CASE REPORT

Juvenile aggressive cemento-ossifying fibroma of the sphenoid sinus with proptosis: a surgical dilemma

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SUMMARY

The term fibro-osseous lesion has currently grown in popularity as an overall designation for a number of rare, histologically benign head and neck lesions that are made up of bone, fibrous tissue and cementum.

Cemento-ossifying fibroma is a variant of cementifying fibroma and is a fibro-osseous disease. They are usually small innocuous lesions which follow a slow benign course and are commonly seen in the skull bone rather than the sphenoid. It is rare for these tumours to attain large size, behave aggressively, destroy bone and require a radical surgery. One such rapidly growing juvenile cemento-ossifying lesion of sphenoid in our 10-year-old young patient causing proptosis and impaired vision is reported here because of its uncommon nature and its surgical dilemma. Selection of surgical approach to resect this tumour becomes difficult because it is deeply seated and needs a multidisciplinary approach.

BACKGROUND

Cemento-ossifying fibroma belongs to the group of tumour and tumour-like lesions composed of fibro-osseous tissue. It is commonly seen in the skull bone rather than the sphenoid. Incidence of proptosis in this type of tumour is rare. A typical cemento-ossifying fibroma follows a benign, innocuous course, but rarely, in certain instances, can it demonstrate a much more aggressive behaviour and has been designated as juvenile aggressive cemento-ossifying fibroma (JAOF).¹ We describe such a rare case of JAOF of the sphenoid sinus presenting with proptosis.

CASE PRESENTATION

A 10-year-old boy had presented with proptosis and decreasing vision in the left eye of 6 months duration. This was associated with bilateral nasal obstruction.

Craniotomy and deroofting of the optic canal wall was performed by the neurosurgeon following which the patient had further visual deterioration in the left eye and was referred to us for further management. On admission, the best-corrected visual acuity was recorded as 20/20 in the right eye and 20/200 in the left eye. Hertel's exophthalmometry revealed a measurement of 13 (right) and 19 mm (left) suggestive of proptosis in the left eye. All the extraocular movements were normal and full in both the eyes. Anterior and posterior segment examination in the right eye showed normal findings, except for sluggish consensual pupillary reflex. Anterior segment in the left eye showed a relative afferent pupillary defect, while the rest was within

normal limits. The posterior segment in the left eye showed temporal pallor of the optic disc, while the rest of the macula and background was normal. A nasal endoscopy revealed a sphenoidal swelling abutting the choanae. The tumour material was resected and sent for histopathological examination.

INVESTIGATIONS

CT of the paranasal sinus showed an expansile lesion of the sphenoid sinus (6×5×3.4 cm). Superiorly the sinus bulged into the sella turcica displacing the pituitary gland. The bony wall was seen to abut the optic chiasma. Inferiorly, the sinus was seen bulging into the posterior half of the nasal cavity. The left lamina papyracea was seen to be displaced laterally into the left orbit causing proptosis. There was minimal compression of the left optic nerve. The anterior ethmoidal cells were displaced. Posteriorly, the mass was seen to abut the clivus. The bony wall of the expanded sphenoid showed ground glass appearance (figure 1).

Histopathological evaluation of the tissue from the sphenoid sinus revealed a fibro-osseous lesion composed of thin anastomosing and curved anastomosing trabeculae of woven bone and globular cementicles surrounded by unduly cellular fibrous stroma exhibiting occasional mitotic figures. Also seen were features of cystic degeneration, with dense haemorrhage, numerous osteoclast type of giant cells and diffuse infiltration of inflammatory cells, suggesting a diagnosis of JAOF (figure 2).

Figure 1 CT scan of paranasal sinus (coronal cut) showing expansile lesion of the sphenoid sinus.

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Figure 2 Histopathology showing a fibro-osseous lesion characteristic of juvenile aggressive cemento-ossifying fibroma.

DIFFERENTIAL DIAGNOSIS

Fibrous dysplasia.

TREATMENT

The patient underwent an emergency transnasal endoscopic optic nerve decompression on the left side, which helped in reducing the intralesional pressure by uncapping the tumour. Along with cystic tumour marsupialisation, total ethmoidectomy and middle meatal antrostomy were also performed on the left side. However, the approach was converted to a midfacial degloving approach for complete removal of the tumour.

OUTCOME AND FOLLOW-UP

Postoperative exophthalmometry on day 7 revealed 10 mm in the right eye and 18 mm in the left eye, and his best corrected vision improved to 20/32 in the left eye.

The child has been on regular follow-up. He has been asymptomatic with no recurrence at 1 year of follow-up.

DISCUSSION

Cemento-ossifying fibroma of the paranasal sinuses is a rare benign fibro-osseous lesion arising from the periodontal membrane. JAOF is an aggressive variant that primarily occurs in young patients; in total 60–80% have been identified in patients between 5–15 years. Involvement of paranasal sinuses other than sphenoid and craniofacial bones are commonly encountered. JAOF of the sphenoid is comparatively rare² and the origin of a cemento-ossifying fibroma in the sphenoid sinus is uncertain.

One possibility is that it may arise from ectopic periodontal membrane.³

A second explanation may be that since periodontal membrane is of mesodermal origin, primitive mesodermal cells in the sinus could differentiate as periodontal membrane under certain conditions and form cementum containing fibro-osseous lesions.

The aggressive behaviour may be determined by the cell of origin or accentuated growth due to local conditions in the sinus.⁴

Expanding lesions involving the sphenoid sinus manifest themselves by applying pressure on the surrounding structures particularly IV to VI cranial nerves that are closely related to the sinus, resulting in various ophthalmoplegias and retro-orbital pain. Proptosis and hypophyseal compression occur less frequently.⁵

In addition to swelling and asymmetry, other complications include decreased vision, headache, nasal obstruction, epistaxis and mucocoele.

Although most reports indicate a slow growth, there can be aggressive growths, as in our case. Ossifying fibroma of the mid-facial region and paranasal sinuses are more aggressive than those in the mandible.

CT scans and MRI are invaluable in differentiating other fibro-osseous lesions such as fibrous dysplasia and determining the extent of the lesion.

Management of symptomatic fibro-osseous lesion of the cranial bone varies from wait and watch policy to surgical resection via nasal endoscopic surgery or craniofacial resection. Craniofacial resection is required for very aggressive tumours but may have high morbidity. Endoscopic approach is effective, convenient and safe, carries lesser morbidity and provides better cosmetic results. However, recurrence is common.⁶ Because of the aggressive behaviour, a surgical procedure that is more aggressive than that employed for the typical ossifying fibroma is required.⁷ Therefore, wide exposure and excision remains the treatment of choice. Adjuvant therapy such as radiotherapy is contraindicated because these types of lesions are essentially benign and it carries the risk of malignant transformation.⁸

Learning points

- ▶ An aggressive variant of a rare tumour has been highlighted.
- ▶ Commonly seen in other sinuses. But extremely rare in the sphenoid. Hence, it should be considered during the diagnosis.
- ▶ Wide surgical exposure and removal has a good prognosis.
- ▶ By overcoming this diagnostic challenge, it is possible to attain good visual outcome and no recurrence.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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