Cementoossifying fibroma of the paranasal sinuses: A review of two cases

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Abstract

Cementoossifying fibroma (COF) of the paranasal sinus is a rare benign fibroosseous tumour arising from the periodontal membrane. It contains multipotential cells capable of forming cementum, lamellar bone and fibrous tissue. Cementoossifying fibroma peaks in the third and fourth decades and occurs more frequently in women than in men. Occasionally COF may grow aggressively and extend to involve the orbits and skull base, resulting in serious cosmetic and functional problems. We present CT and magnetic resonance imaging (MRI) findings of two young adults with cementoossifying fibroma of the paranasal sinuses who presented with progressive proptosis and facial deformity. Previously unreported correlations with Cerebral Angiography and MR spectroscopy are discussed.

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1. Introduction

Cementoossifying fibroma (COF) of the paranasal sinus is an uncommon benign fibroosseous tumour arising from the periodontal membrane. It contains multipotential cells capable of forming cementum, lamellar bone and fibrous tissue [1,2]. Occasionally cementoossifying fibroma may grow to a massive size and extends to involve the orbit and skull base causing serious cosmetic and functional problems [3]. The aggressive nature of this tumour may resemble malignant lesions such as fibrosarcoma and osteosarcoma which can give rise to a diagnostic challenge.

We present CT and magnetic resonance imaging (MRI) findings in two young adults with cementoossifying fibroma of the paranasal sinuses who presented with progressive proptosis and facial deformity. Previously unreported correlations with cerebral angiography and MR Spectroscopy are discussed.

2. Case report 1

A 19-year-old man presented to his family physician with gradual right eye proptosis of 1-year duration and progressive right nasal obstruction that had been continuing for 4 months. There was no history of epistaxis or any visual disturbances. Clinical examination revealed a large papillomatous smooth mass arising from the middle meatus causing deviation of the posterior portion of the nasal septum. There was proptosis of the right eye with fullness on the right side of the face. Extraocular muscle movements were normal.

An elective contrast enhanced axial CT scan of the paranasal sinus was performed (GE Lightspeed 16, GE Medical System, Milwaukee, Wisconsin, USA), with 80 ml of Iopromide 300 mg/ml (Ultravist 300, Schering AG, Berlin, Germany). The scan revealed a heterogeneous vascular calcified soft tissue mass, measuring 5.2 cm × 6.0 cm × 5.3 cm occupying the right maxillary sinus and nasal cavity causing expansion of the right medial orbital wall and erosion of the lamina papyracea. Coronal reconstructed CT showed the tumour mass extending superoanteriorly into the right ethmoidal sinus with associated erosion of the cranial base (Fig. 2.1). Proptosis of the right globe was present. A ten-
Fig. 2.1. Case 1: reconstructed CT image in coronal plane showed a heterogeneous vascular calcified soft tissue mass occupying the right maxillary sinus and nasal cavity, extending superoanteriorly into the right ethmoidal sinus with associated erosion of the cranial base.

A tentative diagnosis of extracranial sinonasal meningioma with a differential diagnosis of sarcoma was made at the time. Juvenile angiofibroma was considered less likely as this tumour usually arises from the nasopharyngeal space, characteristically causing widening of the pterygopalatine fossa with anterior bowing of the posterior antral wall. The long-standing history and insidious behaviour of this tumour however did not fit the diagnoses of either osteosarcoma or chondrosarcoma.

Magnetic resonance (MR) imaging examination using the Siemens Magnetom Vision (1.5 Tesla) demonstrated the mass to be isointense to muscle on T1-weighted images and low signal intensity on T2-weighted images with vivid enhancement seen on post-gadolinium images (Fig. 2.2). MR Spectroscopy of this mass showed a high alanine peak, which was suggestive of a meningioma (Fig. 2.3). MR angiogram using the time of flight sequence (2-D TOF) demonstrated dilated feeding vessels from the right external carotid and the right ophthalmic arteries.

Cerebral angiogram showed marked tumour blush which was supplied by the right ophthalmic artery, right maxillary and right facial artery (Fig. 2.4). These vessels were selectively cannulated and embolized with (polyvinyl alcohol 150–200 μm) particles. Satisfactory devascularization of tumour was achieved prior to surgical resection.

3. Case report 2

A 24-year-old lady presented to the ENT clinic with a history of progressive worsening nasal blockage for 6 months associated with blurring of vision. Clinical examination revealed a smooth solid mass arising from the lateral wall of the nasal cavity.
Fig. 2.4. Case 1: cerebral angiogram pre- and post-superselective embolization showing marked tumour blush which was supplied by the right ophthalmic artery, right maxillary and right facial artery. Marked devascularization was seen following tumour embolization.

An elective contrast enhanced axial CT scan of the paranasal sinus was performed (GE Lightspeed 16, GE Medical System, Milwaukee, WI, USA), with 80 ml of iopromide 300 mg/l (Ultravist 300, Schering AG, Berlin, Germany). The scan demonstrated a huge lobulated heterogeneous vascular calcified mass, occupying the ethmoidal sinuses and extending into both the frontal sinuses and the sphenoidal sinuses. Coronal reconstructed CT showed the tumour mass extending into the nasal cavity associated with erosion of the cranial base and planum sphenoidale. There was expansion of both the medial orbital walls and erosion of the lamina papyracea. The left medial rectus muscle was stretched and displaced (Fig. 3.1). The optic chiasma was also displaced upward. Proptosis of the left globe was present.

