

CASE SERIES AND REPORTS

Endonasal endoscopic resection of ossifying fibroma involving the ethmoid sinus, orbit and anterior skull base: case report and literature review

Resezione endoscopica di un fibroma ossificante interessante il seno etmoidale, l'orbita e il basicranio anteriore: case report e revisione della letteratura

M. JURLINA¹, N. SKITARELIĆ², D. PASSALI³, F.M. PASSALI⁴, R. MLADINA¹

¹ Department of Otolaryngology Head and Neck Surgery, University Hospital Rebro, Zagreb, Croatia;

² Department of Otolaryngology Head and Neck Surgery, General Hospital Zadar, Zadar, Croatia;

³ Department of ORL, University of Siena, Italy; ⁴ Department of ORL University of Rome Tor Vergata, Italy

SUMMARY

Ossifying fibroma is a benign fibro-osseous tumour that rarely involves the ethmoid sinuses and orbit. It is classified as a benign fibro-osseous lesion, a term that is synonymous with a variety of lesions reported in the literature. Recurrence rate with deleterious effects in cases of extramandibular ossifying fibroma is the impetus for open en bloc resection of the tumour. Continuously evolving techniques in endonasal endoscopic sinus surgery has rendered resection of large benign sinonasal and cephalonasal tumours possible. The authors report a case of ossifying fibroma involving the ethmoid sinus, orbit and anterior skull base in a 65-year-old previously healthy woman completely resected by endonasal endoscopic sinus surgery. The patient was free from postoperative complications and was dismissed from hospital on the sixth postoperative day. At present, the patient is disease-free at a regular five-year postoperative follow-up. Endonasal endoscopic resection of sinonasal ossifying fibromas is an excellent therapeutic option when performed by a surgeon experienced in endoscopic sinonasal surgery. The advantages of an endonasal endoscopic approach include direct visualization, enhanced visibility and magnification resulting in decreased intraoperative and postoperative morbidity. Aesthetic outcome is excellent in the absence of facial scars.

KEY WORDS: Ossifying fibroma ethmoid • Endonasal endoscopic sinus surgery

RIASSUNTO

Il fibroma ossificante è un tumore fibro-osseo benigno che solo raramente interessa il seno etmoidale e l'orbita. Viene classificato come una lesione fibro-ossea benigna, una dicitura che raggruppa una discreta varietà di lesioni riportate in letteratura. Una tendenza alla recidiva con importanti sequele ha rappresentato la spinta verso una resezione open en bloc nelle forme extramandibolari di questo tipo di lesione. La continua evoluzione delle tecniche di endoscopia endonasale ha reso possibile la resezione delle grandi lesioni benigne nasali e cefalo-nasali. Gli autori descrivono l'asportazione completa di un voluminoso fibroma ossificante interessante seno etmoidale, orbita e basicranio anteriore in una paziente di 65 anni in buone condizioni generali. La paziente non ha avuto complicanze postoperatorie ed è stata dimessa in sesta giornata. La paziente è al momento al quinto anno di follow-up e si presenta libera da malattia. L'asportazione endoscopica del fibroma ossificante endonasale è un'ottima scelta terapeutica nelle mani del chirurgo esperto. I vantaggi della tecnica includono la visualizzazione diretta della neoformazione e la sua maggiore magnificazione, che portano a una riduzione delle complicanze intra e postoperatorie. L'outcome estetico è ovviamente eccellente per l'assenza di cicatrici.

PAROLE CHIAVE: Fibroma ossificante • Etmoide • Chirurgia endoscopica nasale

Acta Otorhinolaryngol Ital 2016;36:144-148

Introduction

Ossifying fibroma (OF) is a rare, benign fibro-osseous tumour typically found in the mandible, and therefore has been reported mainly by oral surgeons^{1,2}. About 10% of all cases are found in the maxilla³. OF rarely involve the ethmoid sinuses and orbit^{1,4,5}. The authors present an unusual case of OF involving the left ethmoid sinus, or-

bit and anterior skull base occurring in an elderly woman who was completely resected by endonasal endoscopic sinus surgery.

Case report

A 65-year-old previously healthy woman who presented with the sensation of pressure in the left orbit. She did not

refer any disturbances of smell or vision. The patient was not taking any medications and had no known drug allergies. Ophthalmologic examination showed no pathologic findings. There was no conjunctival oedema or haemorrhage. The showed any pathologic changes.

