

## Case Report

# Benign cemento-ossifying fibroma: a rare case report

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### ABSTRACT

Cemento-ossifying fibroma (COF) is a benign fibro-osseous lesion commonly seen in the head and neck regions. It is considered as a benign, locally aggressive neoplasm that requires surgical excision. COF has traditionally been considered to be slow growing. We report a case of 11 year-old girl who presented to the ENT Department of our hospital with 7 months history of nasal obstruction, proptosis and headache. Computed Tomography scan images showed a mass in the right nasal cavity. This case is notable because involvement of the sphenoid sinus is rare.

**Keywords:** Cemento-ossifying fibroma, Sphenoid sinus, Computed tomography

### INTRODUCTION

Ossifying fibroma (OF) is a rare benign fibro-osseous lesion which was first described by Menzel in 1872. He considered it as a form of Osteoma but the term "ossifying fibroma" was subsequently coined by Montgomery in 1927.

The etiology of OF is unknown but odontogenic, developmental and traumatic origins have been suggested.<sup>1</sup> The most common sites are mandible and maxilla.<sup>2</sup> Other sites reported are parietal, occipital, temporal and sphenoid bone, nasopharynx, sella turcica and nasal cavity. Fibro-osseous lesions of the face and paranasal sinuses are relatively uncommon. Women are affected more often than men with a female to male ratio of 2:1.<sup>3</sup> OF lesions have woven or metaplastic bone as their primary component. However, within certain OF lesions, hard tissue identifiable as woven or metaplastic bone is seen mixed with cementum-like tissue. Tumors in which the main mineralized component is cementum are known as cementifying fibromas. Tumors in which the main mineralized component is bone are known as OFs.

Tumors with a mix of mineralized components are labelled cemento-ossifying fibromas (COF).<sup>4</sup>

COF has been considered to be a slow growing tumor but lesions occurring in the paranasal sinuses and midface display more aggressive behaviour. Histologically, it is composed of calcified material and fibroblastic stroma. The calcified structures are composed of bony trabeculae with prominent osteoblastic rims and some osteoclasts. Cementum-like bodies are scattered throughout the lesion. The connective tissue consists of spindle fibroblastic cells in a storiform pattern.

Radiologically, COF lesions are typically well-defined unilocular radiolucencies with scattered radiopaque foci.<sup>5</sup> The lesions are also expansile and circumscribed by a thick shell of bone density with a multiloculated internal appearance and a content of varying densities.<sup>6</sup> The cementum-like bodies appear radiolucent or like ground glass. On MRI, a COF is hypointense to muscle on T1- and T2-weighted images. The well-demarcated bony walls are isointense to soft tissue on T1-weighted images and hypointense on T2-weighted images.<sup>7</sup>

## CASE REPORT

An 11 year old female presented to ENT Department, with complaint of nasal obstruction, proptosis and headache. No history of epistaxis and diminution of vision were reported.

On anterior rhinoscopy, pinkish mass in the right nasal cavity and deviated nasal septum to the left were noted. On endoscopic examination the right nasal cavity was completely occupied by a firm to hard mass and the origin of mass could not be assessed.

CT imaging using thin contiguous axial post contrast scan of orbits was performed. It revealed evidence of marked thickening involving the sphenoid sinus with cortical thickening, sclerosis and bony remodelling. The lesion was seen extending postero-superiorly to sellar region and antero-inferiorly to nasal cavity. There was associated lateral displacement of the medial wall of orbit without erosion of the wall. It suggested possibility of cemento-ossifying fibroma.

An operative procedure was performed with a trans-nasal endoscopic approach. The origin of the mass was found to be from sphenoid sinus. Haemorrhagic fluid was aspirated from the mass. Mass was resected from the nasal cavity and the walls of the sphenoid sinus were also cleared off disease. Lamina papyracea was found to be intact. Haemostasis was achieved and mass was sent for histopathological examination (HPE). The HPE report confirmed the pathology as cemento-ossifying fibroma.

Patient reported relief from symptoms on follow up. Endoscopic evaluation of the right nasal cavity showed complete wound healing 1 month after surgery.

## DISCUSSION

Ossifying fibroma is one of the benign fibro-osseous lesions.<sup>8</sup> Currently, the term benign fibro-osseous lesion is used in the literature to describe a spectrum of lesions ranging from fibrous dysplasia to ossifying fibroma, including cementifying or cemento-ossifying dysplasia, ossifying fibroma and juvenile active ossifying fibroma.<sup>9,10</sup>

The etiology of OF is unknown but these lesions are presumed to originate from periodontal ligaments of teeth because of their capacity to produce cementum and osteoid material.<sup>11</sup> Other theories include traumatic and developmental causes. Brademann et al explained that ectopic periodontal membrane may have differentiated from primitive mesenchymal cells in the petrous bone as a potential cause for development of OF in this area and speculated that trauma may be an inducing factor for OF.<sup>12</sup> Many authorities prefer to designate the cementum-like materials present in ossifying fibroma as a variation of bone. So, the designations ossifying fibroma, cement-

ossifying fibroma and cementifying fibroma are the same lesions and classified best as osteogenic neoplasm.<sup>10</sup>

The clinical presentation of these tumors is variable, depending on the site and rate of growth.<sup>8</sup> It ranges from an asymptomatic bone lesion found incidentally on imaging taken for another reason, to symptoms due to mass effect of sinonasal lesions such as nasal obstruction, anosmia, hyposmia, headache or epistaxis. Ocular symptoms include visual loss, diplopia, proptosis and epiphora. Larger tumors may also lead to a painless swelling of the involved bone.

Differential diagnoses include fibrous dysplasia, sinonasal psammomatous meningioma, and well-differentiated osteosarcoma.<sup>13</sup> Fibrous dysplasia (FD) is an idiopathic non-neoplastic disease affecting patients during the first 2 decades of life. Craniofacial involvement is seen in 50% of patients with polyostotic lesion and 25% with monostotic lesions.

Most lesions are treated with excision or curettage with or without bone grafting.<sup>2</sup> Total removal must be performed to avoid recurrence.<sup>1</sup>

In conclusion, knowledge of clinical, imaging and histopathological characteristics of OF in this rare location is helpful in differential diagnosis of tumors involving this region of head and neck.

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