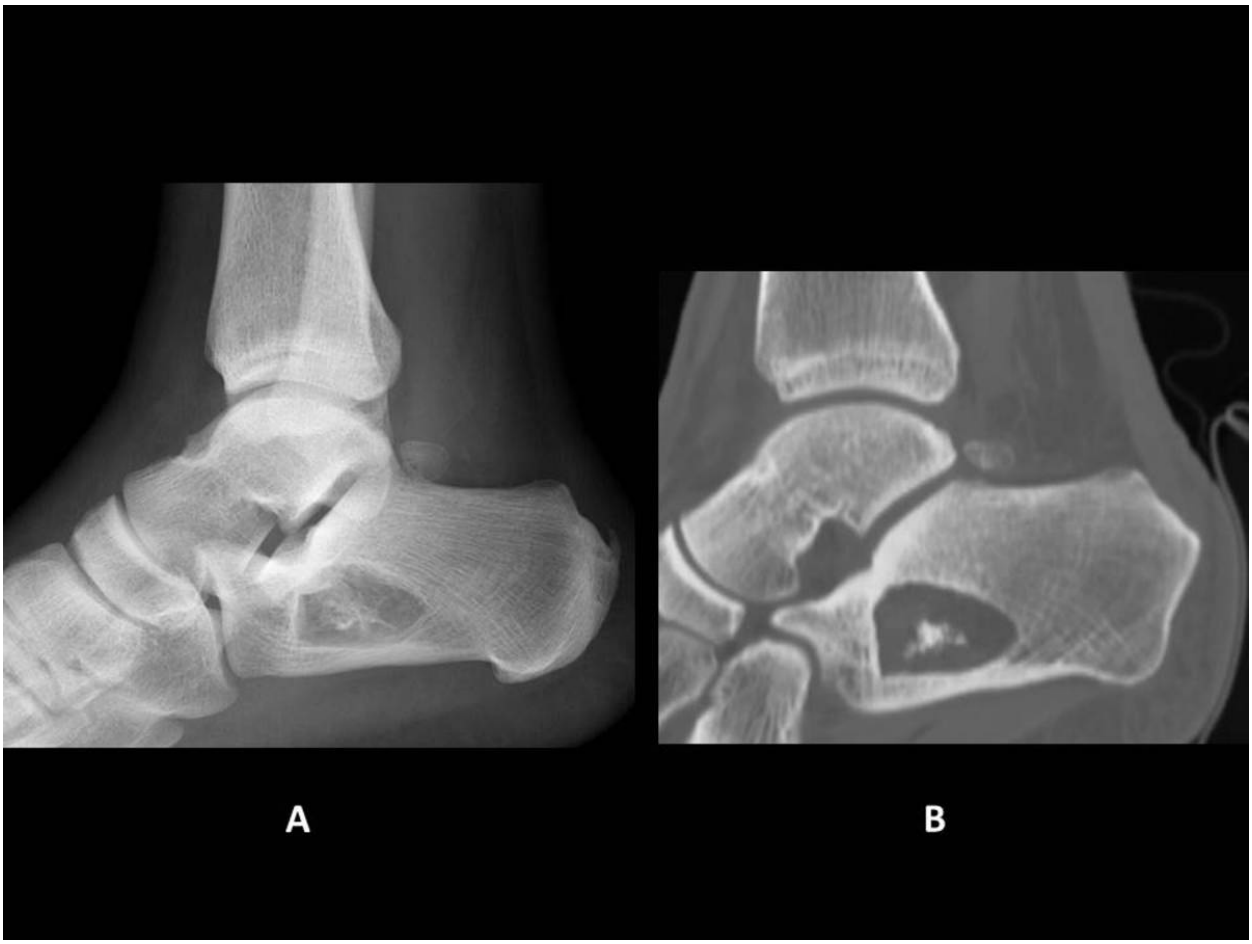


Fig. 17: Stage II intraosseous lipoma. Lateral radiograph (A) and sagittal computed tomography reformation (B) of the foot reveals a lucent lesion with a thin well-defined sclerotic border with central calcification involving the calcaneous. The area of lucency seen on the radiograph corresponds to fat attenuation visible on CT.

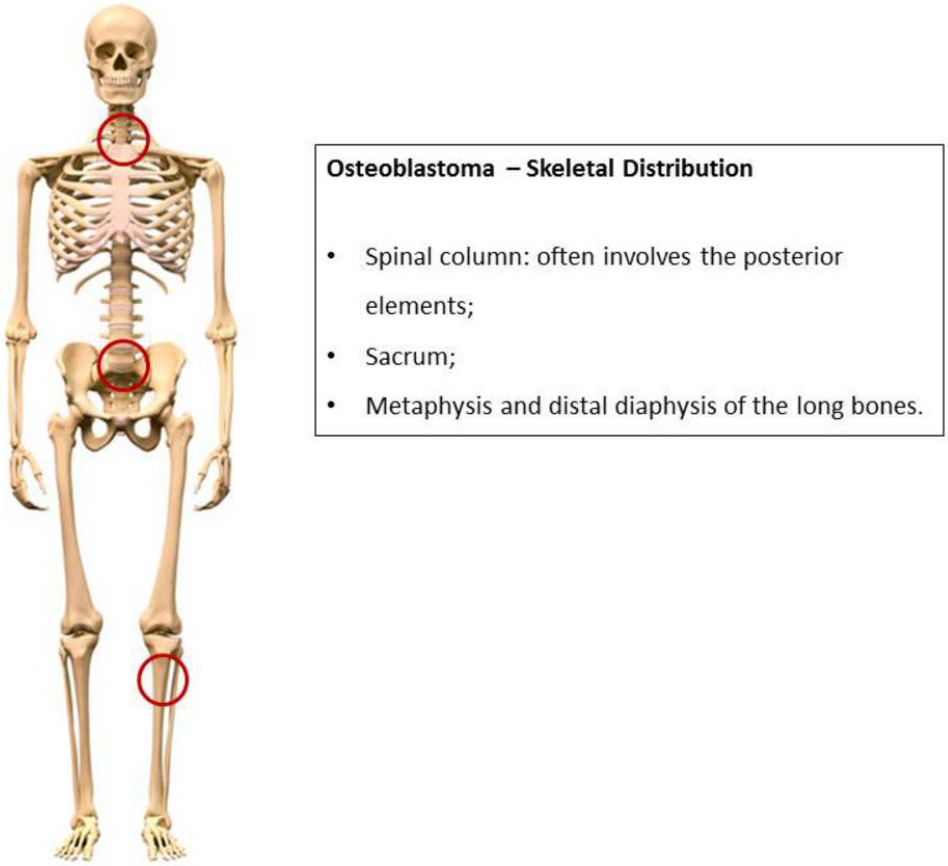


Fig. 18: Osteoblastoma - Skeletal Distribution.

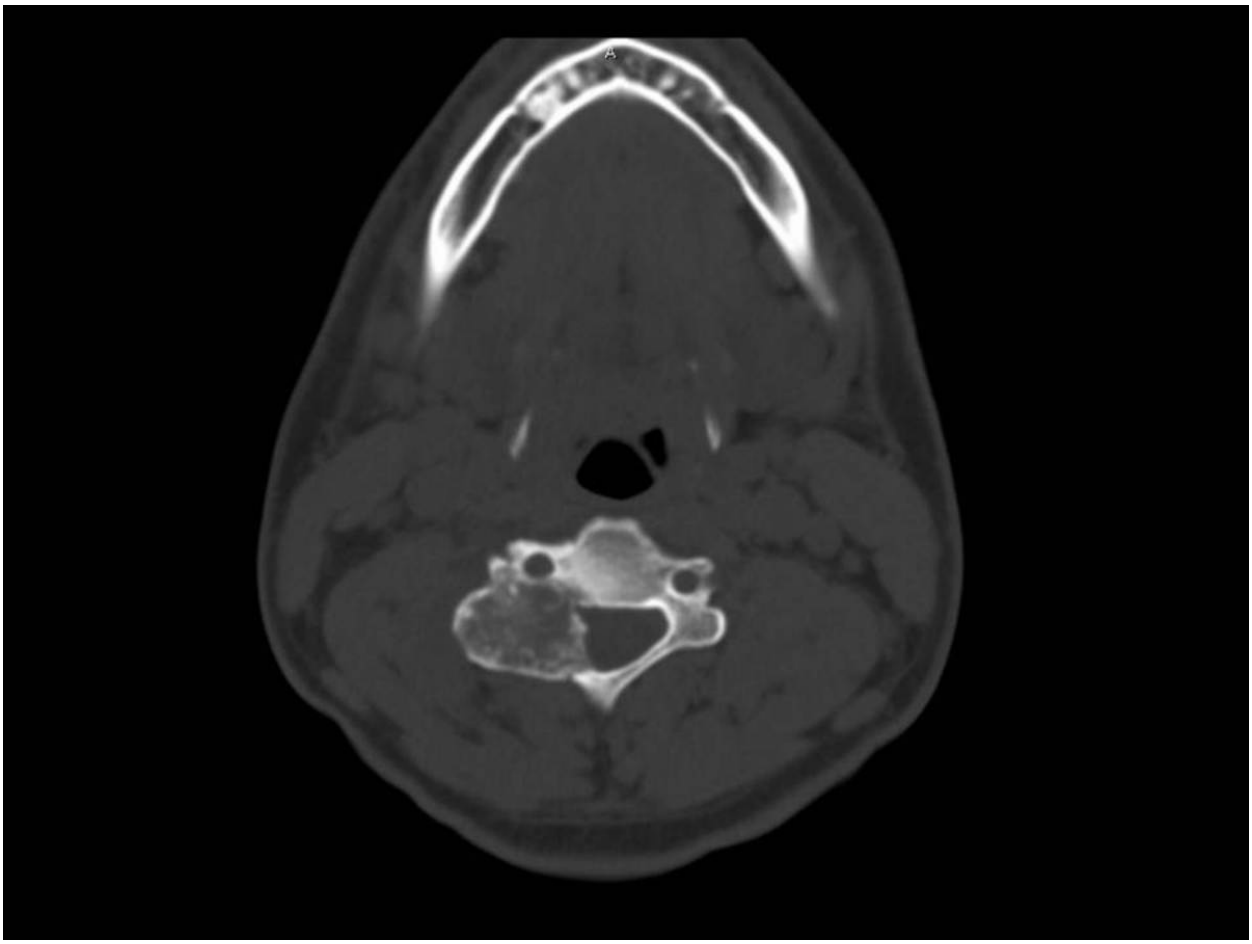


Fig. 19: Osteoblastoma is seen on computed tomography in the posterior cervical spine (lamina), demonstrating marked bone expansion with central mineralization.