Endoscopic evaluation of the left nasal cavity showed oedema within the left middle meatus. Multi-slice computed tomography (MSCT) of the paranasal sinuses demonstrated an ovoid mass expanding within the left ethmoid complex. On coronal MSCT, the tumour was seen to occupy the left nasal cavity and eroding the lamina papyracea. It was attached to the anterior skull base by a narrow, stalk like base (Fig. 1).

Based on the physical examination findings and imaging studies, the patient was admitted to the hospital and prepared for surgery. The patient was operated under general, hypotensive anaesthesia. During the procedure, the tumour was found to be attached to the anterior skull base and eroding the left lamina papyracea. A 30° endoscope was used to proceed to delicate preparation of the anterior skull base. The tumour was completely resected endoscopically using straight powered drill bits with clear intraoperative presentation of cribriform plate. The intraoperative tumour is presented in Figure 2. The left periorbita and skull base were not surgically violated and orbital fat did not protrude into the surgical field at the end of procedure.

There were no postoperative complications. Routine postoperative endoscopic examination of the left sinonasal cavity was performed and no evidence of cerebrospinal fluid leaks from the anterior cranial fossa was present.

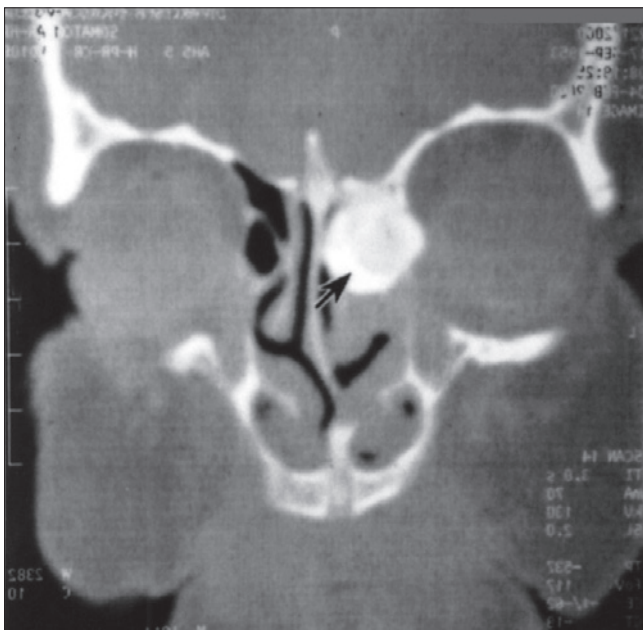


Fig. 1. Preoperative coronal MSCT scan showing the tumour occupying the left nasal cavity and eroding the lamina papyracea. It was attached to the anterior skull base by a narrow, stalk-like base (black arrow).



Fig. 2. Intraoperative tumour specimen.

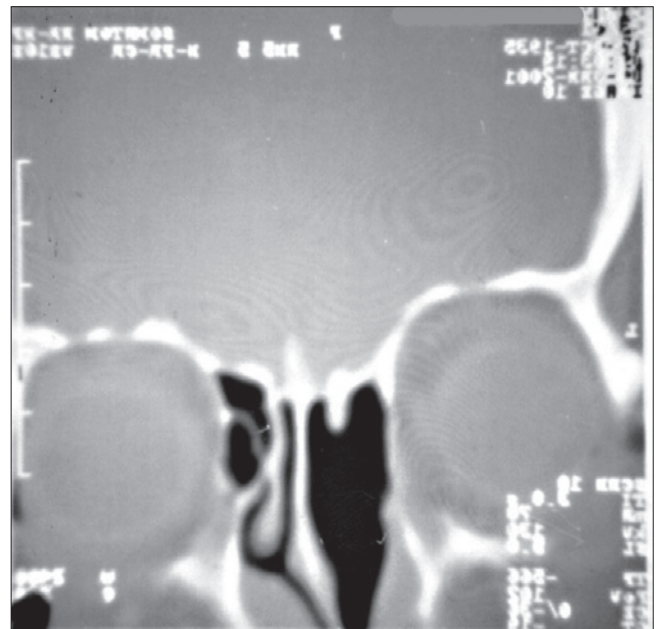


Fig. 3. Postoperative coronal MSCT scan at one year postoperative follow-up.

