CASE REPORT

Multiple Osteomas of Mandible in a Patient with Gardner's Syndrome - Report of a Case.

Arun George, Sankar Vinod Vichattu, Joy R Das, Atic Thomas, Jinu Elizabeth James

Abstract

Introduction: Osteomas are benign tumors of the bone usually seen in craniofacial bones which arise from either the cortex or medulla. They are usually asymptomatic with slow painless growth. Multiple osteomas are often associated with Gardners syndrome. Solitary osteomas are typically non syndromic.

Case Presentation: The patient presented with remarkable facial asymmetry following the growth in the mandible. Multiple osteomas were noted which invoked suspicion of a syndrome. On endoscopic evaluation the patient was found to have multiple intestinal polyps, pathgnomonic for Gardners syndrome.

Management and prognosis: The lesion was surgically removed under local anaesthesia. as an excisional biopsy and the asymmetry was rectified. Histopathologic examination revealed it to be ivory osteoma with dense compact bone and less marrow spaces. The jaw lesions would precede the development of colonic polyps and therefore may contribute to early diagnosis of Gardner's syndrome. There is also a chance of malignant evolution associated with the polyps

Conclusion: People with the condition have a higher risk of developing other family adenomatous polyp (FAP)related cancers including pancreatic cancer and liver cancer. The patient should be in close follow up

Keywords: Benign, Facial asymmetry, FAP, Gardners Syndrome, Intestinal polyps, Malignancy

Oral and Maxillofacial Pathology Journal (2023): ***https://www.ompj.org/archives***

Introduction

Osteoma is a benign, slow growing, painless growth that is usually seen in bones formed by membranous ossification. They usually develop on the surface of the craniofacial skeleton and have a propensity to involve the bones of the paranasal and frontal sinuses and the orbit where they are found in 1% to 3% of the adult population. Rarely, osteomas are located in the vertebral column and the appendicular skeleton where they develop on the surfaces of long tubular bones.[1] They are usually composed of compact mature bone. They are mostly seen in the skull, facial and jaw bones with a predilection to paranasal sinuses. When seen as multiple osteomas in the craniofacial skeleton, they raise a strong suspicion of Gardners syndrome. component of Gardners syndrome. Gardners syndrome is a combination of intestinal polyps mainly in the colon and lower gastrointestinal tract, multiple bony osteomas especially in the jaw bones, supernumerary or unerupted tooth and epidermoid cysts.[2]

Because osteomas of the bones, including the jaws may be the initial symptom or the clinical finding in the Gardner's syndrome, this entity should always be included in the differential diagnosis.[3] Gardner and Richards described the syndrome along with its genetic pattern in 1953.

Osteomas might be composed of either dense compact bone with scanty marrow spaces (compact/ivory osteoma) or cancellous bone with abundant bony trabeculae along with fibro-fatty marrow tissue (cancellous/trabecular/spongy osteoma).[4] Based on its site of origin they can be classified as central (arising from the medullary cavity) or peripheral (arising from the cortex).[5]

In view of the finding of a mutation in gene adenomatous polyposis coli (APC) located on the long arm of chromosome[5] in both Familial Adenomatous Polyposes (FAP) and Gardner's syndrome, the latter was considered as a variant

Department of Oral and MaxilloFacial Surgery, Mar Baselios Dental college, Kothamangalam, Ernakulam- 686 691

Corresponding author: Joy R Das, Department of Oral and MaxilloFacial Surgery, Mar Baselios Dental College, Kothamangalam, Ernakulam - 686 691. E-mail: drjoy303@gmail.com

How to cite this article: George A, Vichattu SV, Das RJ, Thomas A, James JE. Multiple osteomas of mandible in a patient with gardners syndrome - report of a case. Oral Maxillofacial Pathol J 2023;14(1): page no. 131-133

Source of Support: Nil

Conflict of Interest: None

of FAP. It is called Gardner's syndrome only when both colonic and non colonic components are present.

Case Presentation

Patient reported to Department of Oral and Maxillofacial Surgery with a complaint of slow growing, painless swelling over right side of face since two years (Figure 1). She had a history of tooth extraction in the right lower jaw 6 years earlier. She had few missing teeth also for which she gave a vague history. There was no history of trauma in the related area. Her medical history was non contributory. An extra oral examination showed multiple swellings over right mandible region anterior to ear.

On palpation, the swelling was not tender, with no local rise in temperature, bony hard in consistency, and attached to the body of mandible. The overlying skin was normal in color and showed no adhesion to the mass. One swelling of size 2 cm x 1cm, roughly oval in shape, 4 cm anterior to posterior border of the mandible, and another swelling of 2 cm x2 cm size 1 cm anterior to posterior border of mandible was found. Another diffused bony hard swelling extending over ramus region 1.5 cm superior to lower border of mandible extending up to right ear tragus region was also found.

