Case Report

Osteochondroma of maxillofacial region: Tumor arising from two different developmental bones

Sujata Mohanty, Himanshu Gupta, Jitender Dabas, Priyadarshan Kumar

Department of Oral and Maxillofacial Surgery, Maulana Azad Institute of Dental Sciences, New Delhi, India

Abstract

Osteochondromas are benign bony tumors which are commonly believed to originate by the proliferation of epiphyseal cartilage into the surrounding tissues. However, this hypothesis cannot explain the occurrence of this tumor in the intramembranous bones and soft tissue. Since most of the craniofacial bones have intramembranous origin, the occurrence of this lesion in this territory is considered rare. Contrary to the above hypothesis, Lichtenstein proposed that this entity arises from the metaplastic changes in the periosteum which explains the occurrence of this tumor in endochondral as well as intramembranous bones and also in soft tissues. Complying with Lichtenstein's hypothesis, the authors are presenting two cases of osteochondromas with one arising from the endochondral bone (the coronoid process of the mandible) and the other from an intramembranous bone (lateral pterygoid plate of the sphenoid).

Key Words: Coronoid, Jacob's disease, osteochondroma, sphenoid bone

Address for correspondence:
Dr. Sujata Mohanty, Department of Oral and Maxillofacial Surgery, Maulana Azad Institute of Dental Sciences, New Delhi - 110 002, India.
E-mail: drsm28@gmail.com

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INTRODUCTION

Osteochondromas are cartilage-capped bony protrusions originating juxta-cortically from the endochondral bones. It is considered as the most common tumor of the axial skeleton found mainly in the metaphysis of long bones. It commonly occurs in adolescence or childhood with 80% of the cases seen in the first two decades,[1] comprising 20–50% of the benign tumors and 10–15% of the bony tumors overall.[2] Most of the cases are solitary, but multiple lesions are found to occur in context with hereditary multiple osteochondromas (HMO) syndrome. This syndrome has inherited in autosomal dominant manner and is caused by mutation in two genes, namely, EXT1 on chromosome 8 and EXT2 on chromosome 11.[3] The solitary lesions undergo malignant transformation in about 1–2% of patients. Whereas, lesions associated with HMO syndrome have 1–25% chances of malignant transformation.[4] Various theories of etiopathogenesis have been proposed for this lesion, out of which Lichtenstein's theory is the most widely accepted which proposes that osteochondroma develops by the metaplastic changes in the periosteum, as the pluripotent periosteum has the potential to develop osteoblasts and chondroblasts.[5] However, what triggers this metaplastic change is still unknown.

CASE REPORTS

Case 1

A 22-year-old female reported with the chief complaint of difficulty in swallowing and a swelling on the left side of the

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palatal region. First, she noticed a small swelling intra-orally on the left posterior palatal region 1 year back which gradually increased in size over a period. Initially, the swelling was asymptomatic but later on as the swelling increased in size, the patient experienced difficulty in deglutition. No history of associated trauma to the region was present. The clinical examination revealed facial symmetry, stable occlusion and no limitation in mouth opening. Intraoral examination revealed a fixed, well-defined bony apophysis present submucously on the left side of soft palate just posterior to the tuberosity [Figure 1a]. The mass was approximately 1.5 cm in diameter and was hard and tender on palpation. Color and texture of the overlying mucosa were normal without any ulceration and discharge. Based on these clinical findings, exostosis, osteoma or osteoblastoma was suspected.

Orthopantomogram (OPG) was done which only revealed an increased radiopacity over the left coronoid region as compared to other side probably due to superimposition. Hence, a computed tomography (CT) scan was advised.

CT revealed a well-defined hyperdense mass measuring 1.0 cm × 1.5 cm arising from the left lateral pterygoid plate with a continuous medullary cavity extending up to left retromolar trigone region [Figure 1b]. The cortex of the lesion was continuous with the lateral pterygoid plate. This continuity of medullary cavity of the lesion and lateral pterygoid plate was highly indicative of osteochondroma. Mandibular region and bilateral temporomandibular joints (TMJs) appeared normal.