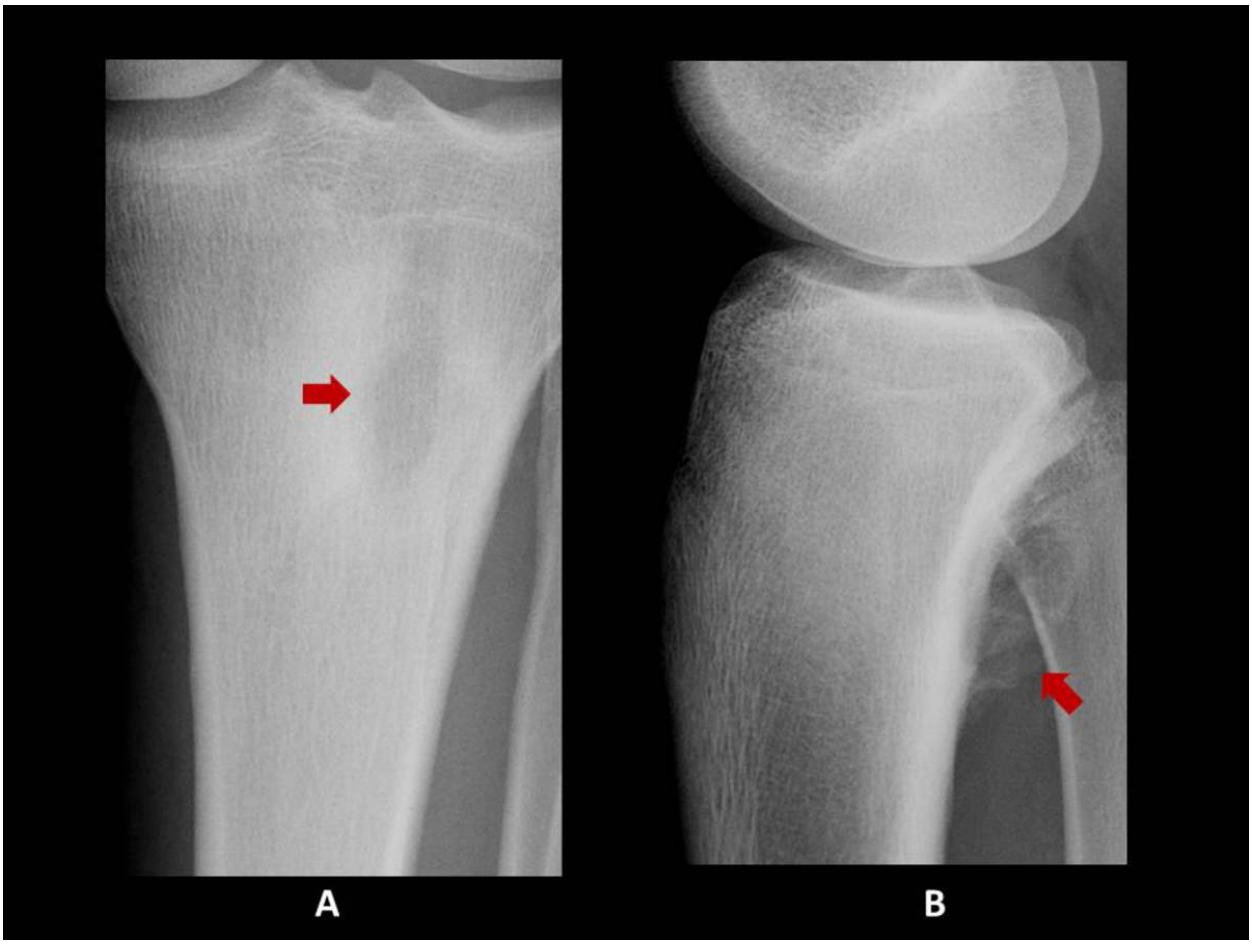


Fig. 20: Osteoblastoma. Anterior-posterior (A) and lateral (B) radiographs of the proximal left tibia reveals a geographic lytic lesion in the posterior tibia with cortical expansion and internal matrix mineralization (B, arrow).

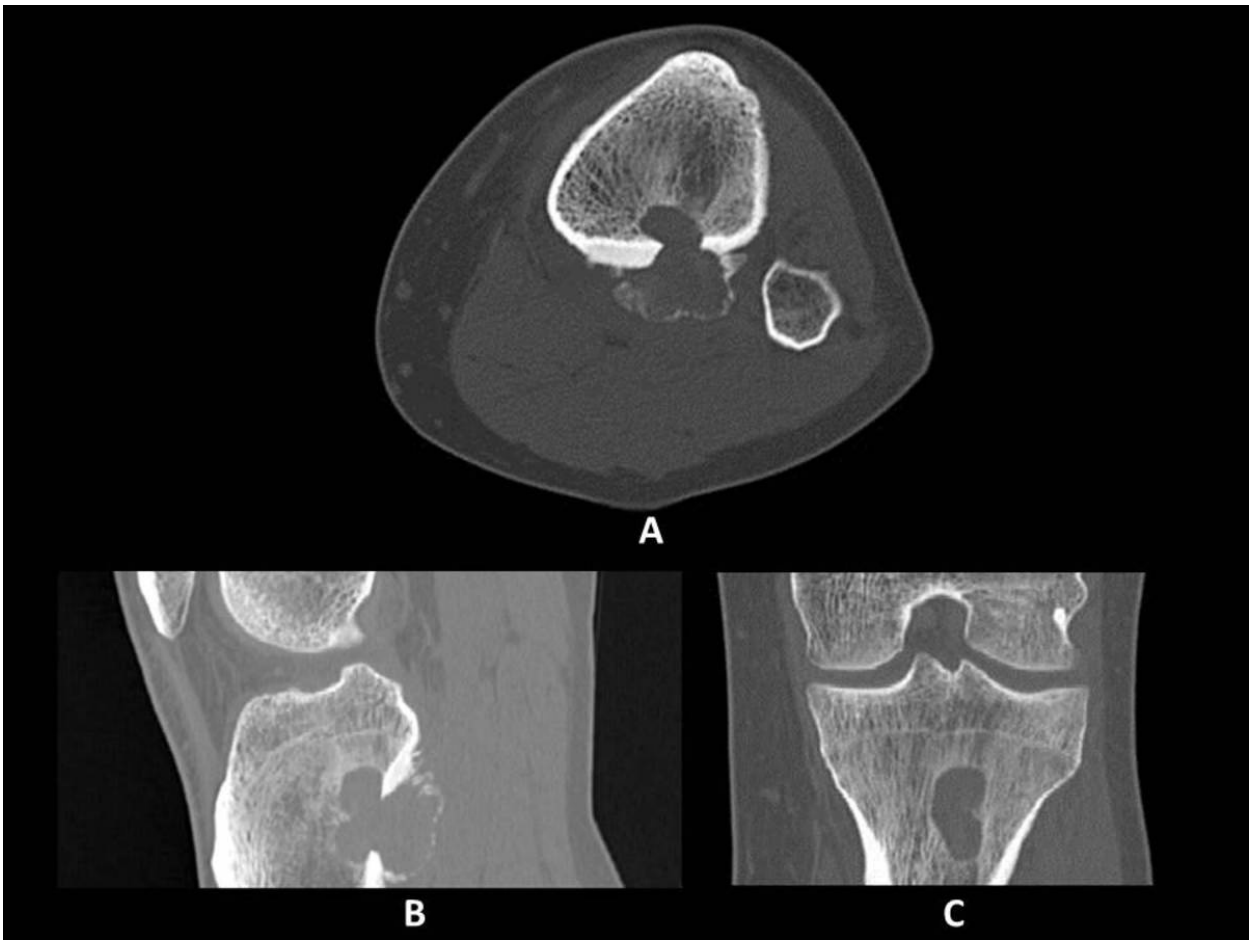


Fig. 21: Osteoblastoma. Same bone lesion seen in figure x. Similar to the radiograph, lesion is predominantly lytic, with cortical expansion and cortical destruction, latter is better appreciated on CT.

