

Frontal sinus cementifying ossifying fibroma

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ABSTRACT

Cementifying ossifying fibroma is a fibro-osseous lesion which produces cementum. Cementifying ossifying fibroma is a mesodermal, non-odontogenic tumor of ectopic multipotential periodontal membrane blast cells. Although, it is a benign bone tumor, it has an aggressive destructive behavior in patients under 20 years of age with high tendency for recurrence. Cementifying ossifying fibroma can be recognized microscopically by the presence of cementicles. This is a case of frontal sinus cementifying ossifying fibroma presenting with proptosis in a 39-year-old male.

Saudi Med J 2005; Vol. 26 (3): 470-472

Fibro-osseous lesions demonstrate replacement of normal bone architecture by a benign fibrous tissue containing varying amounts of mineralized material.¹ These lesions include fibrous dysplasia at one end (monostotic and polyostotic) and at the other end, a group of lesions arising from multipotential mesenchymal blast cells situated in the periodontal membrane that have the capacity to produce cementum, bone and fibrous tissue. These lesions are ossifying fibroma, cementifying fibroma, the cementomas, fibrous osteoma, osteoblastoma and osteoma. In 1971, the World Health Organization (WHO) classified 4 types of fibro-osseous lesions: fibrous dysplasia, ossifying fibroma, cementing fibroma and cementifying ossifying fibroma (COF).² But, this suggestion of origin is not suitable to explain its occurrence in non-tooth-bearing locations. Therefore, others have hypothesized that it represents a disturbance in the normal reparative pattern of bone,³ or trauma may act as a trigger to sudden growth of the ectopic periodontal tissue.⁴ Ossifying fibroma is one of the fibro-osseous lesions. It was first reported in a 35-year-old woman with a large tumor of the mandible that had been present for 25 years.⁵ Fifty-five years later, it was described as a specific clinical entity.⁵ It was described in 3 cases that

may have been fibrous dysplasia.⁵ It has many synonyms such as osteofibroma, fibro-osteoma, monostotic fibrous dysplasia and localized Paget's disease. Ossifying fibroma is an expansile lesion with well-defined margins. Calcification, cementum and bone are dispersed through the lesion. These lesions has predilection for the mandible and maxilla.^{5,6} Its existence in the nasal bones, orbit, ethmoid sinus, sphenoid sinus, occiput and the temporal bone is rare.^{3,5} Involvement of the frontal sinus is very rare. Cementifying ossifying fibroma produces cementum. Cementum is a hard mineralized dental tissue, which has all the characteristics of compact bone. There are 4 types of cementum-producing tumors. They are periapical cement dysplasia, cementifying fibroma, benign cement-blastoma and gigantiform cementoma. Cementifying ossifying fibroma can be recognized microscopically by the presence of homogeneously distributed spherical calcifications (cementicles). These cementicles (Psammoma-like bodies) contain cementum-like, avascular and basophilic bone. The peripheral portion consists of a bony shell with osteoblastic activity on the internal surface touching the tumor and osteoclastic activity on the convex external surface.¹ They are present in the cellular tissue in 60% of the cases.¹ Thus, they are not

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Received 27th September 2004. Accepted for publication in final form 12th December 2004.

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Figure 1 - A 39-year-old male with left proptosis due to cementifying ossifying fibroma.

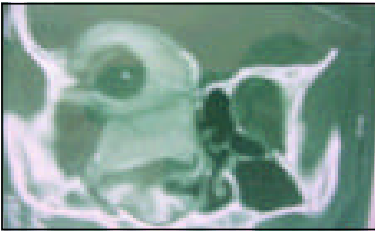


Figure 2 - Coronal computerized tomography shows left frontal, ethmoidal and nasal cementifying ossifying fibroma with orbital and intracranial expansion.