The histopathological finding was consistent with an OF. The tumour mainly consisted of two components: a fibrous stroma, rich in fibroblasts and small vessels surrounded with bony lamellae that were rimmed by osteoblasts and occasionally by osteoclasts.

A postoperative coronal MSCT scan after one year postoperative follow-up is shown in Figure 3. The patient is presently completely free of disease at a regular five-year follow-up.

Discussion

OF holds a tenuous place in the classification of bony lesions. It was first mentioned by Menzel in 1872, who considered it to be a form of osteoma⁶. OF is classified

as a benign fibro-osseous lesion, a term that is synonymous with a variety of lesions reported in available literature (Table I).

The origin of OF is debated with a predominant theory claiming the tumour originates from periodontal roots because of their capacity to produce cementum and osteoid tissue⁶. The tumour has the capability of producing cement, lamellar bone and fibrous tissue in widely differing proportions. The periodontal membrane is a mesodermal germ layer product. The normal migration of the medial part of nasal anlage occurs through the ethmoid sinus region, and a small portion of this mesenchyme differentiates into the periodontal membrane⁶. The ethmoidal localisation of OF could therefore be explained by incomplete migration and maturation of the periodontal membrane. Marvel et al. stated that OF originates from primitive mesenchymal cells that are believed to produce cementum at sites distant from odontogenic tissue⁷.

Both benign and malignant fibro-osseous lesions should be considered in the differential diagnosis of OF. These include well-differentiated osteosarcoma, and the spectrum of fibro-osseous lesions of the head and neck⁸.

Regardless of the theory of origin, fibro-osseous lesions and OF differ from fibrous dysplasia not only histologically, but also in their distinct clinical behaviour. Some authors have suggested that sharply defined calcifying spherulation is an important differential finding not seen in fibrous dysplasia, but most pathologists agree that it is very difficult to differentiate between these two entities using histological criteria alone. No hereditary tendencies have been observed, and no pigmentary or endocrine changes have been reported associated with ossifying fibromas. Moreover, abnormal serum calcium, phosphorous or alkaline phosphatase levels have not been reported.

OF is usually well circumscribed rather than diffuse and most often involves a single bone. The tumour produces moderate expansion of the tables of the involved bone, but usually leaves a thin "egg shell" boundary on either side with no periosteal reaction. It has definite bounda-

ries but is not truly encapsulated⁹. All previously mentioned features were present in our case.

OF typically presents in the mandible (75%) and thus is usually reported and treated by oral surgeons. Other reported locations of OF are maxilla, frontal bone, sphenoid bone, ethmoid bone, temporal bone, orbit, anterior cranial fossa and auricula^{1 5 6 8-12}. The otolaryngologist's concern lies with the extramandibular presentation because OF is believed to behave more aggressively than its mandibular counterpart and requires complete surgical resection¹³.

Presenting symptoms of OF depend on the location of the tumour and range from nasal obstruction to disfigurement. Individual patients may present with ophthalmologic symptoms, such as proptosis and diplopia. Intracranial extension of OF fibroma itself or associated mucoceles may give rise to neurological symptoms¹⁴. Patients with OF range from newborns¹⁴ to those in their eighth decade of life, with more aggressive behaviour at an earlier age¹⁵⁻¹⁷. The highest incidence of OF is reported between the ages of 20 to 40 years with a female predilection^{7 18 19}.

Although malignant transformation in OF has not been reported¹, surgery is the treatment of choice²⁰. Radiotherapy is contraindicated for OF because it may increase the possibility of malignant transformation. Reported rates of malignant transformation range from 4% to 40%⁴. Mandibular OF is traditionally treated with curettage because of the favourable results in this particular anatomical location. Recurrence rate with deleterious effects when OF is located in extramandibular regions is the impetus for open en bloc resection of the tumour^{3 7 21 22}. Complete resection of extramandibular OF is curative, and the exact surgical approach depends on the location and extent of the tumour. For the accomplishment of that task a variety of open approaches have been reported including: Caldwell-Luc operation, curettage, with peripheral ostectomy when the tumour is located in the floor of the maxillary sinus; lateral rhinotomy with medial maxillectomy when the tumour is located in the medial wall of the maxillary sinus; external ethmoidectomy for recurrent ethmoid tumour, transcranial/subcranial approach, and subfrontal approach^{1 3 4 7 23 24}.

