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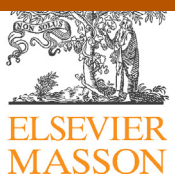

LETTER /*Musculoskeletal imaging*

Imaging features of systemic cystic angiomatosis

Keywords Diffuse angiomatosis; Angioma; Splenic nodules; Biphosphonates

Systemic cystic angiomatosis is a rare disease, which corresponds to disseminated angiomatous or lymphangiomatous cysts [1—4]. Although this disease mainly develops in the skeletal structures, visceral involvement can occur. The imaging features of systemic cystic angiomatosis can mimic and be mistakenly diagnosed as a primary carcinoma with disseminated secondary skeletal involvement. Histologically, the diagnosis is difficult. We report the case of a 57-year-old man with systemic cystic angiomatosis presenting with typical skeletal and splenic involvement, which was discovered incidentally.

Case study

A 58-year-old man presented to the emergency room for assessment of suspected infectious lung disease. Chest X-rays revealed an asymptomatic centimetric area of bone loss on the left rib. Chest and abdominopelvic computed tomography (CT) was performed as well as a whole body MR imaging examination (Fig. 1). They revealed multiple areas of bone loss mainly in the axial skeleton and the ribs. The appearance of the lytic lesion suggested a ''pin head'' arrangement in certain areas. MR imaging showed splenomegaly associated with numerous splenic nodules presenting with heterogenous enhancement. Serum protein electrophoresis was normal and there was no inflammatory syndrome. PET-CT showed non-specific, moderately increased metabolic activity in certain areas of bone loss. Three PET-CT guided biopsies, which were at first inconclusive, finally suggested the diagnosis of systemic cystic angiomatosis (Fig. 2). Histopathologically the lesions corresponded to lymphangiomatous-type dilation of the vascular spaces (Fig. 3). No treatment was initiated. The patient, who was asymptomatic, will undergo splenic ultrasound and radiographic bone assessment every 6 months as long as he remains asymptomatic.

Discussion

Systemic cystic angiomatosis is a rare entity that is mainly characterized by disseminated and multifocal lytic lesions, but which may be associated with visceral involvement. Several organs may be affected, including the lungs, kidneys, spleen, colon, small bowel and skin [1—3]. The exact pathogenic etiology of this disease is unknown; it seems to be associated with sequestration of liquid in the lymphatic system during embryogensis [5]. Skeletal involvement is predominant.

Systemic cystic angiomatosis has been referred to either angiomatosis or lymphangiomatosis. This is because vascular dilations may contain blood or lymph [4,5].

CT usually shows vertebral involvement with bone loss and osteosclerotic radiolucencies. In the axial plane, CT