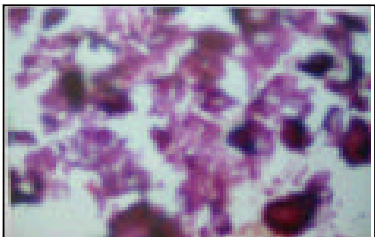


Figure 3 - Cementifying ossifying fibroma cementicles and fibrous cellular stroma (hematoxylin and eosin original x10).

pathognomonic and they are also found in other tumors such as meningioma and thyroid tumors.⁶ In a review of 70 cases of ossifying fibroma and COF involving the paranasal sinuses reported by Bertrand et al¹ in 1993, the average age of patients was 18.6 years and the male to female ratio was 1:1. It encountered in the ethmoid sinus in 40 cases, in the frontal sinus in 14 cases, with orbital extension in 26 cases and anterior cranial fossa extension in 8 cases. Recurrence occurred in 22 cases out of 63 cases (3 of 6 in the COF group). The behavior of these tumors is aggressive, highly destructive and shows a marked tendency to recur; thus, radical surgery is recommended.^{1,6} Radiotherapy is contra-indicated for fear of malignant transformation. It has an incidence of malignancy from 0.4-44%.¹

Case Report. A 39-year-old male was presented with an insidious onset of left proptosis (**Figure 1**). He had no history of epistaxis nor headache. On examination, he was in good general condition. Vision was normal as was the clinical neurologic examination. Nasal examination revealed that the bridge of the nose was broadened resulting in hypertelorism. Left nasal cavity showed medialization of the lateral nasal wall with a mass which is firm to hard with red intact mucosal covering reaching nearly to the nasal floor causing complete obstruction. It did not bleed on touch, from which a biopsy was taken. The right nasal cavity was normal with a good airway. Posterior rhinoscopy showed a smooth red mass in the left choana. A computerized tomography sections (**Figure 2**) demonstrated a large, well-circumscribed mass, involving the left frontal sinus mainly and extending to the ethmoidal sinuses, the nasal cavity reaching its floor and expanding into the anterior cranial fossa. The lesion was heterogeneous due to foci of ossification. The adjacent bones were merely displaced by the tumor which was well-defined by a thin eggshell osseous capsule. The histopathological report revealed evidence of ill-defined involvement by relatively cellular proliferation of bland appearing spindle cells of apparent fibroblastic nature with prominent associated ossification. Although somewhat active appearing, the proliferating cells were generally free of remarkable anaplasia or significant elevation of mitotic activity. There are spheroidal cementum-like calcifications (**Figure 3**), which homogeneously distributed in the fibrous cellular stroma. Trabeculae of woven bone rimmed by osteoblasts. The diagnosis of a COF was made on the basis of clinical, medical-imaging findings and the histopathological report.

The patient was admitted to our department, and an external approach was performed and reduction of mass was performed.

Discussion. Histopathologically, it is difficult to separate ossifying fibroma from fibrous dysplasia as they share in many pathological features. Radiological findings are helpful in differentiation.⁷ Ossifying fibromas are radiologically expansile lesions with sharp demarcation from the adjacent bone. It contains areas of calcifications and ossifications. On the other hand, fibrous dysplasia shows diffuse changes and margins. On histopathological differentiation, the ossifying fibroma is well demarcated from the surrounding non-neoplastic tissue. It has a variability in bone to fibrous tissue ratio, in mixture of woven and lamellar bone and in degree of mineralization.⁶ The bone trabeculae is commonly rimmed by osteoblasts in ossifying fibroma. On the other hand, fibrous dysplasia merges with the adjacent non-lesional bone. It has a classic ground-glass appearance on radiographs. It expands the bone throughout its length rather than in a localized fashion.⁸ It has a uniform bone-to-fibrous tissue ratio. The bone trabeculae is lacking the osteoblastic rimming. Ossifying fibromas occur in all age groups with an average age of 31 years, more common in females and in 75% of cases occurs in the mandible.⁷ Treatment of ossifying fibroma is curettage and osteotomy and if recurrence is detected, conservative excision is recommended. In aggressive lesions, radical surgical resection or bloc resection is advised for definitive therapy. However, recurrence occurs in 0%⁷ to 28%⁹ of cases. The most complete excision results in deformity and functional losses that may be more damaging to the patient than the disease itself and is accompanied by significant morbidity such as serious infections, leakage of cerebrospinal fluid, orbital damage and intracranial injury.¹⁰

Cementifying ossifying fibroma can be diagnosed histopathologically by the presence of spherical

calcifications in a fibrous stroma. However, it is not pathognomonic, as it is present in 60% of cases. Then, it is too difficult to differentiate it from ossifying fibroma and fibrous dysplasia. At this stage, the diagnosis of a COF should be made on the basis of clinical, medical-imaging findings and the histopathological report.⁶

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