Continuously evolving techniques in endonasal endoscopic sinus surgery have made resection of large benign sinonasal and cephalonasal tumours possible^{25 26}. A total of 15 cases of sinonasal OF endoscopically treated have been published in the available literature^{12 27-30}. The advantages of endonasal endoscopic approach include direct visualisation, enhanced visibility and magnification resulting in decreased intraoperative trauma and postoperative morbidity. The advent of 3D navigation devices further enhances previously mentioned advantages. Aesthetic outcome is excellent in the absence of facial scars. Postoperative surveillance is accurate and flaw-

Table I. Various terms reported in the literature for different histological variations of ossifying fibromas.

Ossifying fibroma
Cementifying fibroma
Cemento-ossifying fibroma
Desmo-osteoblastoma
Psammo-osteoid fibroma
Psammonmatoid ossifying fibroma
Juvenile ossifying fibroma
Juvenile aggressive ossifying fibroma
Juvenile active ossifying fibroma

less. All these features make an endoscopic approach a preferable option wherever possible. Complications of the endoscopic technique include injury to the skull base with resultant cerebrospinal fluid leak, infection, bleeding and injury to orbital structures²⁷⁻³¹.

Conclusions

In this report, we present a case of a 18 mm OF that was completely resected endoscopically without complications or recurrence after five-year follow-up. The tumour's well-demarcated borders allowed for complete resection and ensured assurance of tumour-free margins. The unusual feature of OF in our case was its adherence to the anterior skull base in the form of a narrow stalk and without dural involvement. It significantly complicated the operating procedure because of imminent endocranial complications. The tumour was successfully and completely drilled off from the skull base and further extracted through the choanae and mouth. Endonasal endoscopic resection of sinonasal ossifying fibromas is an excellent therapeutic option when performed by a surgeon experienced in endonasal endoscopic sinus surgery. The endoscopic examination also provides excellent postoperative surveillance for early detection of recurrence. It should be anticipated as a treatment of choice in case of sinonasal OF with hesitation only in cases of extensive orbital or intracranial extension.

