

**CLINICAL CASE****Ossifying fibroma of the nose,  
ethmoid and maxillary sinuses****Codruț Sarafoleanu, Claudiu Manea, Raluca Enache**

ENT&amp;HNS Department "Sfanta Maria" Hospital, Bucharest, Romania

**ABSTRACT**

**BACKGROUND:** Ossifying fibroma is a benign lesion commonly found in the mandible, but it may also involve the maxilla and the paranasal sinuses. Being a locally aggressive tumor, with a high neoplastic potential, the complete surgery excision is the treatment of choice.

**MATERIAL AND METHODS:** We report a 6-year-old boy presenting with a voluminous tumor occupying the left nasal fossa extended to the ethmoid and maxillary sinuses, which later was diagnosed as ossifying fibroma.

**RESULTS:** The nature of the disease, the diagnosis and treatment protocols are presented. A transnasal endoscopic approach was performed with a piece-meal resection of the tumor.

**CONCLUSIONS:** The diagnosis of an ossifying fibroma consists in a combination of clinical, radiological and pathological criteria. Complete surgical excision of the ossifying fibroma of the nose and sinuses by endoscopic approach can be the treatment of choice in selective cases.

**KEYWORDS:** ossifying fibroma, endoscopic approach, ethmoid sinus, maxillary sinus

**INTRODUCTION**

Ossifying fibroma is a relatively uncommon benign fibro-osseous tumor affecting the cranio-facial region. It mainly develops in the mandible (75% to 89% of the cases<sup>1</sup>), but it may also involve the paranasal sinuses. This lesion most often occurs during the 2<sup>nd</sup> to 4<sup>th</sup> decades of life, with a predilection to men (male/female = 5:1)<sup>2</sup>. Histologically, it consists of fibro-cellular tissue, well demarcated by the surrounding bone<sup>3</sup>. It is usually asymptomatic, but with a high aggressiveness and recurrence rate. Due to these characteristics an early diagnosis and complete surgery excision are needed.

**CASE REPORT**

A 6-year-old boy patient presented with a 6-month history of left nasal obstruction, anterior left rhinorrhea, associated with left periorbital swelling and left-sided epiphora, symptoms that appeared 2 weeks prior to examination.

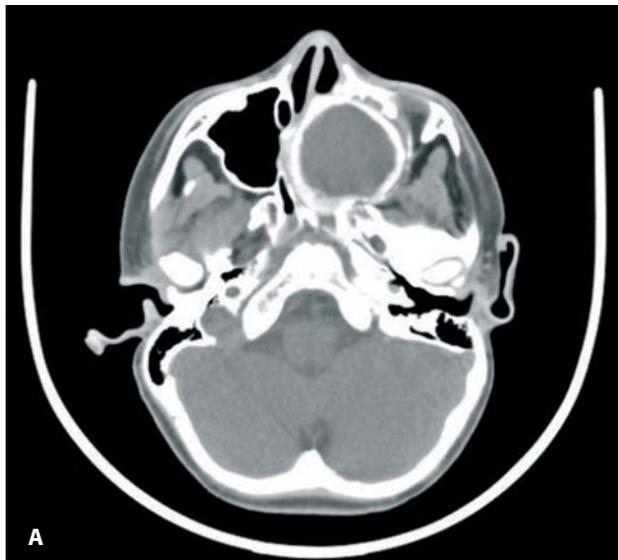
Clinical examination revealed, despite the periorbital swelling, no alteration of the visual field and normal ocular motility in all directions.



**Figure 1** Endoscopic view of the tumor (white arrow) occupying the left nasal fossa coated with a reddish mucosa

The nasal and nasopharyngeal endoscopic examination revealed a large, reddish tumor occupying the left nasal fossa (Figure 1) and a global septum deviation to the right nasal fossa.

Following the diagnosis protocol a CT scan was performed (Figure 2A) which was suggestive for a well-defined soft-tissue density mass surrounded by an eggshell – thin rim and occupying the left nasal fossa, with ex-



**Figure 2** CT scan (axial view - A) and MRI (coronal view - B) - tumor occupying the left nasal fossa, with an eggshell-thin rim of bone surrounding, involving the left maxillary sinus and ethmoid, compressing the posterior part of the nasal septum, with the tendency of orbital invasion.

tension towards the left ethmoid and maxillary sinuses and with a compression effect upon the posterior part of the nasal septum.