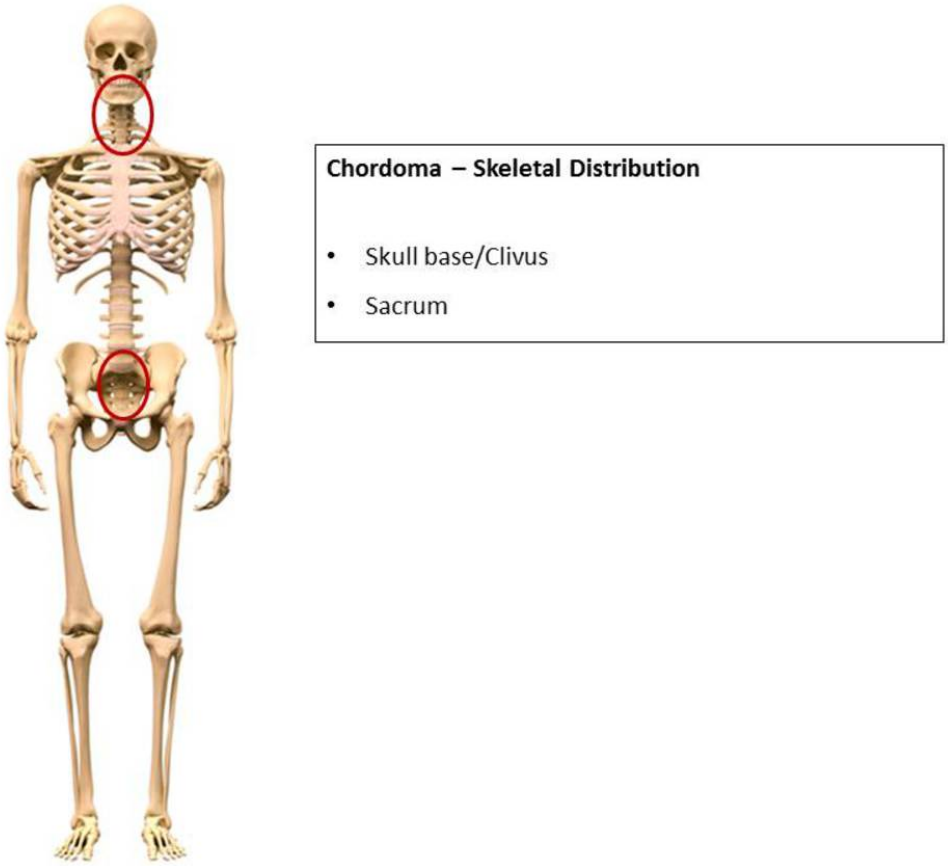


Fig. 22: Chordoma - Skeletal Distribution.

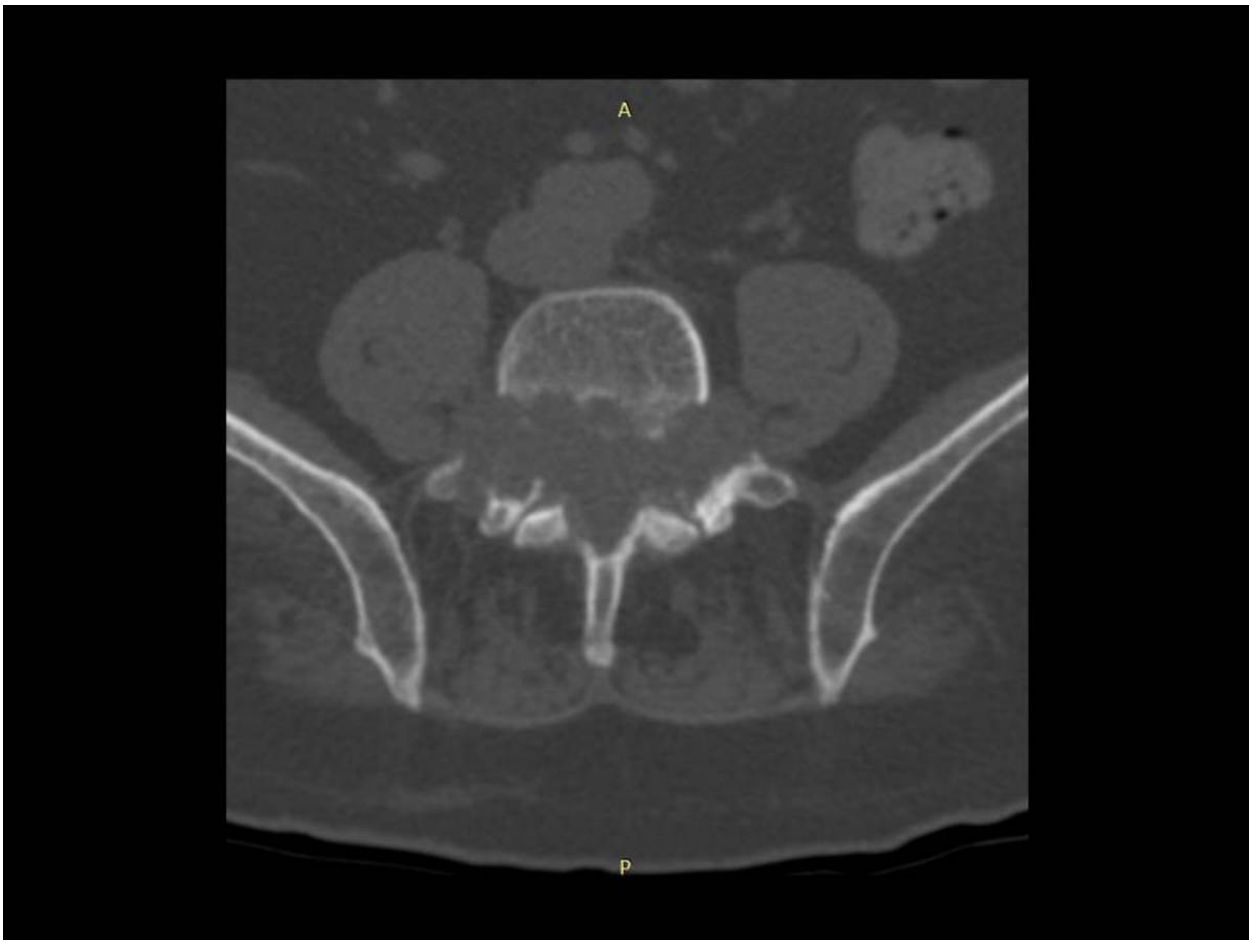


Fig. 23: Chordoma. CT shows extensive bone destruction and a large soft tissue mass.

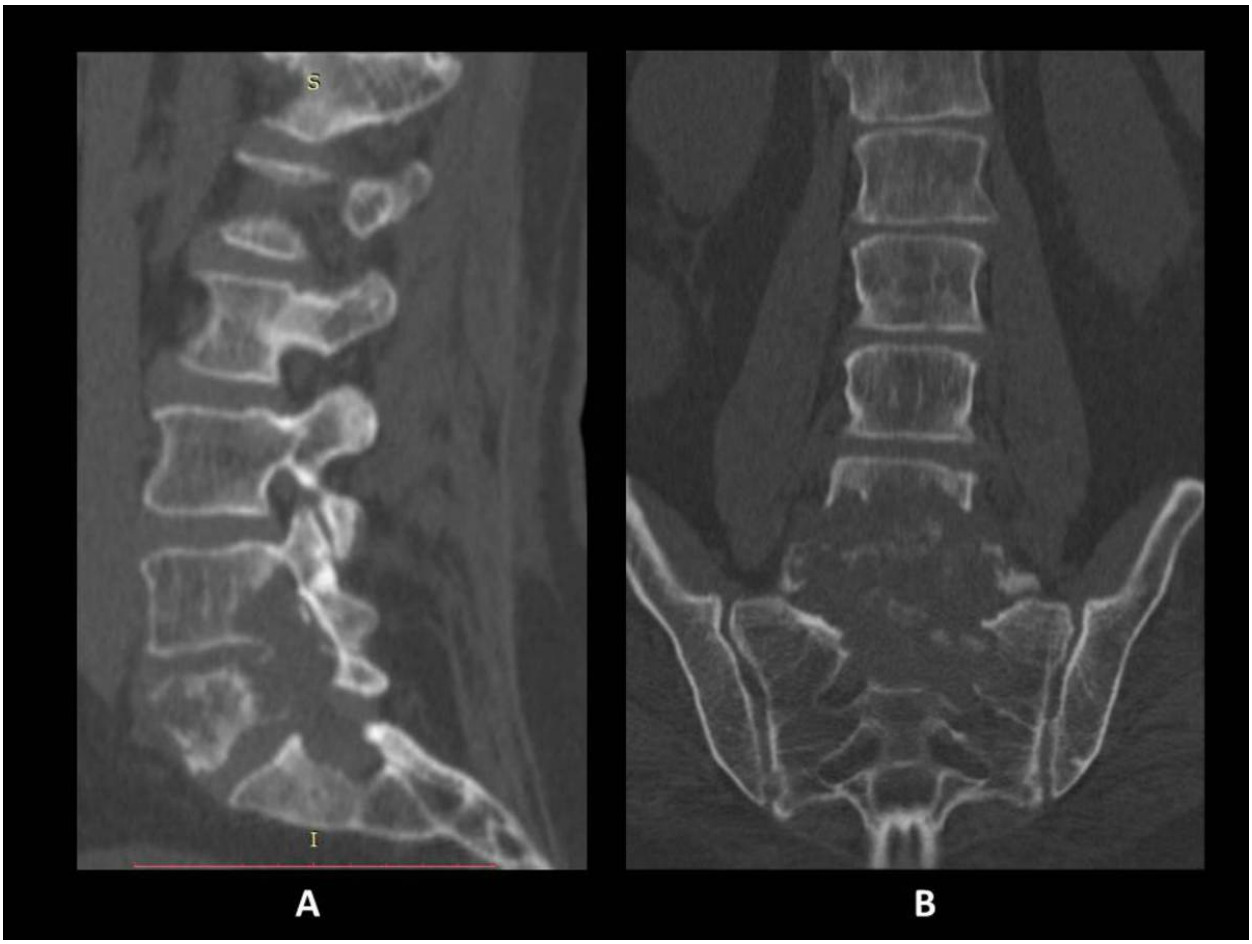


Fig. 24: Chordoma. Sagittal (A) and coronal (B) computed tomography reformation reveals expansive lesion with scalloped borders, with amorphous calcifications in the tumor matrix and partial destruction of the upper sacrum.

