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Case Report

Ossifying fibroma of the ethmoid sinus: Report of a rare case and review of literature

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Abstract

Ossifying fibroma (OF) is a benign fibro-osseous lesion which was first described by Menzel in 1872. It is commonly seen in the head and neck regions and represents an aggressive pattern when the midface and paranasal sinuses are involved. We report a 36 years old white woman with OF in the right ethmoid sinus. Computed Tomography (CT) scan images showed a hyperdense mass. Transnasal endoscopic resection was performed and histopathological examination confirmed the diagnosis of OF. The present case is notable because involvement of the ethmoid sinus is rare in this condition with only 48 cases reported in the literature until June 2011.

KEYWORDS: Ossifying fibroma, paranasal sinus, ethmoid

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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[1, 2] but the term of "Ossifying Fibroma" was subsequently coined by Montgomery in 1927.[3,4]

The etiology of OF is unknown but odontogenic, developmental and traumatic origins have been suggested.[3, 5] It is commonly found in the tibia and fibula of children 10 years or younger[6] but young adult individuals with an average age of 20 to 30 years are the most commonly affected groups[3] with involvement of the head & neck region. In this site, the lesions arise in the mandible in about 62% to 89% of patients[7] followed by the maxilla and rarely the orbit, skull base and calvarium.[8] Women are affected more often than men with a female to male ratio of 2:1.[9] Ossifying fibroma of the sinonasal tract occurs at a slightly older age (3rd to 4th decade of life), and preferentially in black women.[10] There is no evidence of hereditary predominance.[9]

Involvement of the sinonasal tract is extremely rare and only 48 cases were reported in the literature from 1971 to 2011 on the basis of a search in Pubmed. The reported cases were between 3 weeks and 41 years of age, and 23.7% to 56% were female.[1, 3-7, 9-38]

OFs manifest typically as painless slow growing tumors but extramandibular lesions such as those occur in the paranasal sinuses and midface tend to display more aggressive behaviour and rapid growth.[3] Radiologically, such lesions are typically well-defined unilocular radiolucencies with scattered radiopaque foci.[39] Pathologically, they are characteristically sharply demarcated lesions, containing fibrous tissue and trabeculaes of osteoid and lamellar

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bone or basophilic spherules that resemble cementum[40] with no mitotic activity.[9]

Mandibular OFs are usually resected using curettage with favourable results. Open or endoscopic en bloc resection is the treatment of choice in the extramandibular OFs regarding to recurrence and deleterious effects when OF is located in these regions.[13] Here, we presented a rare case of OF in the right ethmoid sinus and reviewed the literature of this rare tumour.

Case Report

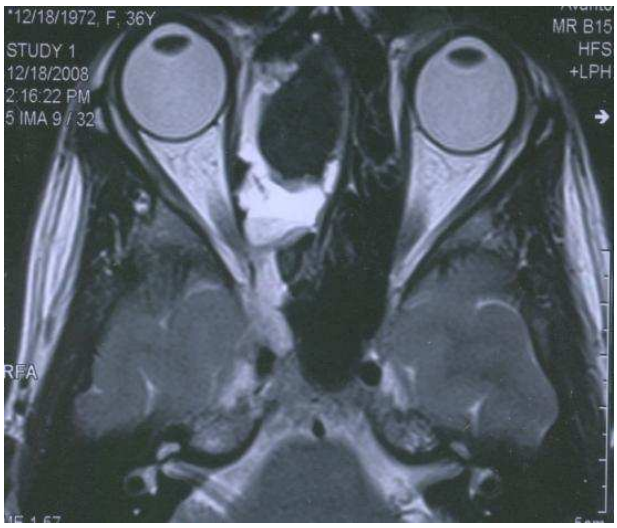
A 36-year-old woman was referred to the otolaryngology clinic of Taleghani Hospital, Shahid Beheshti University of Medical Sciences, with a right intranasal mass (January 2009). This was an incidental finding on follow-up Magnetic Resonance Imaging (MRI) which was advised her due to a history of pituitary microadenoma and galactorrhea from 1 year ago. The patient has been treated with Bromocriptine and her serum prolactin level was at normal range of value. She did not suffer from hyposmia, epistaxis, nasal obstruction, any visual disturbances, epiphora and headache.

On physical examination, there was no sign of proptosis or facial swelling but anterior rhinoscopy revealed a nonhemorrhagic mass in the right middle meatus, with the same color of the surrounding mucosa and with deviation of the nasal septum to the left side.