Intraoral examination revealed a well-defined subperiosteal mass on the buccal plate of the right mandible, which extending from the area of right first molar region and another swelling over the angle region.

On the basis of history and clinical examination, the provisional diagnosis was given as osteoma of the right body of the mandible, and the differential diagnosis as bony exostosis, osteoblastoma, osteosarcoma, ameloblastoma, odontogenic fibroma, dentigerous cyst, central giant cell granuloma, and fibrous dysplasia. The patient was subjected to routine hematological and radiographic examination.

Orthopantamogram revealed multiple radiopaque masses over right ramus, body and angle region (Figure 2). Although CT was advised, patient opted against it due to financial constraints. Excision biopsy under local anaesthesia was planned after routine blood investigations. Vestibular incision was made on right side mandible body region extending from ramus to canine region. A full thickness mucoperiosteal flap was raised to expose underlying bony deformity over the angle and body region (Figure 3). Bony projection was chiselled and removed in total and bony contouring was done. Wound was sutured. Excised specimen was sent for histopathologic examination (Figure 4).

Multiple osteomas in the mandible raised a suspicion of Gardners syndrome. The patient was advised a medical consultation regarding the possibility of the syndrome. The patient was advised lower gastrointestinal tract endoscopy by the physician, which revealed multiple polyps in the intestine. This was suggestive of Gardner's syndrome.

Histopathology revealed lamellated dense cortical bone exhibiting osteocytic lacunae with osteocytes in few marrow spaces with fibro fatty marrow. Dense compact bone with scanty marrow spaces was pathognomonic of ivory osteoma, which is a variant of osteoma. Histologically, it consisted of dense compact bone with no evidence of active or previous

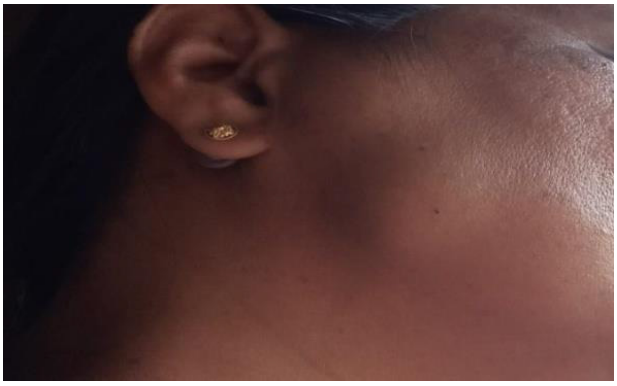

Fig. 1: Extraoral swelling that caused facial asymmetry

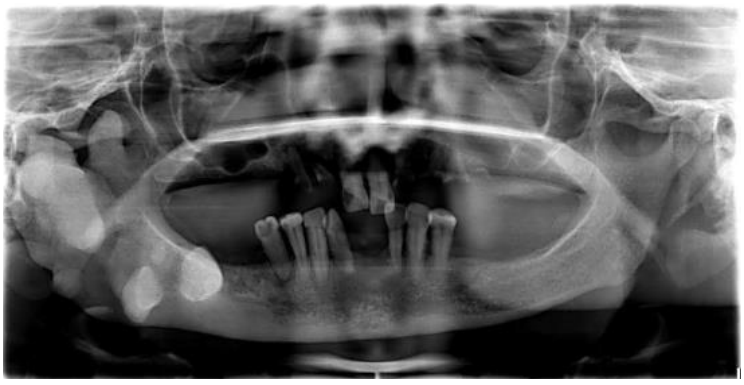

Fig. 2: OPG

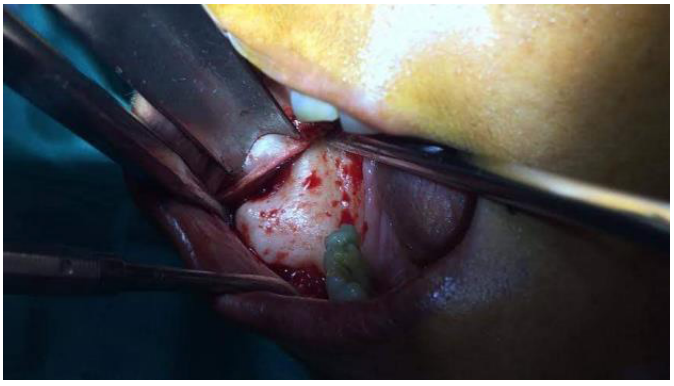

Fig. 3: Intraoral exposure of osteoma

Fig. 4: Gross Pathology

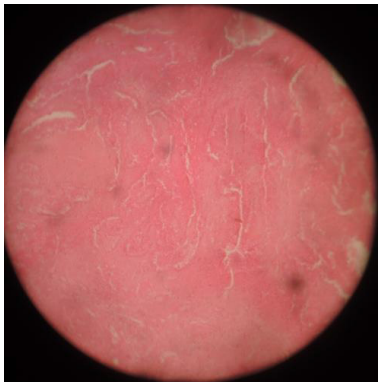

Fig. 5: H&E x5

inflammation. The tissue structure differed significantly from the normal bone structure. No haversian system was visible (Figure 5).

