CASE REPORT

Ossifying fibroma of the middle turbinate revealed by infection in a young child

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KEYWORDS
Ossifying fibroma; Ethmoiditis; Functional endoscopic sinus surgery (FESS)

Summary

Introduction: Ossifying fibroma (OF) is a rare benign fibro-osseous tumor, mainly located in the head and neck region. Most often, it affects the mandible but rare involvement of paranasal sinuses has been reported, associated with more locally aggressive behavior.

Case report: We report the case of an 8-year-old boy with OF of the middle turbinate, revealed by ethmoiditis. Total resection was performed on an endoscopic approach. The patient was free of clinical or radiological recurrence at 3 years’ follow-up. This was the youngest patient with OF of the middle turbinate so far reported in the international literature.

Discussion: Presumptive diagnosis is established by clinical examination and CT scan (location, oval-shaped mass, heterogeneous tumor with a thin bony rim). Definitive diagnosis is founded on histological examination (psammomatous bodies, osteoblastic rim, trabecular bone). Treatment in paranasal sinuses of OF is surgical, preferentially on an endoscopic approach. Resection should be as complete as possible to minimize risk of recurrence, especially in sinonasal locations, known to be more aggressive. Ethmoiditis in an unusual age-range should suggest tumoral etiology.

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Introduction

Ossifying fibroma (OF) is, like osteoma or fibrous dysplasia, a benign fibro-osseous tumor [1]. Presentation varies according to location, which is mainly mandibular [1]. It is usually asymptomatic, discovered serendipitously on radiology. The most frequent clinical sign is tumefaction. Sinonasal forms are rarer, but tend to be symptomatic. The usual rhinologic symptomatology is found: nasal obstruction, pain, epistaxis. Locoregional complications, such as ophthalmic involvement, may be found in ethmoid-frontal forms. Radiological assessment is essential to diagnosis, preoperative assessment and assessment of complications. Definitive diagnosis is founded on histologic examination of the specimen, biopsy often proving insufficient. Clinical and radiological surveillance is mandatory, due to the risk of recurrence.

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1879-7296/ $ – see front matter © 2013 Published by Elsevier Masson SAS.
http://dx.doi.org/10.1016/j.anorl.2013.04.002
Case report

An 8-year-old boy presented in Emergency with febrile left palpebral edema. CT explored for ethmoiditis (Fig. 1). The discovery of a large ethmoid mass led to MRI (Fig. 2). Given the tumoral etiology, the patient was referred to our department. Flexible endoscopy found a left middle turbinate mass, covered by normal mucosa. Ophthalmologic examination found isolated exophthalmia. Surgical biopsy diagnosed OF. Functional endoscopic sinus surgery (FESS) was performed. At the end of the procedure, the lamina papyracea and cribiform plate were visible, showing erosion. Postoperative course was simple, with removal of the nasal packing and discharge home at D + 1. Histology confirmed the diagnosis of OF, finding a storiform tumoral proliferation, a bone matrix with numerous spherules and a peripheral osteoblastic rim (Fig. 3). Follow-up comprised clinical and endoscopic examination at 3, 6, 18 and 36 months and CT at 18 and 36 months. At the time of writing, there had been no recurrence.

Discussion

OF was first reported by Menzel in 1872 [2] and described as a clinical entity by Montgomery in 1927 [3].

It is one of the benign fibro-osseous tumors. The more aggressive forms are found in younger patients. OF mainly affects the facial bones (85%) [4], with a clear mandibular predominance (77%) [5]. Mean age at diagnosis is variably reported as 20 to 40 years [3,6] (range, 2 to 63 years [4,7]), with female predominance [2,6]. Sinus involvement is more rare, and mainly concerns the ethmoid. The present patient is the youngest in whom OF of the middle turbinate has been reported [3,8].

Pathognomic symptoms for sinonasal OF are non-specific, but involve slowly progressing tumoral extension. They may be acute, as in the present patient, in case of complications (infectious, visual, endocranial).

Radiologic assessment is fundamental and comprises facial-bone CT or MRI. It guides diagnosis, as the CT characteristics of OF differ from those of other fibro-osseous tumors. OF generally presents as a round or oval mass of heterogeneous content featuring calcium, surrounded by a thin bony cap, pushing back but not invading neighboring tissue. CT also assesses extension and thus the possibility of resection via an endonasal approach. Finally, the preoperative scan serves for comparison during postoperative radiologic surveillance.

Biopsy is rarely diagnostic; complete histologic examination of the surgical specimen is required. The lesion is round or oval, well-contoured by its thin cap, with avascular fibrous cellular struma. The bone is trabecular/laminar/woven, the trabeculae surrounded by osteoblasts [5]. Presence of spherules, known as psammomatous bodies, is highly characteristic.
Ossifying recurrence. Surgery, concerning (such mention)

the resection has been used to replace it, to reduce morbidity.

Adjuvant radiation therapy is contraindicated due to risk of malignant transformation and lack of proven efficacy.

Spontaneous malignant transformation is rare but has been reported, whereas metastasis has not.

Prognosis is thus strictly local, due to complications and the risk of long-term recurrence.

**Conclusion**

Sinonasal ossifying fibroma is a rare benign tumor. Treatment is surgical, and preferably functional endoscopic sinus surgery, indicated for symptomatic lesions or ophthalmic infections or infections complications or any atypical symptomatology (such as ethmoiditis at the age of 8 years). Long-term clinical and radiological surveillance is required due to risk of recurrence.

**Disclosure of interest**

The authors declare that they have no conflicts of interest concerning this article.

**Acknowledgements**

Thanks to Dr Carla Fernandez of the anatomopathology department of the La Timone Hospitals for the pathology slides and their interpretation.

**References**