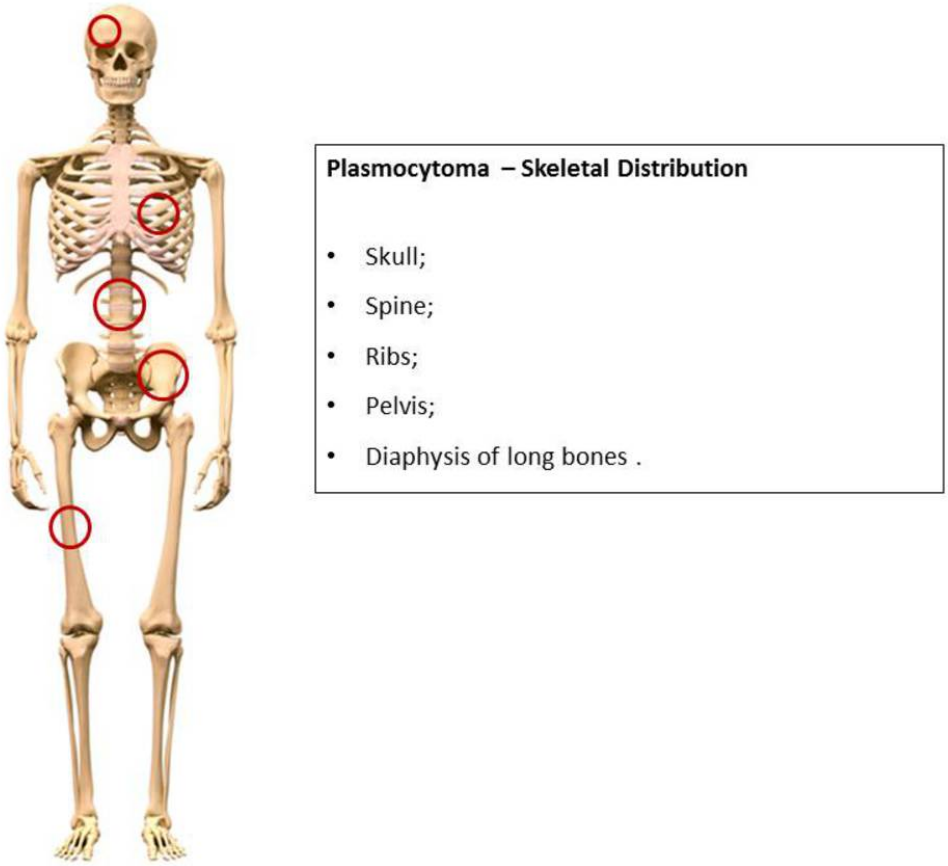


Fig. 25: Plasmocytoma - Skeletal Distribution.

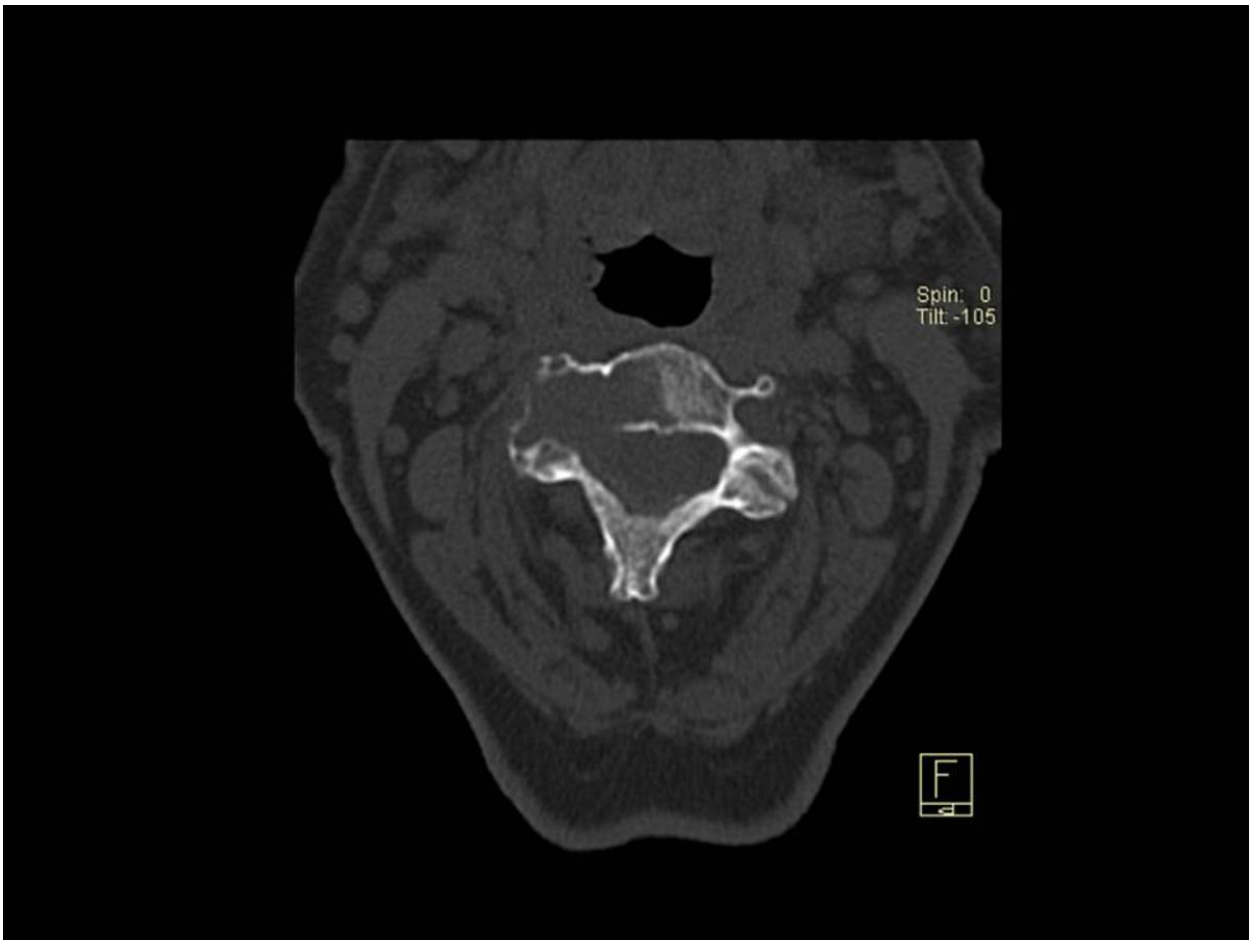


Fig. 26: Plasmocitoma. Expansive lytic lesion is seen on computed tomography in the anterior cervical spine. Evident marked erosion, expansion and cortical destruction. No matrix mineralization.

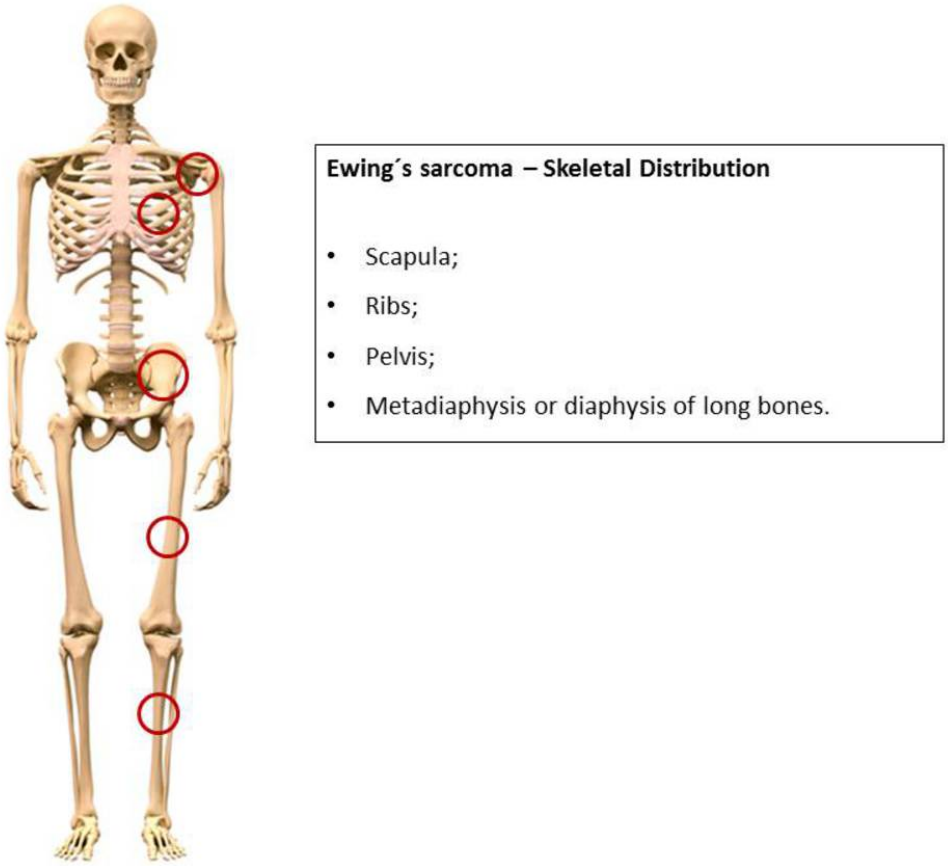


Fig. 27: Ewing´s sarcoma - Skeletal Distribution.