The radiologic examination was completed by the MRI (Figure 2B) which showed a tumor with an intermediate signal density, with significant contrast enhancement that confirmed the tumor tendency of orbital invasion.

The treatment strategy consisted in transnasal endoscopic approach. Due to the large size of the tumor, only a piece-meal resection was possible in order to avoid destruction of the surrounding structures (Figure 3). During the surgery we discovered that the lamina papyracea was eroded and this is why it was entirely removed leaving a dehiscence area of the medial orbital wall, but with normal orbital content.

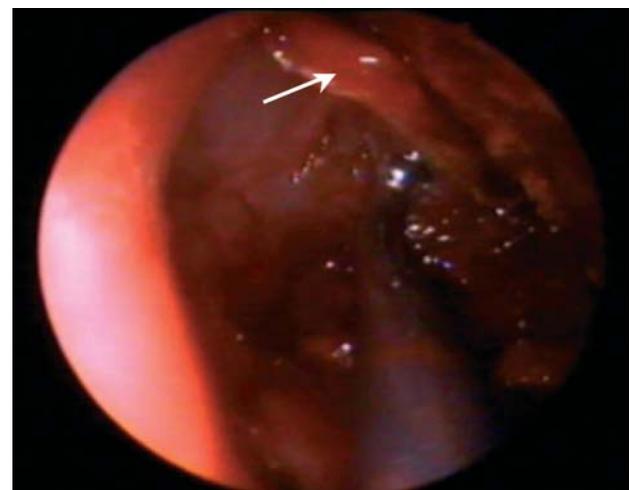
The frozen sections and the final histopathologic examination revealed an ossifying fibroma.

The postoperative period was free of complications and the patient was discharged five days after the surgery.

The nasal endoscopic examination performed at 6 months after surgery showed no signs of local recurrences.

## DISCUSSION

Ossifying fibroma is a slow growing, destructive, benign lesion that can occur almost anywhere in the facial skeleton. Sloopweg reported that maxilla as being the most frequent site<sup>4</sup>. In the same time, Johnson reported that 90% of the ossifying fibroma of the facial bone de-



**Figure 3** Endoscopic view during the resection of the tumor, with the eggshell-thin rim of bone (white arrow)

velops in the paranasal sinuses and only 10% in the mandible<sup>5</sup>.

This uncommon tumor can present a diagnosis dilemma for both the clinician and pathologist because of the different histopathologic features. During time, ossifying fibroma was referred to as psammomatoid ossifying fibroma, psammoosteoid fibroma, juvenile ossifying fibroma, juvenile active ossifying fibroma, cementifying or cemento-ossifying fibroma<sup>6</sup>.

Many pathologists consider the ossifying fibroma a variant of fibrous dysplasia. Even though some authors opine that ossifying fibroma and fibrous dysplasia cannot be differentiated by microscopy due to the histological overlap, there are several parameters used for

the histopathologic distinction between one and another: prevalence of osteoblastic rimming around the bone trabeculae, lesional circumscription and variability in tissue composition within the lesion from one microscopic field to another are said to favor the diagnosis of ossifying fibroma<sup>7</sup>. In fibrous dysplasia, the stroma is more collagenized and less cellular, with abortive bony trabeculae and woven trabecular bone. A generally uniform bone-to-fibrous tissue ratio with fairly evenly scattered bone trabeculae suggests fibrous dysplasia.

The radiological features of ossifying fibroma may also help in the differential diagnosis. Although the radiography of paranasal sinuses is not out of use, CT scan and MRI are essential tools in the correct management of this tumor. Ossifying fibromas of the head and neck are circumscribed lesions of varying density on CT scan. Even if most sinonasal ossifying fibromas showed high density and a dense, calcified outer shell, some authors<sup>8</sup> found a tumor of low density on CT scan, equal to skeletal muscle with only a few foci of calcification and no outer shell. Also, the MRI is useful in the differential diagnosis between ossifying fibroma and fibrous dysplasia because of the different signals on T1 and T2.