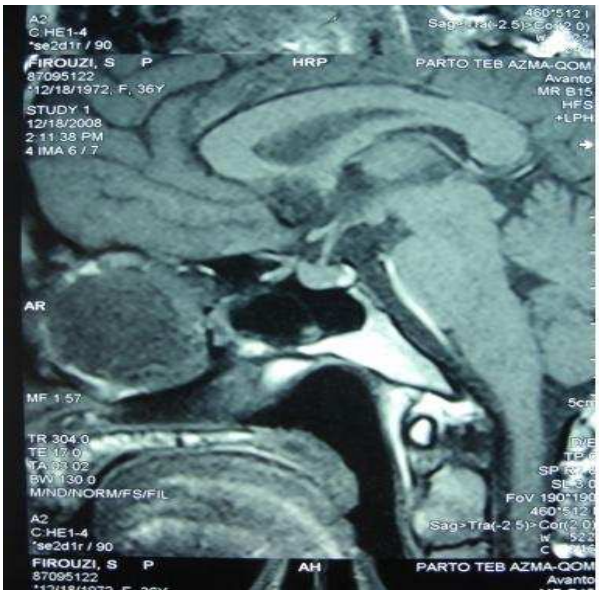
Computed Tomography (CT) scan images revealed a large calcified hyperdense mass in the right ethmoid sinus which obliterated the middle meatus. Fluid accumulation in the right frontal air cells as well as both maxillary sinuses was also detectable but the orbital walls appeared intact without any lytic or erosive lesion. Laboratory values were normal.

An operative procedure was performed with a transnasal endoscopic approach. Due to proximity of this lesion to the base of the skull and attachment to the lamina papyracea, en bloc resection was not advisable. Therefore, the lesion was removed in several fragments. Histopathological examination reported abundant irregular immature (woven) bone partially surrounded by an osteoblastic layer within fibrous connective tissue which was consistent with an ossifying fibroma.

Endoscopic evaluation of the right nasal cavity showed complete wound healing 3 months after surgery and the patient was clinically well. Unfortunately, the patient did not come back for follow up imaging that was advised her 6 months postoperatively.