As the lateral pterygoid plate is an unusual site for osteochondroma, other entities such as osteoma and osteoblastoma were considered as differential diagnosis. An incisional biopsy was performed and based on clinical and histological examination; a diagnosis of osteochondroma of left pterygoid process of the sphenoid was made. She was operated under general anesthesia with nasotracheal intubation. Intraoral incision was placed over the palpable mass, medial and posterior to the left maxillary tuberosity and the mass was exposed. Resection of the bony mass was done and the surgical site was closed primarily. The excised mass was ovoid, yellowish white and hard in consistency. The whole specimen was then sent for histopathological examination which confirmed the incisional biopsy finding. Histopathological examination revealed a core of tissue covered by a cartilaginous cap. The cartilage cells were regularly aligned representing hyaline cartilage. Areas of cancellous bone formation were seen extending from this in a perpendicular direction. The degree of mineralization of bone varied at areas. The marrow space showed adipose tissue associated with stromal tissue [Figure 2]. Thus, a final diagnosis of osteochondroma of the lateral pterygoid plate was arrived at. The patient was kept on regular follow-up and no recurrence or any complication has been noticed for the past 3 years of surgery.

Case 2

An 18-year-old male patient reported to our institute with a chief complaint of reduced mouth opening and facial asymmetry. On clinical examination, a nontender, bony hard swelling of approximately 2 cm diameter was palpated on the right preauricular region along with expanded zygomatic arch [Figure 3]. On mouth opening, a mechanical "stop" was noticed after an opening of 11 mm. Clinically, a differential

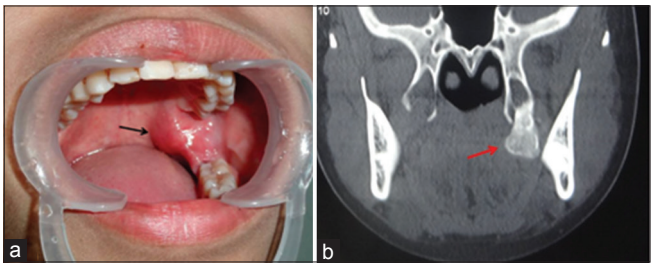

Figure 1: (a) Intra-oral submucosal bony hard swelling on left side of soft palate. (b) Well-defined hyperdense lesion arising from the left pterygoid process of sphenoid. Cortical continuity between bone and lesion can be seen

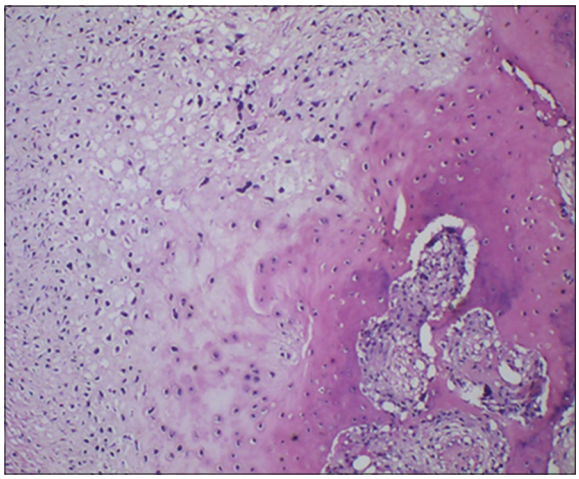

Figure 2: Photomicrograph showing chondroid matrix with proliferating chondroblasts and cartilaginous tissue is seen blending with the cancellous bone (H&E stain, x100)

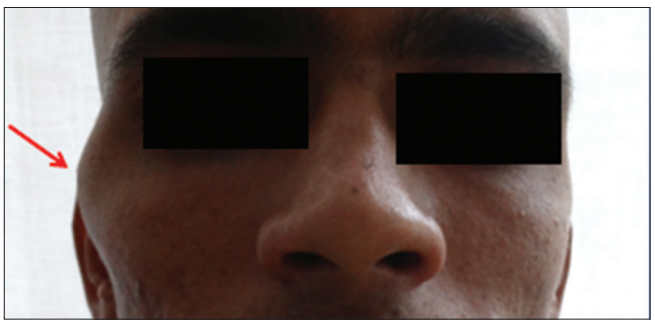

Figure 3: Swelling over right side zygoma causing facial asymmetry

diagnosis of benign tumor/exostosis of the zygoma, TMJ ankylosis, or condylar/coronoid tumor was considered. OPG revealed a radiopaque lesion seemingly attached to the right coronoid process of the mandible. CT examination revealed a well-defined heterogeneous hyperdense mass seemingly arising from the right coronoid process of the mandible. The mass appeared mushroom shaped in coronal section proliferating laterally while the axial section showed a pseudo-joint formation between the coronoid process and the zygomatic arch [Figure 4]. All these features were highly suggestive of osteochondroma of the coronoid process (Jacob's disease).