In view of these findings, it was estimated that further surgical approach to the lesion would carry a high risk of tissue damage with minimal benefit. The patient was scheduled for regular follow up, both medical and dental because of a possible chance of malignancy in the intestine and recurrence of the lesion in the mandible was expected.

Discussion

Osteomas usually occur singly, but when they are multiple, Gardner's syndrome must be ruled out. Gardner's syndrome includes familial adenomatoid polyposis (FAP), multiple osteomas, supernumerary, impacted tooth and epidermoid cysts. Osteomas in this case usually affect membranous bones as mandible and maxilla. Osteomas of the skull may be compact, spongy, or mixed. The differential diagnosis will include most of the bony lesion that cause facial asymmetry.[5]

Non syndromic osteomas are typically solitary. However, it is possible to find cases of multiple osteomas, with the risk that the patient has other underlying conditions, such as Gardner's Syndrome.[6] This is why other clinical signs associated with it should be investigated, such as digestive polyps especially in the lower gastrointestinal tract with possible malignant evolution, cutaneous tumours, exostosis of the skull, and supernumerary teeth. The GI tract polyps are usually identified with lower GI tract endoscopy as was done in our case. There could also be rectal bleed in certain cases with rectal polyps. Gardner and Richards described the syndrome along with its genetic pattern in 1953.[7]

Studies showed that 62-80% patients with Gardner's syndrome have osteomatous lesions and the average number of lesions is between 2.9 and 5.9.[8] In our patient we had five lesions in the mandible out of which only three showed significant growth and caused asymmetry of the face.

The jaw lesions would precede the development of colonic polyps and therefore may contribute to early diagnosis of Gardner's syndrome, but they do not seem to have prognostic value regarding the development of colonic cancer. Also jaw lesions grow gradually in childhood and adolescence but continue to develop in adulthood, careful and periodical radiographic examination of the jaws must be performed during postoperative follow-up.

It is known that Gardner's syndrome is inherited in an autosomal dominant fashion. Familial adenomatous polyposis (FAP) is an autosomal dominant form of intestinal polyposis and colorectal cancer caused by germ line mutations in the adenomatous polyposis coli (APC) gene. Thus, the patient's family members should also be screened for polyps. However, no other individual in the patient's family ever had this disorder.[9]

The other components like epidermoid cysts which usually happen in 50-60% of Gardner's syndrome was absent in our case. But the patient had history of some unerupted and supernumerary tooth which was removed years back.

Conclusion

Though multiple osteomas usually do not have a malignant transformation, the patient is under constant follow up since there is a chance of malignant transformation following the intestinal polyps. It is to be kept in mind that multiple osteoma warrants additional investigations as they usually precedes intestinal polyps.

References

1. G. Petur Nielsen, Andrew E. Rosenberg, Bone-Forming Tumors Bone and Soft Tissue Pathology, 2010, Chapter 14 2010 page 309.
2. Bilkay U, Erdem O, Ozek C, Helvaci E, Kilic K, Ertan Y, Gurler T. Benign osteoma with Gardner syndrome: review of the literature and report of a case. J Craniofac Surg. 2004 May;15(3):506-9.
3. Baykul, Timuçin & Heybeli, Nurettin & Oyar, Orhan & Doğru, Harun. (2004). Multiple huge osteomas of the mandible causing disfigurement related with Gardner's syndrome: Case report. Auris, nasus, larynx. 30. 447-51
4. Samir K. El-Mofty, Chapter 9 -Bone Lesions, Diagnostic Surgical Pathology of the Head and Neck (Second Edition), 2009, Pages 729-784.
5. Kim JY, Park GM, Cho BH, Nah KS. Odontogenic myxoma: report of 2 cases. Korean J Oral Maxillofac Radiol 2002; 32: 231-4.
6. Nilesh K, V Vande A, K Veerabhadrappa S. Solitary peripheral ivory osteoma of the mandible presenting with difficulty in deglutition: a case report. J Dent Res Dent Clin Dent Prospects. 2017 Winter;11(1):56-60
7. Gardner EJ, Richards RC. Multiple cutaneous and subcutaneous lesions occuring simultaneously with hereditery polyposis and osteomatosis. Am J Hum Genet 1953;5:139/47.
8. Katou F, Motegi K, Baba S. Mandibular lesions in patients with adenomatosis coli. J Cranio-Max Fac Surg 1989;17:354-8.
9. Gómez García EB, Knoers NV. Gardner's syndrome (familial adenomatous polyposis): a cilia-related disorder. Lancet Oncol. 2009 Jul;10(7):727-35. doi: 10.1016/S1470-2045(09)70167-6