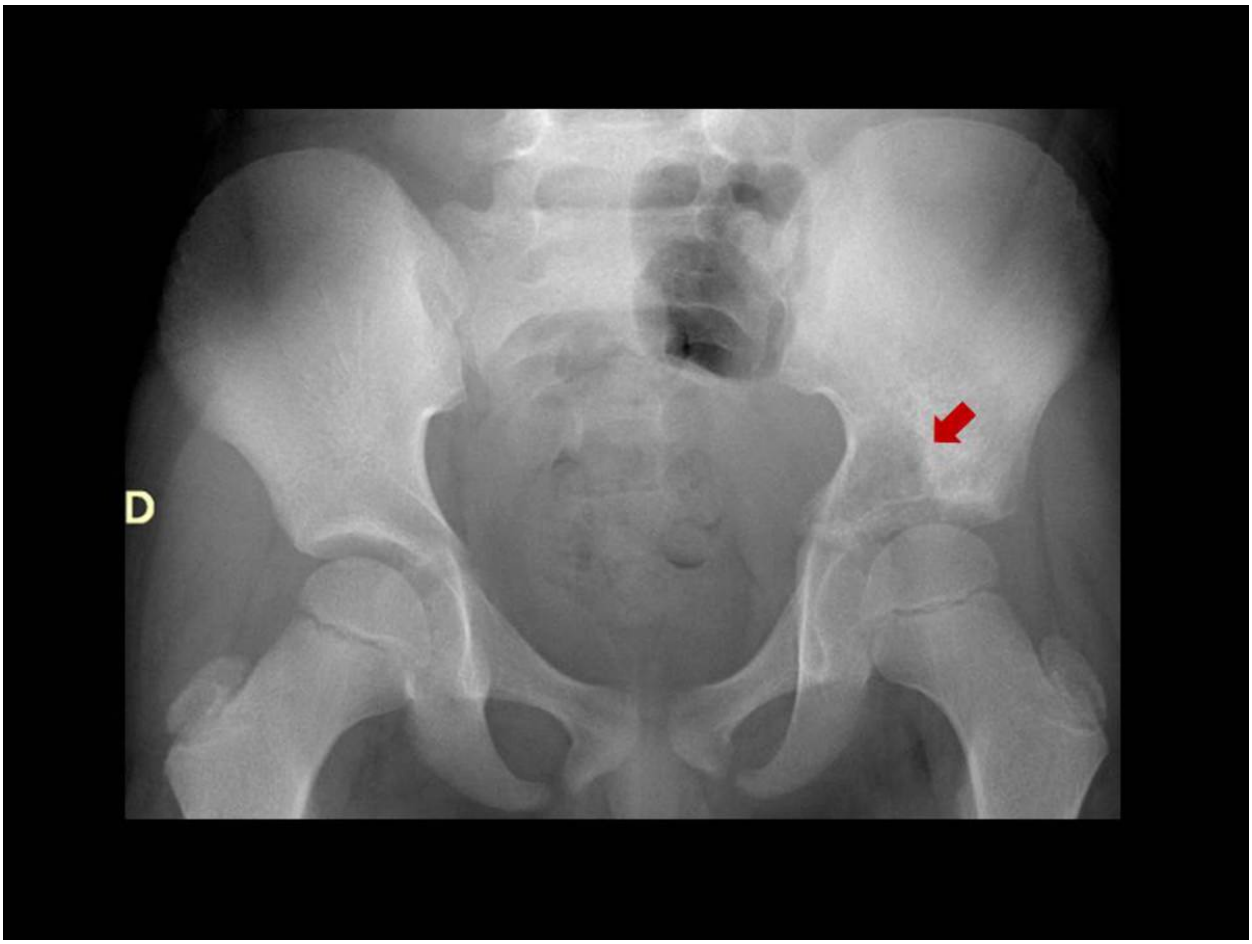


Fig. 28: Ewing sarcoma. Anterior-posterior radiography of pelvis shows ill-defined lytic lesion in the iliac bone (arrow).

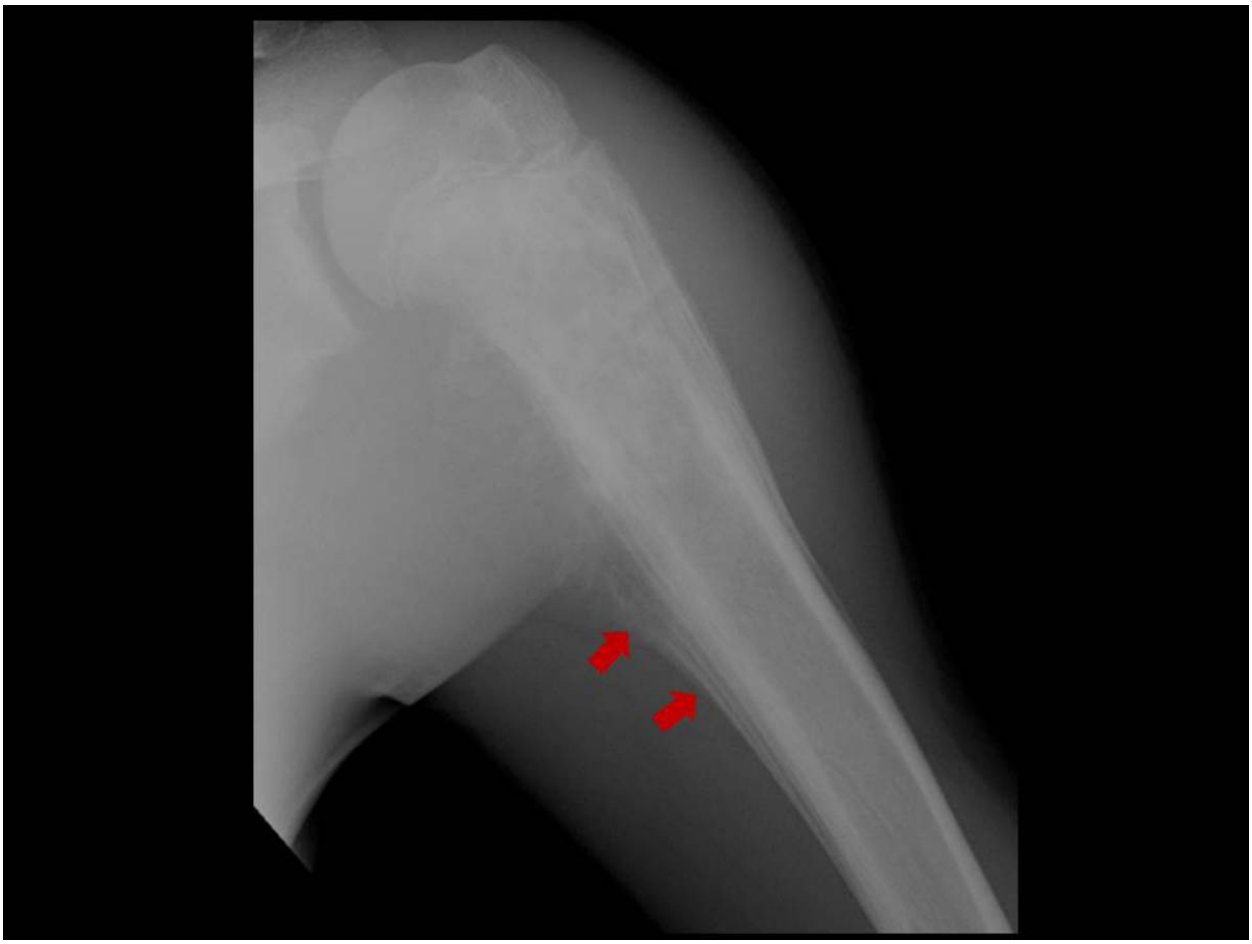


Fig. 29: Ewing sarcoma. Anterior-posterior radiography reveals a poorly defined osteolytic lesion in the proximal metaphysis of the left humerus, with permeative bone destruction and periosteal "sunburst" and "onion skin" like reaction.