Clinically, the ossifying fibroma is potentially aggressive, with infiltration of the surrounding areas (orbital walls, anterior skull base and anterior cranial fossa). The common clinical features of the lesion arising from the paranasal sinuses and orbit include nasal obstruction, exophthalmia or intracranial manifestation. However, in most patients imaging has revealed displacement rather than destruction of the near bony elements<sup>9,10</sup>. Also, in histology, in several cases, the shell was not part of the proliferating tumor, but represented reactive bony change<sup>10</sup>.

The treatment of choice is surgery. To prevent the recurrences, the complete tumor resection is mandatory, some authors recommending also the resection of a minimal margin of surrounding bone, because of the strong neoplastic potential of ossifying fibroma<sup>11</sup>. This goal can be achieved in open or endoscopic surgery, depending on the localization and extension of the tumor. In our case, the endoscopic approach was challenging, especially because of the age of the patient and the extension of the tumor, and proved to be so far the right choice. Radiotherapy does not play a role in the treatment of this tumor, particularly in a growing child.

The possible complications that can occur due to incomplete tumor resection are such as orbital cellulites, blindness, intracranial extension or meningitis. This is why repeated postoperative endoscopic examination and imaging are mandatory<sup>12</sup>.

## CONCLUSIONS

Ossifying fibromas are tumors with rare sinonasal localization. The clinical, radiologic and pathologic diagnosis protocols are very important tools in the positive and differential diagnosis. The complete tumor resection is mandatory to prevent the recurrences and possible complications. Endoscopic approach can be, in selected cases, the treatment of choice.

## REFERENCES

1. Saito K., Fukuta K., Takahashi K., et al. – Benign fibrous lesions involving the skull base, paranasal sinuses, and nasal cavity. *J Neurosurg*, 1998;88:1116-9.
2. Harnsberger H.R., Wiggins R.H. III, Hudgins P.A. et al. – Ossifying fibroma, sinus. In: Harnsberger H.R., Wiggins R.H. III, Hudgins P.A. et al, eds. *Diagnostic Imaging: Head and Neck*. Salt Lake City: Amirsys, 2004.
3. Tamgadge S.A., Tamgadge A.P., Bhatt D.M., Bhalerao S., Periera T., Gotmare S. – Juvenile ossifying fibroma of the maxilla (case report). *Scientific Journal* 2009;vol III:14-16.
4. Slootweg P.J. - Maxillofacial fibro-osseous lesions: classification and differential diagnosis. *Semin Diagn Pathol* 1996;13(2):104-12.
5. Johnson L.C., Yousefi M., Vinh T.N., Heffner D.K., Hyams V.J., Hartman K.S. - Juvenile active ossifying fibroma. It's nature, dynamics and origin. *Acta Otolaryngol Suppl* 1991; 488:1-40.
6. Johnson L.C. - Juvenile active ossifying fibroma. Its nature, dynamics and origin. *Acta Otolaryngol Suppl*, 1991;488:3-40.
7. Chong V.F.H., Tan L.H.C. – Maxillary sinus ossifying fibroma. *Am J Otolaryngol*, 1997;18:419 –24.
8. Engelbrecht V., Preis S., Hassler W., Lenard H.G. – CT and MRI of congenital sinonasal ossifying fibroma. *Neuroradiology*, 1999;41:526-529.
9. Han M.H., Chang K.E., Lee C.H., Seo J.W., Han M.C., Kim C.W. – Sinonasal psammomatoid ossifying fibromas: CT and MR manifestations. *AJNR*, 1990;12:25-30.
10. Wenig B.M., Vinh T.N., Smirniotopoulos J.G., Fowler C.B., Houston G.D., Heffner D.K. – Aggressive psammomatoid ossifying fibromas of the sinonasal region. *Cancer*, 1995;76:1155-1165.
11. Sweet R.M., Bryarly R.C., Kornblut A.D., Corio R.L. – Recurrent cementifying fibroma of the jaws. *Laryngoscope*, 1981;91:1137-43.
12. Baumann I., Zimmermann R., Dammann F., Maassen M.M. – Ossifying fibroma of the ethmoid involving the orbit and the skull base. *Otolaryngol Head Neck Surg*, 2005;133:158-159.