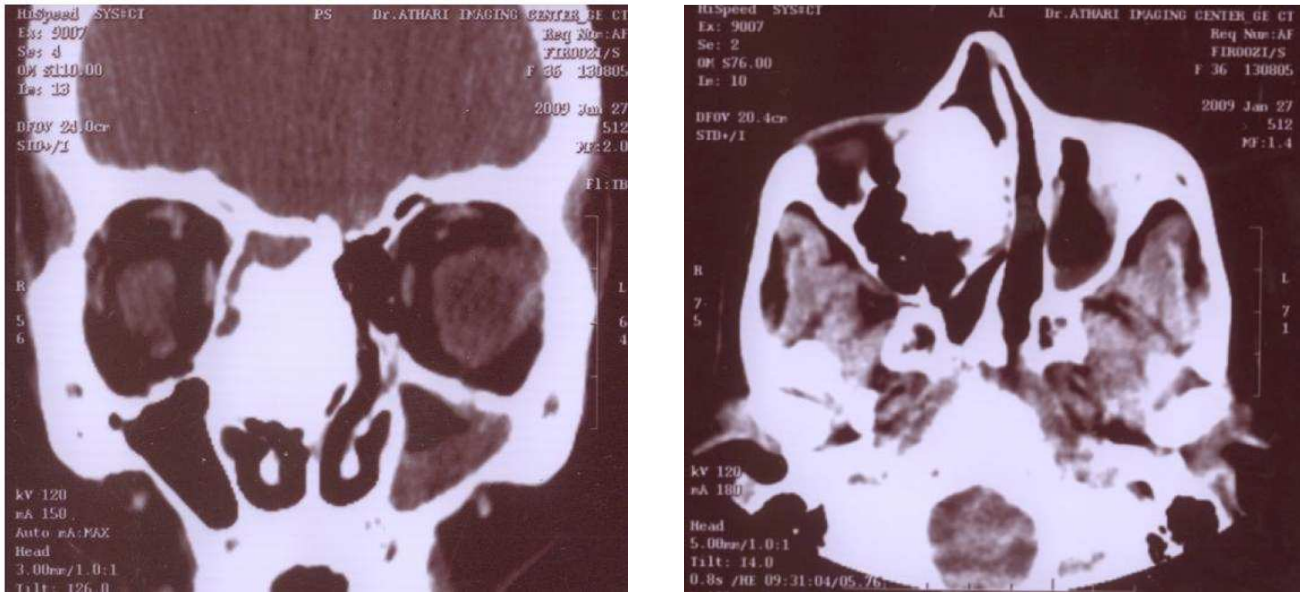


A



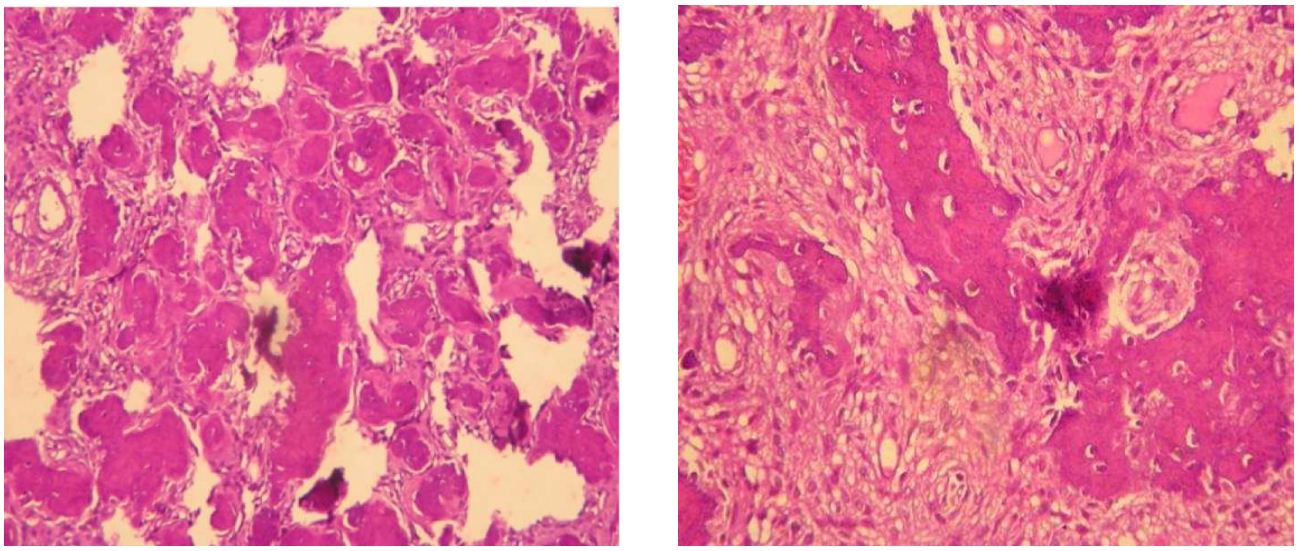
B

Figure 1. A: Coronal T2 weighted MRI image that reveals a mass in the right ethmoid and nasal cavity region.
B: Sagittal T1 weighted MRI image that reveals pituitary microadenoma and sinonasal mass.



A B

Figure 2. A and B, Coronal & axial CT scan of the paranasal sinuses demonstrate a calcified mass in the right ethmoid sinus which obliterated middle meatus.



A B

Figure 3. C and D, Histopathologic sections show irregular immature (woven) bone partially surrounded by an osteoblastic layer within a fibrous connective tissue.

Discussion

Ossifying fibroma is one of the benign fibro-osseous lesions.[8] Although, they are common in the mandible, occurrence in the paranasal sinuses is rare[3, 14] with only 48 reported cases in the literature.[1, 3-7, 9-38]

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.[1, 40]

As previously mentioned, the etiology of OF is unknown[3, 13] but these lesions are presumed to originate from periodontal ligaments

of teeth because of their capacity to produce cementum and osteoid material.[13] Other theories include traumatic and developmental causes. Brademann et al[41] 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. Today, 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 cementyfing fibroma are the same lesions and classified best as osteogenic neoplasm.[40] Accordingly, development of OF in long bone can be explained in this manner. The ethmoidal location of OF in this case may also be explained by incomplete migration of the mesenchymal and its differentiation into periodontal membrane.[42]

Clinical presentation

The clinical presentation of these tumors is variable, depending on the site and rate of growth.[8] It ranges from an asymptomatic bone lesion found incidentally on imaging taken for another reason as in our case, 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. While pain and paresthesia are rarely associated with an ossifying fibroma[40] where the temporal bone is involved, the patient may complain of pain, pulsatile tinnitus, otorrhea with progressive hearing loss. Meningitis and pneumocephalus are two rare intracranial complications of ossifying fibroma.[8]

Imaging

In radiographic studies, the initial lesions may exhibit unilocular radiolucencies and somewhat sclerotic borders[3] with gradual transformation to radioopacitiy.[8, 40] CT slices may clarify well-circumscribed lesions. The central area consists of a nonhomogenous matrix with "ground-glass" opacification representing diffuse calcifications and low attenuation areas containing fibrous tissue with possible contrast enhancement. The walls of the involved sinuses may undergo further remodelling and thickening, sometimes along with erosions.[1, 8]