Extraoral Blair's approach was used to address the tumor. Expansion of the overlying zygomatic process was noted on the way to the coronoid tumor. The coronoid with its associated tumor was excised at a level below the sigmoid notch sparing the condyle. The mass was big enough to be obstructed by the expanded zygomatic arch; hence, hindering its removal. Malarplasty was done by removing a part of the expanded arch which facilitated the removal of the coronoid tumor complex. Excised part of the arch was sculptured and fixed into the arch defect with the help of transosseous wires and wound was closed in layers. After removal of the pathology, an intraoperative mouth opening of 40 mm was achieved. Histopathologic examination revealed a row of proliferating chondroblasts continuing into an area of maturing cartilaginous tissue. Chondrocytes with small nuclei were seen within lacunae. The cartilaginous tissue was seen blending with the cancellous bone. Bone was lined by plump osteoblasts and contained osteocytes within lacunae. Intervening marrow spaces were fibrovascular in nature [Figure 5]. Thus, a final diagnosis of osteochondroma was made. At 3 years follow-up, no recurrence was noticed and the swelling had resolved [Figure 6] with the maintenance of 40 mm mouth opening.

DISCUSSION

Osteochondromas are cartilage-capped bony protrusions from the cortical surface of endochondral bones and are benign tumors arising in long bones of the axial skeleton but rarely seen in the craniofacial bones. Till now, the majority of the reported cases demonstrate that the osteochondromas in the craniofacial region occur on the condylar and coronoid process[1,6] which tend to support the theory of its development from aberrant foci of epiphyseal cartilage. However, this tumor has also been reported to occur in the intramembranous bones as well as soft tissues of the craniofacial region. This seems to challenge the theory of development from epiphyseal cartilage. Some theories that could explain its occurrence in intramembranous bones and soft tissue are as follows:

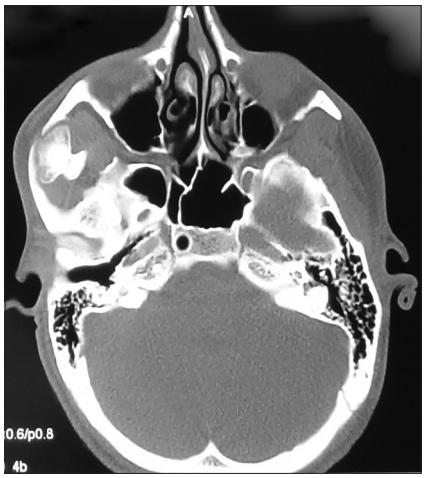

Figure 4: Axial computed tomography image showing the pseudo-joint formation between zygomatic arch and the tumor mass of coronoid process

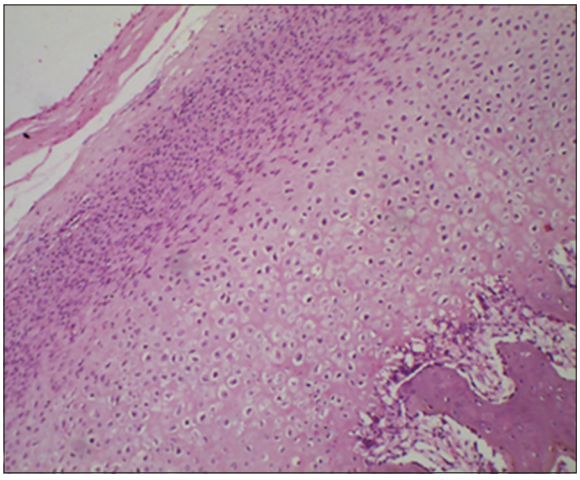

Figure 5: Photomicrograph showing chondrocytes in lacunae and deeper connective tissue showing cancellous bone (H&E stain, x100)