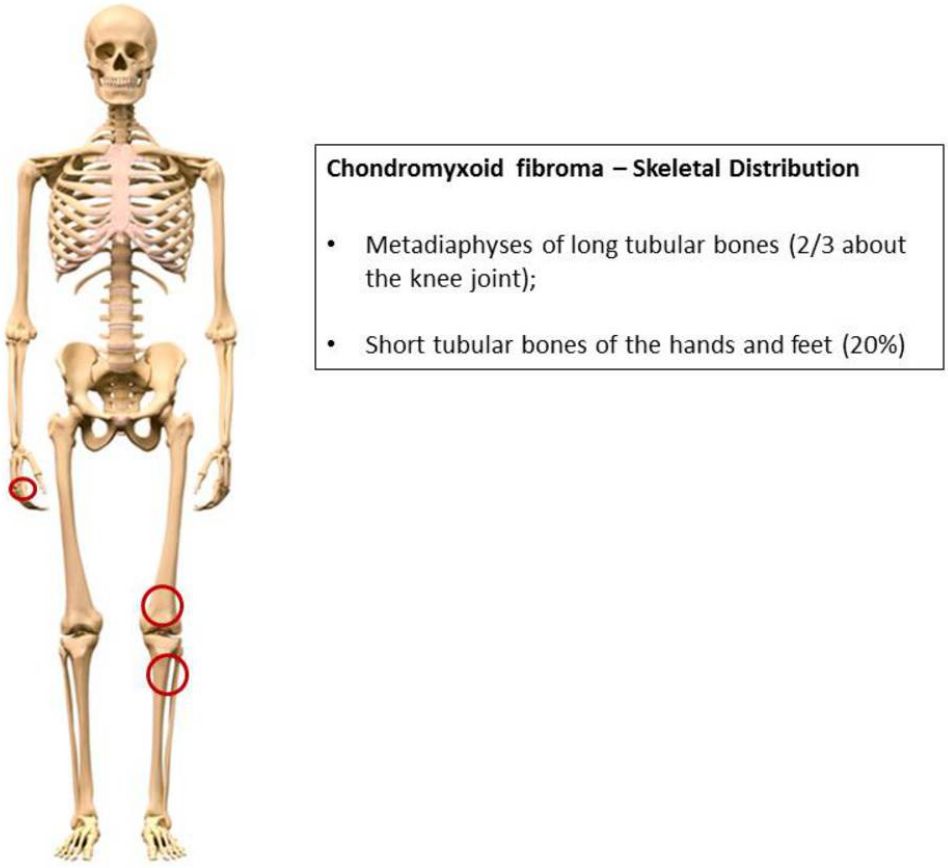


Fig. 30: Chondromyxoid fibroma - Skeletal Distribution.

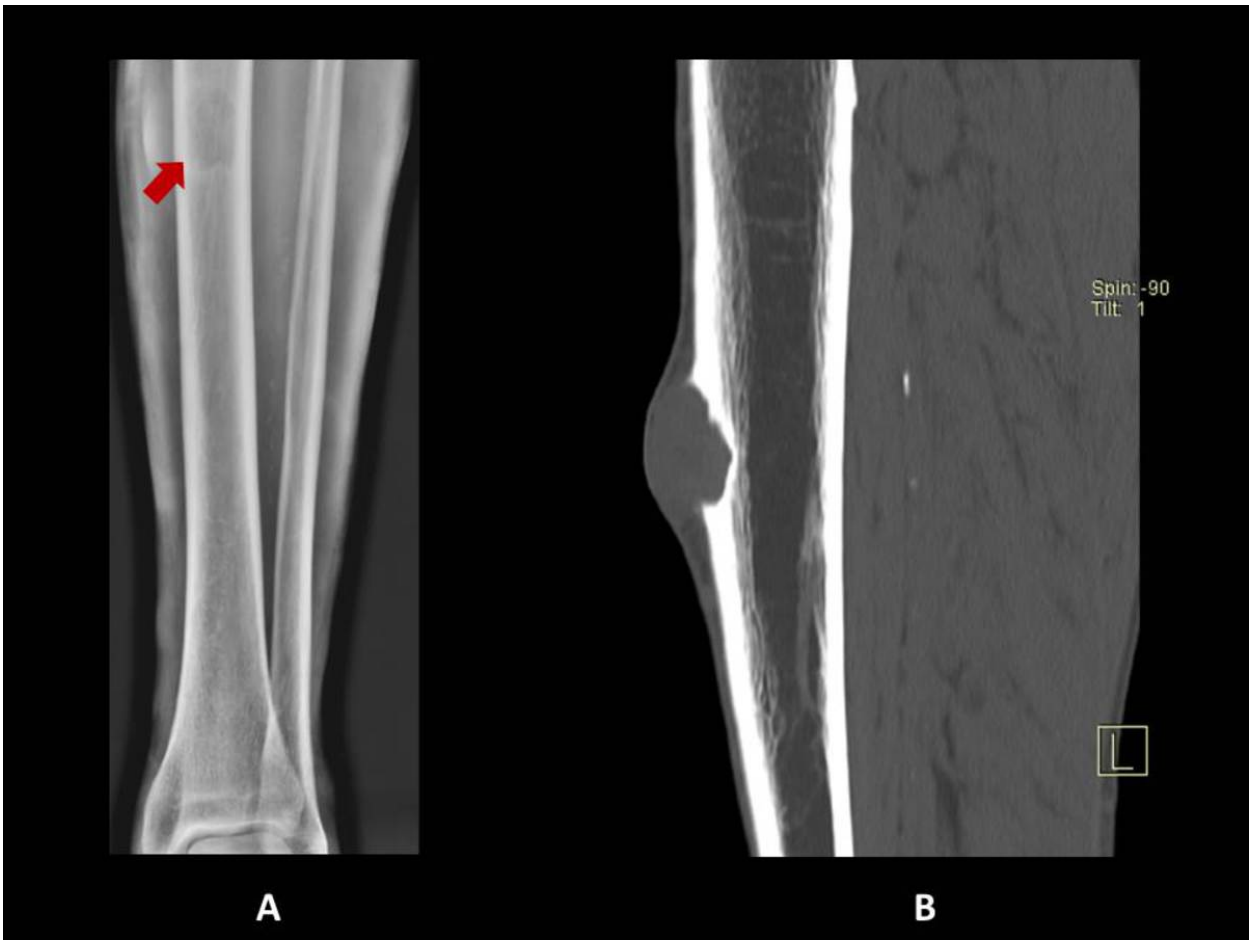


Fig. 31: Chondromyxoid fibroma. Anterior-posterior radiography (A) show a geographic lytic lesion in the diaphysis of left tibia. Sagittal computed tomography reformation (B) reveals an eccentric well-defined lytic lesion with sclerotic endosteal margin.

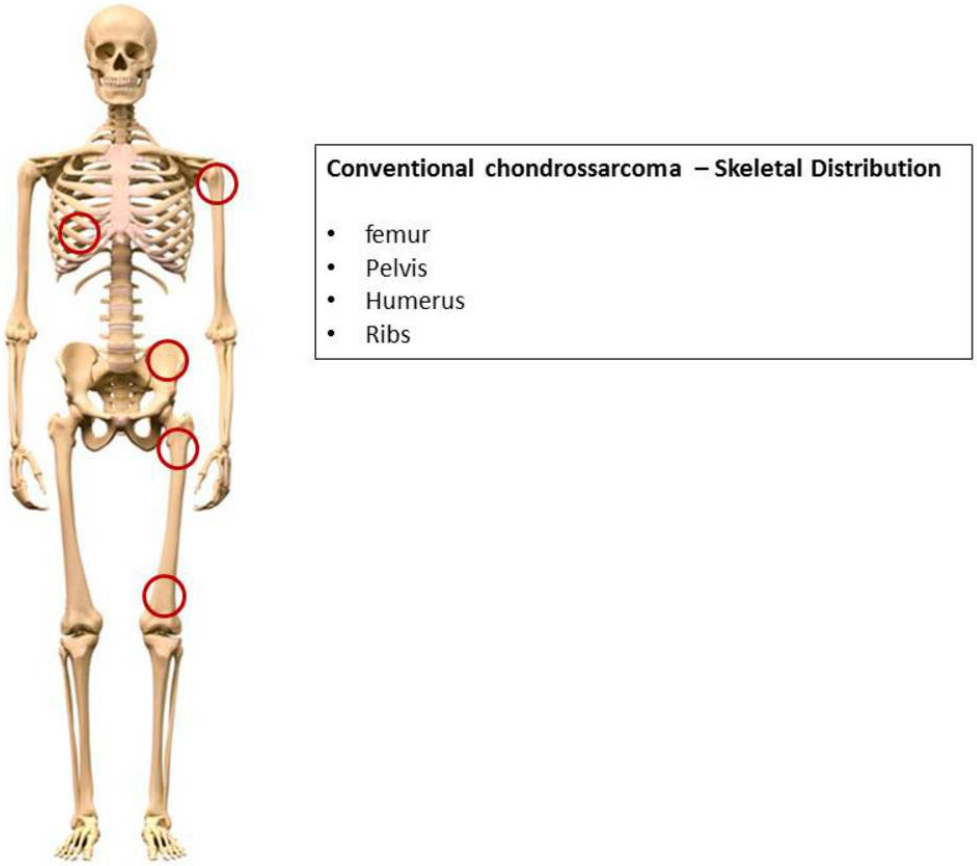


Fig. 32: Conventional chondrossarcoma - Skeletal Distribution.

