Paediatric fibro-osseous lesions of the nose and paranasal sinuses

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Summary

Objective: The term “fibro-osseous lesion” is a generic description for a group of lesions involving the paranasal sinuses and anterior skull base. We aim to improve understanding of the clinical and pathological presentation of fibro-osseous lesions in children.

Methods and results: We report two cases of aggressive “fibro-osseous” lesions arising from paranasal sinuses and anterior skull base in childhood that were successfully managed surgically. We compare our case reports with a review of the available literature and evaluate the management of these lesions.

Conclusions: The clinical behaviour and radiological features of fibro-osseous lesions is variable. Aggressive lesions require a radical surgical approach to ensure complete excision, in spite of an increase in associated morbidity. Incomplete excision of aggressive lesions may result in disease recurrence with severe morbidity or mortality. In contrast a slowly progressive lesion often does not warrant extensive surgical excision. Understanding the nature of fibro-osseous lesions facilitates appropriate clinical management.

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1. Introduction

Fibro-osseous neoplasia is a generic term encompassing several conditions. Makek [1] proposed a ‘working’ classification of fibro-osseous lesions eliminating redundant terminology and taking into account the biological potential of the tumours [1]. They form a spectrum that includes fibrous dysplasia, aneurysmal bone cysts (ABCs), giant (reparative) granuloma, ossifying fibroma, giant cell tumour, fibromyxoma, osteoblastoma and osteosarcoma [2,3]. A distinct group of these fibro-osseous lesions appear to be unique to the nasal cavity, paranasal sinuses and orbit. The nomenclature for this group includes ossifying fibroma, cementifying or cemento ossifying fibroma, psammomatoid ossifying fibroma and juvenile active ossifying fibroma. Although the latter terms suggest a disease of childhood, they are not restricted to the paediatric age group.

The proximity of these lesions to the orbit and anterior skull base and the variety of their clinical and radiological appearance make them a diagnostic and management challenge. The surgical aim should be en bloc surgical excision minimising morbidity.

2. Case reports

2.1. Case 1

An 8-year-old boy presented to the ophthalmologists with proptosis of his left eye noticed after a trivial injury while playing football. At presentation his vision was normal with no ophthalmoplegia and the globe appeared healthy. A CT demonstrated a well-circumscribed bony lesion within the ethmoid air cells, which was expanding into the medial wall of the orbit and was close to the crista galli and skull base. The globe was displaced laterally and it was thought that the lesion represented an ossifying fibroma. He was referred to his local ENT surgeons and underwent a transnasal biopsy. Histology confirmed an “aggressive psammomatoid ossifying fibroma”. During subsequent surgical resection the tumour was approached via a lateral rhinotomy and was found to be compressing the medial rectus and bowing the nasal septum to the right (see Fig. 1). The tumour extended superiorly to the cribiform plate and was found to be abutting the optic nerve. Further dissection was stopped because of the risk to vision and the optic nerve. Postoperatively his vision remained normal.

Four months postoperatively he had slight proptosis of the left eye. A CT scan 4 months later demonstrated a mass within the nasal cavity, which was a similar size to the initial presenting tumour with lateral displacement of the globe. He had no signs of visual disturbance but did have epiphora. He was referred to our centre for further management. He had progressive proptosis of the left eye and his visual acuity had deteriorated to 6/9, which raised concerns that the lesion was affecting the optic nerve. Subsequent visual-evoked potentials supported this suspicion.

He underwent a craniofacial resection of the tumour via a left lateral rhinotomy, bilateral coronal scalp flap and bilateral frontal craniotomy. The whole of the medial wall of the orbit to, and around, the orbital apex and optic nerve, the floor of the paranasal sinuses and a portion of the skull base was excised. A pericranial flap was used to repair the dural defect in the anterior skull base. His postoperative visual acuity was 6/5 with some loss of peripheral vision and his optic disc appeared pale. Follow-up CT scans 3 years postoperatively have shown no evidence of any recurrence and his vision has remained stable (see Fig. 2).

2.2. Case 2

An 11-year-old boy presented with a 10-week history of progressive right-sided proptosis, epiphora, nasal obstruction and periorbital pain. His visual acuity was normal (see Picture 1). Endoscopy showed a lesion based on the anterior skull base. The CT scan had the appearance of an ossifying mass of the right paranasal sinuses, which was biopsied. Histology confirmed a cellular spindle lesion with numerous islands of mineralization and ossification with myxoid areas within mineralised parts. Osteoblastic and osteoclastic activity was seen and the pathologists described it as juvenile cemento ossifying fibroma.