

MAXILLECTOMY FOR CEMENTIFYING OSSEOUS DYSPLASIA OF THE MAXILLA: A CASE REPORT

Sevtap Akbulut¹, Mehmet Gokhan Demir², Kayhan Basak³, Mustafa Paksoy¹

Kartal Dr. Lutfi Kirdar Education and Research Hospital ENT Department Istanbul, Turkey¹; Prof. Dr. Celal Ertuğ Etimesgut State hospital ENT Department, Etimesgut-Ankara, Turkey²; Kartal Dr. Lutfi Kirdar Education and Research Hospital Pathology Department Istanbul, Turkey³

Summary: Objectives: Osseous dysplasias are the most common subtype of fibro-osseous lesions of the maxillofacial bones. They are benign and often present as incidental asymptomatic lesions. Diagnosis can be made with clinical and radiographic examination. Case report: This article reports the case of a 47-year-old man with a huge cementifying osseous dysplasia of the maxilla that presented with secondary infection after teeth extraction and repair of oroantral fistula. A subtotal maxillectomy had to be done after conservative treatment failed to resolve the infection. Discussion/Conclusion: Biopsy, tooth extraction or surgical trauma to the affected bones of osseous dysplasia could easily lead to severe secondary infection, the treatment of which may be very difficult, and thus should be avoided if possible.

Keywords: *Osseous dysplasia; Cemento-osseous dysplasia; Fibro-osseous lesions; Periapical osseous dysplasia; Focal osseous dysplasia; Florid osseous dysplasia; Maxilla; Maxillectomy*

Introduction

Osseous dysplasias (ODs) of the jaw are idiopathic benign lesions showing an admixed production of fibro-col-lagenous tissue and various mineralized materials, including woven bone, lamellar bone, and/or cementum-like materials (1). They are grouped under the “bone-related lesions” in the current World Health Organization (WHO) Classification of Head and Neck Tumors, together with fibrous dysplasia and ossifying fibroma (2). Osseous dysplasias (also referred to as cemento-osseous dysplasias) are located in the periapical region of the tooth-bearing jaw areas. They are subclassified into periapical osseous dysplasia, focal osseous dysplasia, and florid osseous dysplasia according to the clinical presentation (1, 2). Periapical osseous dysplasia occurs in the anterior mandible involving only a few adjacent teeth. Focal osseous dysplasia is the most common type and has a limited lesion occurring in a posterior jaw quadrant. Florid osseous dysplasia is a more extensive disease occurring bilaterally in the mandible or in both maxilla and mandible. This case report demonstrates an unusual case of OD of the maxilla which presented with persistent secondary infection after teeth extractions and was successfully managed with subtotal maxillectomy.

Case Report

A 47-year-old man was referred to our clinic with complaints of dull facial pain and odorous discharge from the

roots of his right upper teeth for the last couple years. The patient’s past medical history included extraction of right upper second and third molar teeth two years earlier, and repair of oroantral fistula with local flaps two times afterwards. Physical examination revealed mild facial asymmetry due to a painless expansive bony swelling in the region of his right maxilla which filled the infraorbital region and obliterated the gingivolabial sulcus. He said he had “fullness of his face” for more than a decade. The swelling extended up to the midline of the hard palate. It was mostly firm and asymptomatic on palpation, although the part of the swelling on the palate region was softer and painful. The overlying mucosa looked normal. Purulent discharge was observed from a fistula tract between the right upper second premolar and the first molar teeth. All laboratory tests, also including serum alkaline phosphatase, calcium, and phosphorus were within the normal limits. A panoramic radiograph displayed a lobular, irregularly shaped radioopaque mass involving the right maxilla extending from maxillary alveolus to inferior orbital border and right maxillary tuberosity. CT scan revealed a right maxillary well-defined, lobulated, inhomogenously high-density, expansile mass with scattered calcifications which also obliterated the maxillary sinus. The inferior maxillary bony cortex was eroded and the orbital floor was thinned and upwardly displaced (Figure 1). Borders of the lesion were indistinct and blended into the unaffected surrounding bone in the palatal region. On the basis of clinical and radiographic examinations, a diagnosis of “infected osseous dysplasia” was made. Initial treatment was performed

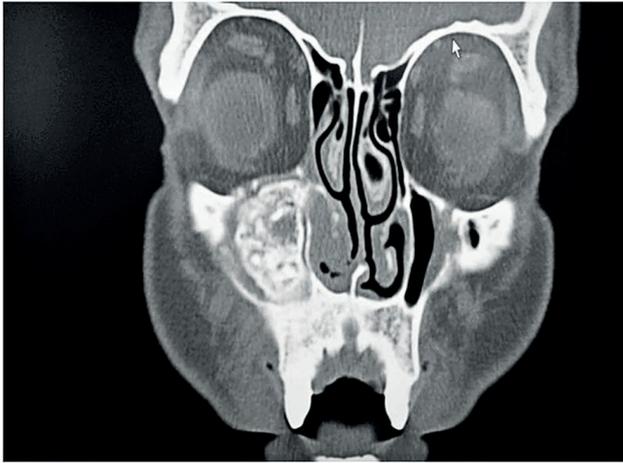


Fig. 1: Paranasal CT image reveals well-defined, lobulated, high-density, expansile mass in maxilla, obliterating the sinus.