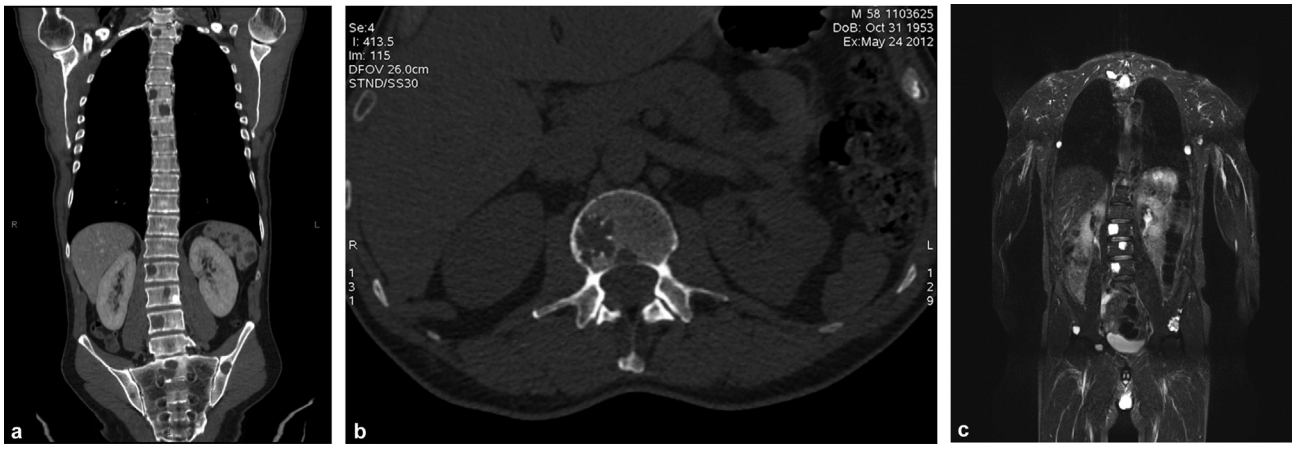

Figure 1. Whole body CT and MR imaging examination: a: in the coronal plane, CT shows lytic vertebral and rib lesions with peripheral osteosclerosis. Multinodular splenic involvement is present; b: in the axial plane at the level of L1 vertebra, CT shows a lytic lesion with ''pin head'' arrangement in certain areas suggesting a vertebral angioma; c: fat-suppressed T2-weighted MR image (TE: 64; TR: 4000; Ti: 150) in the coronal plane shows multiple lytic bone lesions displaying hypersignal associated with multinodular splenic involvement.

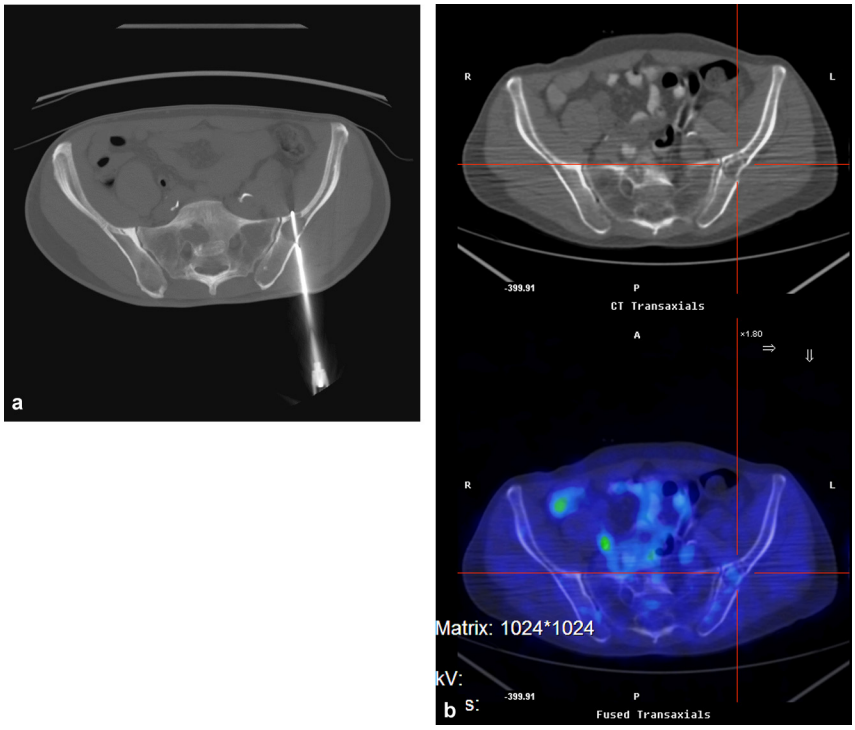

Figure 2. 18 FDG PET-CT guided biopsy (a) of a lytic lesion with moderate uptake on PET-CT allowed definite diagnosis (b).

shows dense bone with areas of bone loss as a ''pin head'' arrangement. In the sagittal plane, CT shows ''vertical stripes''.

MR imaging usually shows hypersignal on T1- and T2-weighted images in relation to the presence of fat as well as mild enhancement after IV administration of a gadolinium chelate [6].

Lesions are non-specific on 18-FDG PET-CT, with no metabolic activity and even moderately increased non-pathological metabolic activity.