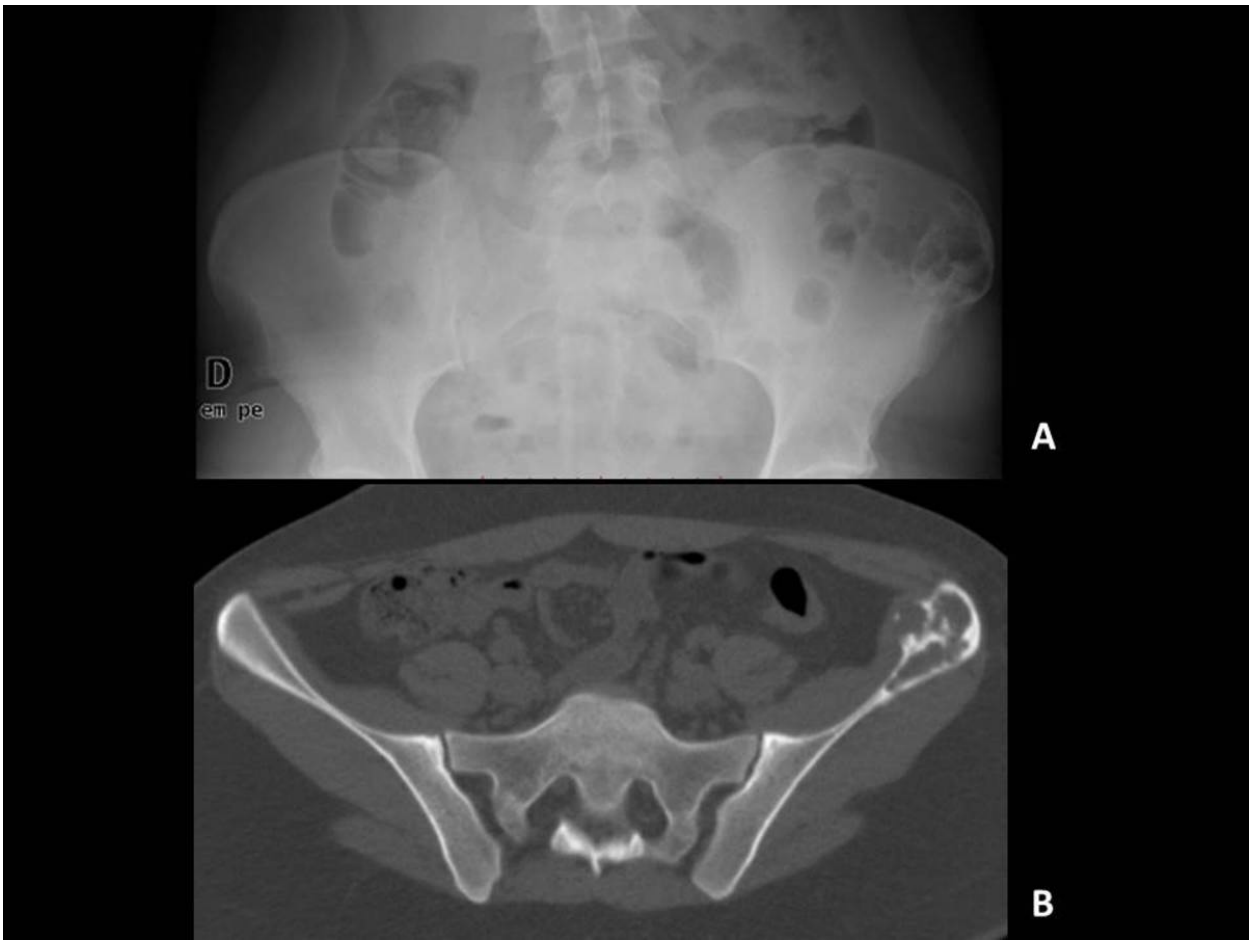


Fig. 33: Conventional chondrosarcoma. Anterior-posterior radiography (A) and CT (B) of the left iliac bone shows a lytic lesion with chondroid matrix mineralization and endosteal scalloping.

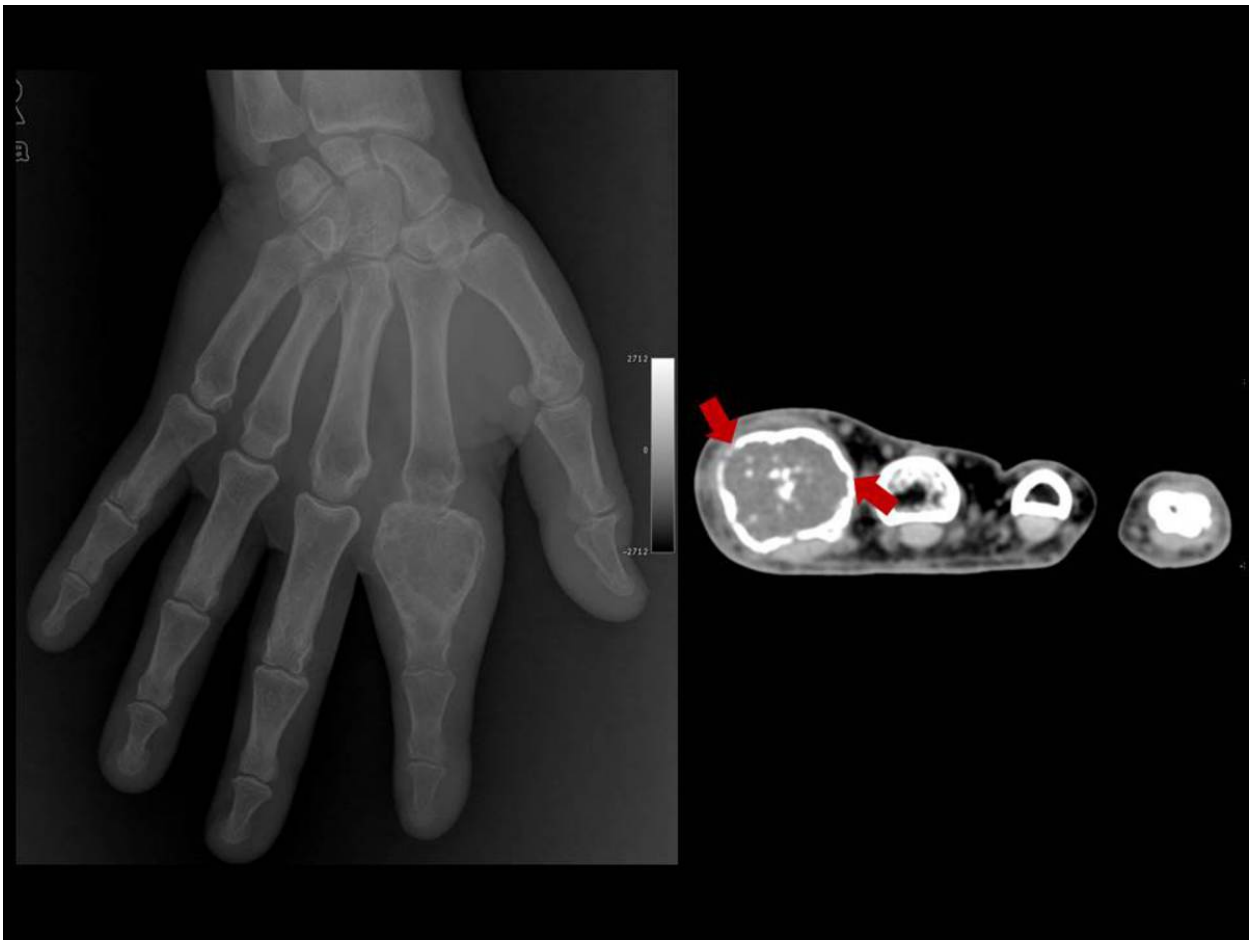


Fig. 34: Secundary chondrosarcoma on preexisting enchondroma. Note the endosteal scalopping (arrows), which is a vital feature in distinguishing low grade chondrosarcoma from enchondroma.