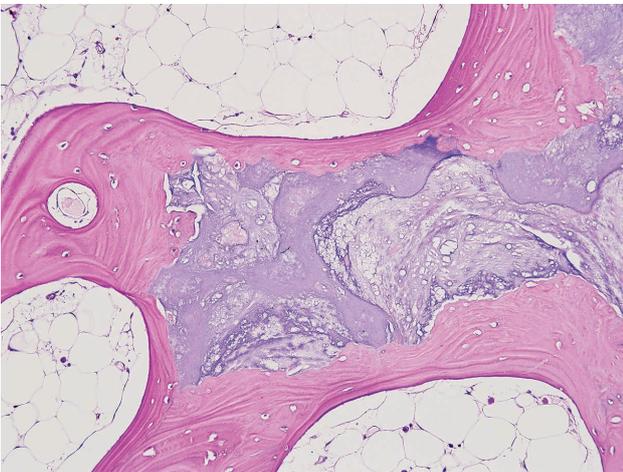


Fig. 2: Histopathological examination shows irregularly shaped cementum-like mineralizations with normal lamellar bone and fibrous tissue in maxilla (original magnification Cementum-like basophilic mineralization within normal bone lamella (original magnification $\times 200$, H&E).

using high dose antibiotherapy and surgical debridement. Histopathological examination of surgical specimen revealed extensive, irregularly shaped cementum-like mineralizations continuous with normal lamellar bone in fibrous tissue. Based on histopathological features a diagnosis of “osseous dysplasia” was confirmed (3, 4). The infection of the maxilla failed to respond to conservative treatment, even after culture-directed antibiotherapy and another session of surgical debridement. A right subtotal maxillectomy was conducted on the patient, and split-thickness skin graft was used for cavity relining. The patient tolerated the procedure well, and the postoperative course was uneventful. The final surgical pathology revealed cementifying osseous dysplasia in maxilla with ulceration and superimposed osteomyelitis, and also accompanying squamous metaplasia of the sinus

mucosa (Figure 2). A maxillary obturator prosthesis was prepared for reconstruction of the maxillary defect. The patient was satisfied with the function of the prosthesis. He has been followed-up for 30 months without any problem.

Discussion

Osseous dysplasias represent the most common subtype of fibro-osseous lesions of the maxillofacial bones (1, 3). They are divided into three subtypes based on clinical and radiographic features, although they appear to represent only variants of the same basic disease process (1, 5). All facial bones can be affected in OD but most of the lesions are seen on the posterior mandible, especially connected to toothbearing areas. The male-to-female ratio for OD is 1 : 4.2 and it shows strong predilection for persons of African descent, although it also occurs in Caucasians and Asians, as in our case (1, 6). Most of the lesions are asymptomatic and detected incidentally. Radiographically, OD evolves through three phases. It presents with a well-defined radiolucency during the early stage. Then it demonstrates a mixed radiolucent-opaque pattern with a well-defined radiolucent rim around the radiopacity. After maturation, OD appears radiologically dense, with lobulated mineralized deposits often with ill-defined borders (6, 7).

Differential diagnoses include fibrous dysplasia, ossifying fibroma, Paget’s disease, and focal or diffuse sclerosing osteomyelitis. Fibrous dysplasia is not radiographically well-defined, and it blends into the surrounding bone. The most typical radiographic feature is that of a ground-glass opacification, although early lesions may be largely radiolucent (5). Ossifying fibroma mostly presents as a well-defined radiolucent-radiopaque image (5). Paget’s disease shows radioopaque lesions without a radiolucent capsule; it is usually polyostotic; and it causes an elevated serum alkaline phosphatase level. Focal sclerosing osteomyelitis is usually encountered in the posterior mandible, and it is extremely rare in the maxilla. It is characterized by an asymptomatic, nonexpansile periapical lesion associated with a tooth. Diffuse sclerosing osteomyelitis presents with cortical osseous expansion and a dull episodic pain that may last for weeks, subside and later become symptomatic again. It is characterized by a unilateral diffuse ground glass opacification without defined boundaries (6). OD can often be diagnosed clinically and radiographically by experienced clinicians. Biopsy should be avoided because of infection, sequestrum formation, and osteomyelitis risks (8). The lesions are avascular and susceptible to bacterial overgrowth. It is difficult to control infections associated with OD due to insufficient antibiotic concentration in lesions because of poor vascularization. Such infections are induced by direct communication through the mucosa or simply by trauma to the overlying tissues (9). Extraction of the tooth is also not recommended due to poor socket healing in the affected area of the bone (8). In our case, the patient remained asymptomatic for a very long period; extraction of teeth and