The possible differential diagnoses in the presence of disseminated lytic angiomatous features include histiocytosis X, hyperparathyroidism, polyostotic fibrous dysplasia, metastases and lymphomas [4,7,8]. Nevertheless for all these conditions, the radiolucent area of osteosclerosis is often absent and the typical angiomatous presentation described above is not present.

Visceral involvement is more rare and can be life threatening. In our patient, splenic involvement may cause splenomegaly, coagulation disorders and even life-threatening splenic rupture. On MR imaging and CT splenic involvement usually presents as multiple slightly enhancing nodules [9].

Patients with systemic cystic angiomatosis are often asymptomatic and the disease is often discovered incidentally following a pathological fracture revealing disseminated skeletal involvement, which is at first mistakenly thought to be neoplastic.

The diagnosis of systemic cystic angiomatosis is obtained from a bone biopsy. Nevertheless, as reported in the literature and confirmed in our patient, the histological diagnosis is difficult, and several biopsies are often needed to reach the final diagnosis [4,6]. The main role of imaging is thus to suggest the diagnosis to guide the pathologist thus avoiding invasive procedures and their associated risks.

There is no confirmed treatment of systemic angiomatosis to date. Some authors have reported treatment with biphosphonates [10]. In our patient, we have decided to perform biannual ultrasound monitoring of the spleen and annual radiological assessment of bone lesions. The patient is asymptomatic at present, but the splenic visceral involvement makes the long-term prognosis guarded.

In conclusion, systemic cystic angiomatosis is a rare disease but a careful analysis of patient history and imaging features help suggest the diagnosis which is often difficult to confirm on histology.

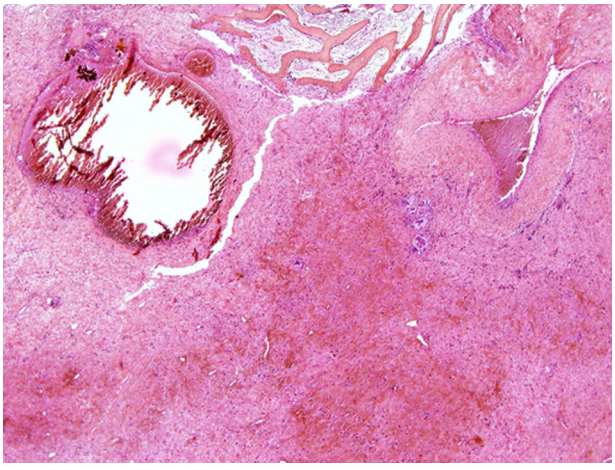

Figure 3. Histopathological analysis of tissue sample obtained from PET-CT guided biopsy (HES, original magnification × 20) shows dilated lymphangiomatous vascular spaces.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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W. Marraoui[a,*], J.-L. Michel[b], J.-L. Kemeny[c], M. Soubrier[d]

[a] *Service de radiologie et imagerie médicale, centre Jean-Perrin, CLCC Clermont-Ferrand, 58, rue Montalembert, 63000 Clermont-Ferrand cedex 1, France*

[b] *Service de radiologie et imagerie médicale, CHU Gabriel-Montpied, CHU de Clermont-Ferrand, 58, rue Montalembert, 63000 Clermont-Ferrand cedex 1, France*

[c] *Service d'anatomie et cytologie pathologique, CHU Gabriel-Montpied, CHU de Clermont-Ferrand, 58, rue Montalembert, 63000 Clermont-Ferrand cedex 1, France*

[d] *Service de rhumatologie, CHU Gabriel-Montpied, CHU de Clermont-Ferrand, 58, rue Montalembert, 63000 Clermont-Ferrand cedex 1, France*

* Corresponding author. UFR médecine, université Clermont 1, 63001 Clermont-Ferrand, France.

E-mail address: Marraouiwissam@yahoo.fr (W. Marraoui)

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