Radionuclide scintigraphy bone scan typically show well-circumscribed area of homogenously intense uptake at the site of the lesion, suggesting with intense osteoblastic activity.[8] Fibroosseous lesions usually have low to intermediate signal intensity on T1 weighted MR imaging usually isointense to muscle and variable signal intensity on T2 weighted images. Ossified areas may appear with low signal intensity and central nonossified areas, cyst or associated mucocele as high signal intensity areas with enhancement on post-Gadolinium images.[4] Magnetic Resonance Imaging is particularly useful for ruling out an intracranial or intraorbital extension.[38] Angiography may be useful to assess the vascularity of the tumour. Embolization may reduce tumour vascularity which may lessen intraoperative blood loss.[7]

Differential diagnosis

Differential diagnoses include fibrous dysplasia, sinonasal psammomatous meningioma, and well-differentiated osteosarcoma.[10] 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.

Radiographically, FD tends to have more diffused margins. Clinically, FD displays self-limited growth with skeletal maturation and often reveals cessation of growth once adulthood is reached.[10] The histological features include irregular shaped trabeculae of osteoid or woven bone ("Chinese letter" configuration) dispersed in a fibrous stroma with variable cellularity, which is directly fused to peripheral normal bone.[10, 40] FD is often polyostotic whereas ossifying fibroma lesions are monostotic. The fibrous stroma in FD is often less vascular and cellular than OF without os-

teoblastic rims,[3] but OF usually presents it.[40] In contrast to OF and other fibroosseous lesion, FD typically demonstrate a rather monotonous pattern throughout the lesions.[40] In general, it is difficult to distinguish them clinically and histopathologically but computed tomography can be helpful in the diagnosis of OF.[41]

Psammomatous meningioma is not a true fibro-osseous lesion and it may show no identifiable connection with the central nervous system (primary/ectopic) or may extend from the central nervous system (secondary). They appear as polypoid masses that cause nasal obstruction.[14]

Meningioma is distinguished from OF by the presence of whorls of meningothelial spindle cells; frequently with empty-appearing nuclei. Psammoma bodies lack the osteoblastic rimming seen in the calcified ossicle of Ossifying fibroma. The diagnosis of meningioma can be confirmed by dual immunoreactivity of the meningothelial cells for epithelial membrane antigen (EMA) and the mesenchymal marker vimentin. The stromal cells in OF and FD may be vimentin positive but lack immunoreactivity for EMA.[2] On imaging meningioma there is heterogenously enhancing mass containing calcification and MR spectroscopy of the lesion shows a high Alanine peak.[7]

Craniofacial osteosarcomas are destructive, poorly defined, osteolytic, osteosclerotic or mixed lesions. Anaplastic tumor cells are seen histopathologically admixed with areas of osteoid formation. They are aggressive tumors that are prone to local recurrence and distant metastasis.[10]

Treatment

Most lesions are treated with excision or curettage with or without bone grafting.[2] Total removal must be performed to avoid recurrence.[1] Based on the present knowledge, the obviously much more aggressive juvenile ossifying fibroma (JOF) should always be totally resected but for the asymptomatic paranasal ossifying fibroma of the adult (COF), a wait-and-scan strategy might be chosen, as recommended for paranasal osteoma and fibrous dysplasia.[38] If it is not possible, complication such as blindness, orbital cellulites, cysts, intracranial extension, meningitis, cerebritis and pneumocephalus can occur. Radiotherapy is contraindicated for OF because it may increase malignant transformation rate from 0.4% to 40%.[1]

Based on improved endoscopes and instruments, the further development of operation techniques (e.g. four-hand-technique) and the routine application of precise navigational systems, the limits of the endoscopic approach could be extended over the last decades.[38] Endoscopic resection of sinonasal OF is an excellent therapeutic option when done by an expert surgeon, as also was successfully performed in this case. Advantages include direct visualization, magnification, no external deformity and decreased morbidity. Complication includes skull base injury with resultant cerebrospinal fluid leakage, which can be repaired endoscopically in the same setting.[13] The craniofacial approach has become the accepted surgical route for large tumors which have breached the anterior cranial fossa and extend into the cranial cavity.[5] Postoperative management must include repeated endoscopic examination and imaging specially in partially excised cases to detect recurrence and prevent long term morbidity.

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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Conflict of Interests

Authors have no conflict of interests.

Authors' Contributions

Sareh Farhadi **(contribution percentage: 30%)** & Behzad Abedin **(contribution percentage: 20%)** obtained the documents of patient, prepared the manuscript and revised it, Zhaleh Mohsenifar **(contribution percentage: 20%)**, Fatemeh Mashhadi Abbas **(contribution percentage: 10%)** and Saeed Allah Nouhi **(contribution percentage: 20%)** read the manuscript and edited it.

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