surgical trauma in connection with fistula repairs, unfortunately, led to infection. In asymptomatic cases treatment is not necessary, and routine follow-ups and protection from infection by improving oral hygiene are recommended. Treatment is required when infection of the lesion occurs and the patient becomes symptomatic. Surgical debridement and enucleation are the first-line treatment alternatives (10). The lack of vascularity of the lesion also complicates the treatment by leading to necrosis and infection which is resistant to conservative treatment. More aggressive and extensive treatment is proposed when conservative treatment fails to resolve the infection (4). However these methods of treatment are not discussed satisfactorily in the literature. Only in a case report were partial mandibulectomy and reconstruction with a vascularized fibular graft described for an aggressive secondary infection of florid OD. Our patient can be classified as “focal” OD as there was only a single lesion of the maxilla. But the location and size of the lesion were unusual. Maxillary bone is affected only in 20% of focal ODs, and it rarely grows more than 2 cm (1). In this case, the lesion obliterated the whole right maxillary quadrant of the jaw, including the maxillary sinus and palate. In order to treat the secondary infection, which did not respond to antibiotherapy and conservative debridement, an extensive surgical resection, i.e., a subtotal maxillectomy, had to be performed. Subtotal maxillectomy is defined as resection of the palate, medial or/and anterior walls of maxilla, preserving the infraorbital plate and posterior wall. It is used for resection of benign lesions of the maxilla or nasal cavity. As far as we are aware, this is the first report of OD which necessitated a maxillectomy for treatment. Our case highlights the very important point that secondary infection of OD can become very difficult to treat. Clinicians should be highly vigilant to prevent patients from getting infected. Diagnosis can be made with accurate clinical and radiographic assessment. As a result, biopsy should be avoided. Tooth extraction or other elective surgical procedures in the affected area of the bone are not recommended, as well. Oral hygiene and regular follow-up should be reinforced in patients.

Conclusion

Osseous dysplasias are the most common subtype of fibro-osseous lesions of the maxillofacial bones. They can be diagnosed with clinical and radiographic examination. Although they are benign and often asymptomatic, treatment of secondary infections may be very difficult. The authors reported a rare case of osseous dysplasia of the maxilla that presented with secondary infection and treated with an extensive surgical resection. Clinicians should ensure the follow-up of the diagnosed patients and take precautions for preventing them from getting infected.

Acknowledgements

The authors wish to thank Daniel B. Magraw from John Hopkins University for editorial assistance in the preparation of this manuscript.

References

1. Phattaratatip E, Pholjaroen C, Tiranon P. A clinicopathologic analysis of 207 cases of benign fibroosseous lesions of the jaws. *International journal of pathology* 2014; 22(4): 326–333.
2. Barnes L, Everson JW, Reichart P, Sidransky D et al. WHO Classification Pathology and Genetics of Head and Neck Tumours. Lyon, France: IARC Press 2005.
3. McCarthy EF. Fibro-osseous lesions of the maxillofacial bones. *Head Neck Pathol* 2013; 7 :510.
4. Kim JH, Song BC, Kim SH, Park YS. Clinical, radiographic and histological findings of florid cemento-osseous dysplasia: a case report. *Imag Sci Dent* 2011; 41: 139–42.
5. Waldron CA. Fibro-osseous lesions of the jaws. *J Oral Maxillofac Surg* 1993; 51: 828–835.
6. Eversole R, Su L, ElMofty S. Benign fibro-osseous lesions of the craniofacial complex: a review. *Head Neck Pathol* 2008; 2: 177–202.
7. Noffke CEE, Raubenheimer EJ. Expansive osseous dysplasia: report of 9 lesions in an African population sample and review of the literature. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2011; 111: e35–41.
8. Sarmiento DJS, Monteiro BVB, Medeiros AMC, Silveira EJD. Severe florid cemento-ossous dysplasia: a case report treated conservatively and literature review. *Oral Maxillofac Surg* 2013; 17: 43–6.
9. Singer SR, Mupparapu M, Rinaggio J. Florid cemento-osseous dysplasia and chronic diffuse osteomyelitis: report of simultaneous presentation and review of the literature. *J Am Dent Assoc* 2005; 136: 927–31.
10. Neville BW, Damm DD, Allen CM, Bouquot JE. Bone pathology. In: Neville BW, Damm DD, Allen CM, Bouquot JE eds. *Oral and Maxillofacial Pathology*. St Louis, Missouri: Saunders Elsevier; 2009. pp. 641–5.

Received: 25/02/2015

Accepted in revised form: 01/04/2015

Corresponding author:

Mehmet Gökhan Demir, Umit mah. Meksika cad. Defne 9 sit. No: 3/21 Umitkoy, Çankaya – Ankara/Turkey; e-mail: mgokhandemir@yahoo.com