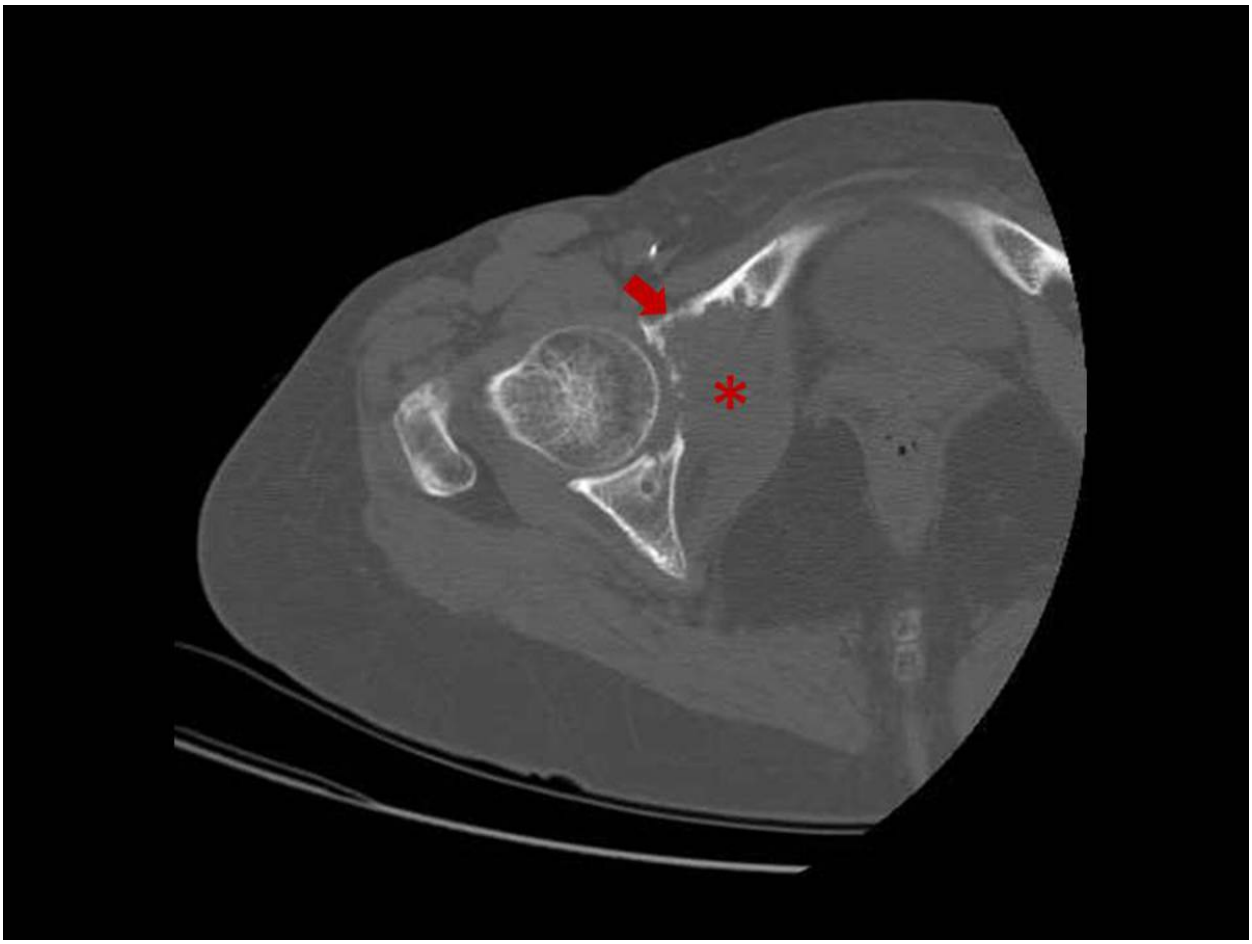


Fig. 35: Dedifferentiated chondrosarcoma. CT reveals a osteolytic lesion in the superior ramus of the right pubis with chondroid mineralization (arrow), which is a feature of a well-differentiated chondrogenic tumor. The cortex penetration and a soft tissue component (*) suggests an aggressive pattern.

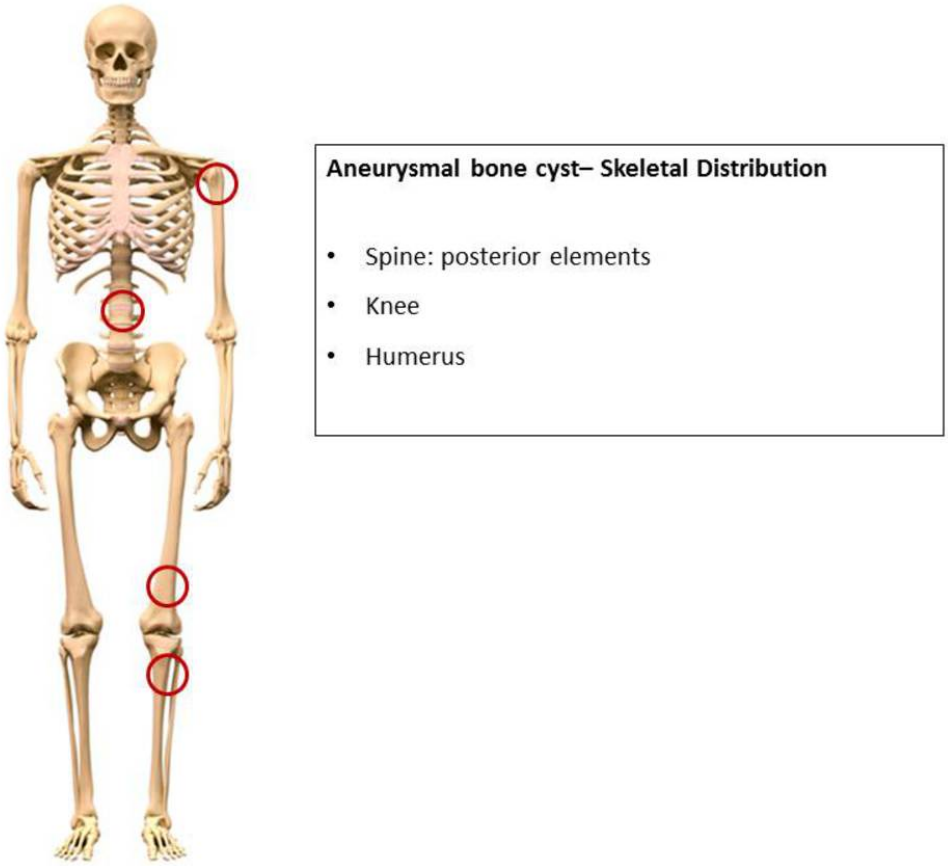


Fig. 36: Aneurysmal bone cyst- Skeletal Distribution.

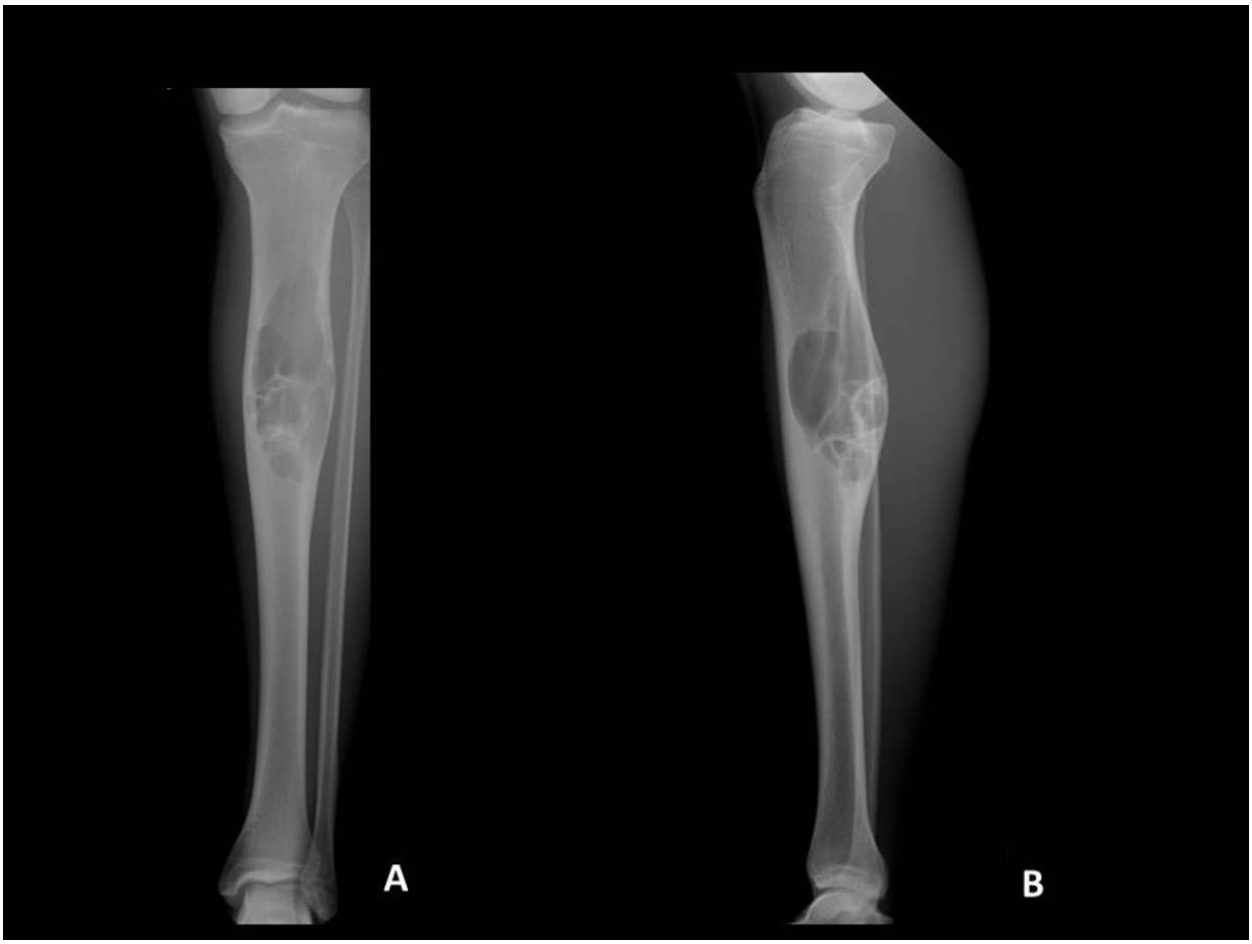


Fig. 37: Aneurysmal bone cyst. Anterior-posterior (A) and lateral (B) radiographs of the left tibia shows an expansive radiolucent lesion in the diaphyseal region of the tibia, eccentric in location, with a narrow zone of transition and the cortex is significantly thinned and bulging.

Conclusion

Bone tumors not always have clinical manifestations, which is why a lot of them are incidental findings on standard radiography, performed for other reasons. Conventional radiology is still the first step in the diagnostic assessment of a bone lesion, thus it is important that radiologists should be familiarized with typical location of bone tumors in the skeleton and their typical imagiologic features as well as the patient's age to help to ensure a presumptive diagnosis.

Personal information

References

- Greenspan A, Remagen W. Differential diagnosis of tumors and tumor-like lesions of bones and joints. Lippincott Williams & Wilkins (2004).
- Brant WE, Helms CA. Fundamentals of diagnostic radiology. Lippincott Williams & Wilkins (2012).
- Miller, Theodore. Bone Tumors and Tumor-like Conditions: Analysis with Conventional Radiography. Radiology: Volume 246: Number 3-March 2008.
- Wodajo, Felasfa M., MD. Visual Guide to Musculoskeletal Tumors A Clinical - Radiologic - Histologic Approach. Saunders 2010.
- Milgram J. W. Intraosseous Lipomas: Radlologic and Pathologic Manifestations; Radiology 1988; 167:155-160.