References

- Nakagawa K, Takasato Y, Ito Y, et al. *Ossifying fibroma involving the paranasal sinuses, orbit, and anterior cranial fossa: case report*. Neurosurgery 1995;36:1192-5.
- Godt A, Gulicher D, Kalwitzki M, et al. *Dislocation of an upper third molar by an ossifying fibroma – case report*. J Craniomaxillofac Surg 2008;36:360-4.
- Cox VS, Rimell FL, Marenttete LJ, et al. *Ethmoidal cemento-ossifying fibroma: The transglabellar/subcranial approach*. Otolaryngol Head Neck Surg 1996;114:335-8.
- Vaidya AM, Chow JM, Goldberg K, et al. *Juvenile aggressive ossifying fibroma presenting as an ethmoid sinus mucocele*. Otolaryngol Head Neck Surg 1998;119:665-8.
- Oukabli M, Akhaddar A, Qamouss O, et al. *Nasoethmoidal psammomatoid cemento-ossifying fibroma with intraorbital extension*. Rev Stomatol Chir Maxillofac 2010;111:43-5.
- Brademann G, Werner JA, Janig U, et al. *Cemento-ossifying fibroma of the petromastoid region: case report and review of the literature*. J Laryngol Otol 1997;111:152-5.
- Marvel JB, Marsh MA, Catlin FI. *Ossifying fibroma of the mid-face and paranasal sinuses: diagnostic and therapeutic considerations*. Otolaryngol Head Neck Surg 1991;104:803-8.
- Kaufmann RA, Padmore RF, Ridge JA, et al. *Primary extraosseous cemento-ossifying fibroma of the auricle*. Head Neck 1999;21:578-81.
- Thomas GK, Kasper KA. *Ossifying fibroma of the frontal bone*. Arch Otolaryng 1966;83:69-72.
- Sterling KM, Stollman A, Sacher M, et al. *Ossifying fibroma of sphenoid bone with coexistent mucocele: CT and MRI*. J Comput Assist Tomogr 1993;17:492-4.
- Shetty SD, Salib RJ, Nair SB, et al. *Ossifying fibromyxoid tumour of the sphenoid sinus*. J Laryngol Otol 2010;124:437-40.
- Singhal A, Ram R, Singhal P, et al. *Cemento-ossifying fibroma of maxillary antrum in a young female patient*. Indian Soc Pedod Prev Dent 2011;29:44-7.
- London SD, Schlosser RJ, Gross CW. *Endoscopic management of benign sinonasal tumors: a decade of experience*. Am J Rhinol 2002;16:221-7.
- Al-Mazrou KA, Mansoor A, Payne M, et al. *Ossifying fibromyxoid tumor of the ethmoid sinus in a newborn: report of a case and literature review*. Int J Pediatr Otorhinolaryngol 2004;68:225-30.
- Johnson LC, Yousefi M, Vinh TN, et al. *Juvenile active ossifying fibroma: its nature, dynamics and origin*. Acta Otolaryngol Suppl 1991;488:1-40.
- De Riu G, Meloni SM, Raho MT, et al. *Complications of mandibular reconstruction in childhood: report of a case of Juvenile Aggressive Fibromatosis*. J Craniomaxillofac Surg 2006;34:168-72.
- Bohn OL, Kalmar JR, Allen CM, et al. *Trabecular and psammomatoid juvenile ossifying fibroma of the skull base mimicking psammomatoid meningioma*. Head and Neck Pathol 2011;5:71-5.
- MacDonald-Jankowski DS, Li TK. *Ossifying fibroma in a Hong Kong community: the clinical and radiological features and outcomes of treatment*. Dentomaxillofac Radiol 2009;38:514-23.
- Efune G, Perez CL, Tong L, et al. *Paranasal sinus and skull base fibro-osseous lesions: when is biopsy indicated for diagnosis?* Int Forum Allergy Rhinol 2012;2:160-5.
- Wenig BM, VinthTN, Smirniotopoulos JG, et al. *Aggressive Psammomatoid Ossifying Fibromas of the Sinonasal Region*. Cancer 1995;76:1155-65.
- MacDonald-Jankowski DS. *Ossifying fibroma: a systematic review*. Dentomaxillofac Radiol 2009;38:495-13.
- Verma P, Rarthore PK, Mrig S, et al. *Cemento-ossifying fibroma of the maxilla: a case report*. Indian J Otolaryngol Head Neck Surg 2011;63:38-40.
- Chong VFH, Tan LHC. *Maxillary sinus ossifying fibroma*. Am J Otolaryngol 1997;18:419-24.
- Shekhar MG, Bokhari K. *Juvenile aggressive ossifying fibroma of the maxilla*. J Indian Soc Pedod Prev Dent 2009;27:170-4.
- Fei Z, Zhang X, Jiang XF, et al. *Removal of large benign cephalonasal tumours by transbasal surgery combined with endonasal endoscopic sinus surgery and neuronavigation*. J Craniomaxillofac Surg 2007;35:30-4.
- Ooi EH, Glicksman JT, Vescan AD, et al. *An alternative management approach to paranasal sinus fibro-osseous lesions*. Int Forum Allergy Rhinol 2011;1:55-63.
- Brodish BN, Morgan CE, Sillers MJ. *Endoscopic resection of fibroosseous lesions of the paranasal sinuses*. Am J Rhinol 1999;13:111-6.
- Choi YC, Jeon EJ, Park YS. *Ossifying fibroma arising*

- in the right ethmoid sinus and nasal cavity.* Int J Pediatr Otorhinolaryngol 2000;54:159-62.
- ²⁹ Garrett P, Stilianos MS, Kountakis E. *Endoscopic resection of large sinonasal ossifying fibroma.* Am J Otolaryngology 2005;26:54-6.
- ³⁰ Selmani Z, Anttila J, Mertakorpi J, et al. *Cemento-ossifying fibroma of the ethmoidal sinus in a child presenting with isolated pain in the nasal region.* J Craniofac Surg 2004;15:215-7.
- ³¹ May M, Levine HL, Mester SJ, et al. *Complications of endoscopic sinus surgery. analysis of 2018 patients – Incidence and prevention.* Laryngoscope 1994;104:1080-3.

Received: February 1, 2015 - Accepted: June 1, 2015