Magnetic resonance imaging examination demonstrated a mass in the ethmoidal sinuses which was isointense to muscle on T1-weighted images and low signal intensity on T2-weighted images with enhancement seen on post-gadolinium images (Fig. 3.2). There were cystic areas noted within. Cerebral angiogram showed tumour blush which was supplied by both the maxillary arteries and branches of the ophthalmic artery. Subsequently, the branches of the right external carotid artery were cannulated and embolized with polyvinyl alcohol particles (150–200 μm).

Fig. 3.1. Case 2: contrast enhanced axial CT scan of the paranasal sinus showed a huge lobulated heterogeneous vascular calcified mass (X) with a Hounsfield’s unit of 347, occupying the whole ethmoidal sinuses and extending into both the frontal sinuses, sphenoidal sinuses with intracranial extension. There was expansion of both the medial orbital walls and erosion of both the lamina papyracea. The left medial rectus muscle was stretched and displaced.

Fig. 3.2. Case 2: magnetic resonance T2-weighted image demonstrated a low signal intensity mass in the ethmoidal sinuses with cystic areas within. The mass was isointense to muscle on T1-weighted images with marked enhancement seen on the post-gadolinium images.
Surgical resection of these tumours was performed by the otorhinolaryngological and neurosurgical teams through combined approaches of cranioorbitomy and lateral rhinotomy under endoscopic guidance. Intraoperatively the infiltration of the medial wall of orbit, planum sphenoidale and the dura matter by these masses were apparent.

Histopathological examination of the specimens showed pieces of tissue consisting of proliferation of spindle shaped cells arranged in whorls surrounded by bony spheres. These bony structures have thin rims of osteoblasts. The spindle cells were uniform and had oval vesicular nuclei. There were also bony fragments and pieces of collagen tissue covered by respiratory epithelium and containing benign mucoous glands. These appearances were consistent with cementoossifying fibroma.

Following surgery the first patient developed bacterial sepsis but was successfully treated with antibiotics. Both the patients were discharged well and are currently under follow up with the ENT clinic.

4. Discussion

Central cementoossifying fibromas are a distinct form of benign fibroosseous lesions of the mandible and maxilla. They are thought to arise from the periodontal ligament and are composed of varying amounts of cementum, bone and fibrous tissue [4]. Cementum is the mineralized connective tissue that covers the root of the tooth. The hybrid name central cementoossifying fibroma is used because there is a spectrum of fibroosseous lesions that arise from the periodontal ligament, ranging from those with only deposition of cementum to those with only deposition of bone [5]. They arise in the mandible in 62–89% of patients, 77% occurring in the premolar region. Most are diagnosed between 20 and 40 years of age [6].

Central cementoossifying fibromas are asymptomatic until they cause expansion. Thus they are generally not diagnosed until the tumor has had time to produce calcifications. Although cementoossifying fibromas of the mandible are common, cementoossifying fibromas of the maxillary sinus are unusual tumors; only 25 have been reported in the literature [4]. Cementoossifying fibromas are typically well-defined, solitary radiolucencies with scattered radiopaque foci. They vary in radiopacity depending on the amount of cementum and bone that have been deposited. Large tumors may involve the nasal septum, orbital floor and infraorbital foramen. Maxillary central cementoossifying fibromas are large at the time of presentation, indicating the capacity of the tumor to expand freely within the maxillary sinus.

Computed tomography typically reveals a well-marginated mixed density tumour with diffuse scattered calcification which may demonstrate varying pattern of aggressiveness. The cementoossifying fibroma imaged with MR in these two cases were isointense to muscle on T1-weighted images and had a diffuse homogenous low signal intensity on T2-weighted images which likely represents the low free water content of the calcific and fibrous tumour.

The differential diagnoses include other lesions that contain radiopacities in the maxillary sinus such as fibrous dysplasia, osteosarcoma, chondrosarcoma and sino-nasal meningioma. The first case had demonstrated radiological features suggestive of sinonasal meningioma, i.e. (1) heterogeneously enhancing mass containing calcification, (2) tissue characteristics coupled with positive MR spectroscopy which is high alanine peak and (3) marked enhancement of tumour which was supplied by the branches of the external carotid artery [7]. However at the time of writing, no literature had reported on the characteristics and vascular pattern of cementoossifying fibroma on conventional or MR angiography. MR Spectroscopy in the first case showed a high alanine peak at 1.5 ppm. We thought that this could be due to the high content of calcium and fibrous tissue.

The recommended treatment of cementoossifying fibroma is excision with or without bone grafting. The entire tumour should be removed including regions of the involved orbital floor and maxillary sinus walls. Radiotherapy is contraindicated because of its radioreistance and post-radiation complications. The recurrence rate of maxillary cementoossifying fibroma is unknown however it has been reported in as many as 28% of mandibular cementoossifying fibroma [4].

In our opinion, angiography would be useful to assess the vascularity of the tumour. In our two patients, embolization had successfully reduced the tumour vascularity consequently minimizing the blood loss during surgery.

References