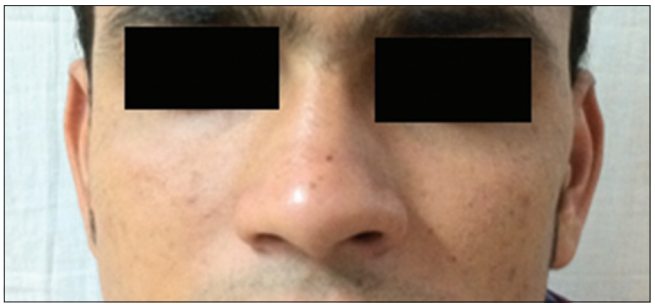

Figure 6: Three years postoperative image showing resolution of bony swelling

- Development from remnants of embryonic cartilage (Meckel's cartilage)
- Osseocartilagenous differentiation of ectopic Mesenchymal cells
- Metaplastic changes in periosteum which have the potential to develop osseocartilagenous tissue.[6,7]

Diagnosis of osteochondromas is made by radiographic and histopathologic examination. Histopathologically, osteochondromas show bony tissue covered by hyaline and fibrous cartilage. On the deeper aspect where cartilage interfaces with the bone, endochondral ossification is seen. Histopathologically, osteochondromas need to be differentiated from osteoma, chondroma, osteoblastoma and chondroblastoma.[8] Recently, the analysis of the gene expression pattern in the extracellular matrix, particularly the expression profile of collagen (COL) using immunohistochemistry and *in situ* hybridization methods, has been forwarded as a modality for the classification and diagnosis of the chondrogenic tumors.[9] The chondrocytes in mesenchymal cell layer overlying the cartilaginous tissue in osteochondroma express COL2 and aggrecan proteoglycan, whereas, the chondrocytes in the deeper zone are hypertrophic and express COL10 along with endochondral ossification.[10,11]

Osteochondroma of the lateral pterygoid plate of the sphenoid bone are extremely rare finding and also represent atypical location for its occurrence. Only one case at this site has been reported in the literature so far.[2] Clinical signs and symptoms usually depend on the size of the lesion and its relation to the adjacent structures. Myriad of symptoms ranging from painless swelling intra-orally, difficulty in deglutition, partial hearing loss, pain in the ear, secretory otitis media and limitation in mouth opening can be present. In the present case, the gradual increase in size with painful deglutition necessitated its removal. On CT imaging, the continuity of cortex and medulla of the lesion with the bone is considered as diagnostic of osteochondroma.[12] The radiographic findings in our case were similar which helped in diagnosing the tumor. The treatment of osteochondroma includes resection of the tumor. The size of the tumor in the present case was small (1.0 cm × 1.5 cm), so intraoral approach was used and the lesion was carefully resected.

Osteochondroma of the coronoid process along with the formation of pseudo-joint with the zygomatic arch (Jacob's disease) is also a rare condition and only 43 cases have been reported so far.[7,13-18] Clinical features include a gradual reduction in mouth opening, thus mimicking TMJ derangement disorders,[18] especially when there is no associated swelling or asymmetry. When suspecting TMJ derangement disorder, magnetic resonance imaging (MRI) of the joint is usually the preferred imaging modality. However since coronoid process usually remains outside the view of TMJ MRI, the lesion remains undetected and the patients are often rendered wrong treatment. Thus, CT is considered as the imaging method of choice in such cases. The valuability of three-dimensional CT in the diagnosis of Jacob's disease has well been described by Akan and Mehreliyeva.[19] In the present case, bony swelling could easily be palpated in the preauricular region and there was no pain during mouth opening. Rather there was a sudden "stop" after an initial mouth opening of 11 mm. This directed the suspicion toward some benign pathology. Following this, noncontrast CT was advised which showed pseudo-joint formation between the zygomatic arch and the lesion with resorption of the arch at few points. This aided in the diagnosis of the tumor. Histopathologically, the lesion can be either osteoma, osteochondroma or exostosis depending upon the bone: cartilage ratio.[20] In the present case, diagnosis of osteochondroma was made after histopathologic examination. Coronoidectomy with either intra- or extra-oral approach is the treatment of choice depending on the size of the tumor. In this case, coronoidectomy was done by the extraoral approach and no recurrence was noted after 3 years of follow-up.

CONCLUSION

With these two cases of osteochondroma in two different developmental bones, i.e., endochondral (coronoid process) and intramembranous (pterygoid plate of sphenoid), the authors agree with the Lichtenstein's theory of the origin of this tumor from the metaplastic changes in the periosteum with differentiation of osteoblasts and chondroblasts from the pluripotent cells.

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Conflicts of interest
There are no conflicts of interest.

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