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Unilateral visual loss resulting from orbital encroachment of an ethmoidal juvenile trabecular ossifying fibroma

NH Ta[1], A Addison[2], B Beigi[3], C Philpott[1]

[1]Norwich Medical School, University of East Anglia, Norwich, UK
[2]Ear, Nose and Throat Department, James Paget University Hospital, Great Yamouth, UK
[3]Ophthalmology Department, Norfolk and Norwich University Hospitals NHS Foundation, Norwich, UK

ABSTRACT

Ossifying fibromas are mainly found in the mandible and maxilla. Reports of them arising in the ethmoid sinuses and orbits are rare. We present a case of an otherwise healthy 20-year-old man with gradual onset of right visual disturbance signified by right relative afferent pupillary defect due to a large unilateral ossifying fibroma arising from the ethmoid sinus compressing the medial half of the right orbit. We emphasise the multidisciplinary combined endoscopic endonasal and external approach to ensure a successful debulking of the fibroma.

KEYWORDS

Fibroma, ossifying – Ethmoid sinus – Sinus surgery – Orbital surgery – Optic nerve

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CORRESPONDENCE TO

Ngan Hong Ta, E: t.ngan@uea.ac.uk

Introduction

Ossifying fibromas are usually slow-growing, benign fibro-osseous lesions. They are formed by normal bone being replaced by a fibrous cellular stroma containing foci of mineralisation or ossification.[1] Ossifying fibromas have various clinicopathological overlaps with the other two fibro-osseous lesions: osteoma and fibrous dysplasia.[2] Thus, to make an accurate diagnosis of ossifying fibroma, a combination of histopathology, clinical presentation and radiological imaging is required.[3]

Ossifying fibromas are subcategorised histopathologically into three main types including juvenile psammomatous, juvenile trabecular and cemento-ossifying fibromas.[3,4] Despite being benign neoplasms, Ossifying fibromas have a high potential to invade local structures including the orbits, leading to various signs and symptoms depending on the compressed structures.[5–7]

The key to managing ossifying fibromas of the sinonasal tract depends on radiological features showing their location, extent and affected and/or nearby structures. Watchful waiting is may be a less favourable option as local compression of these slow-growing tumours in young adults usually results in compressive symptoms. The aim of treatment is to achieve decompression and/or complete resection. The decision to employ either an open surgical approach via transfacial, transoral or craniotomy rather than an endoscopic or a combination of open and endoscopic endonasal approach is thus dependent on whether full resection can be achieved.[8] There is no evidence that either approach is superior to the other.

Case history

A 20-year-old man presented to the ophthalmology department following a referral from his optician with a five-week history of visual disturbance. On examination, there was a marked reduction in visual acuity in the right eye (6/7.5) and colour vision was 1/17 on the Ishihara chart. There was also a right relative afferent pupillary defect and a slight right ptosis. While he was found to have right optic nerve oedema keeping with optic neuritis, it was unusual that he had not experienced any pain on eye movements. He had no relevant past medical history or drug history. Urgent magnetic resonance imaging (MRI) was requested to rule out other causes of his visual deterioration.

On the MRI of the brain and the orbits (Figures 1 and 2), there was a homogeneously enhancing expansile mass centred on the medial wall of the right orbit arising from and involving the anterior and posterior ethmoidal sinuses. The lamina papyracea was significantly displaced medially

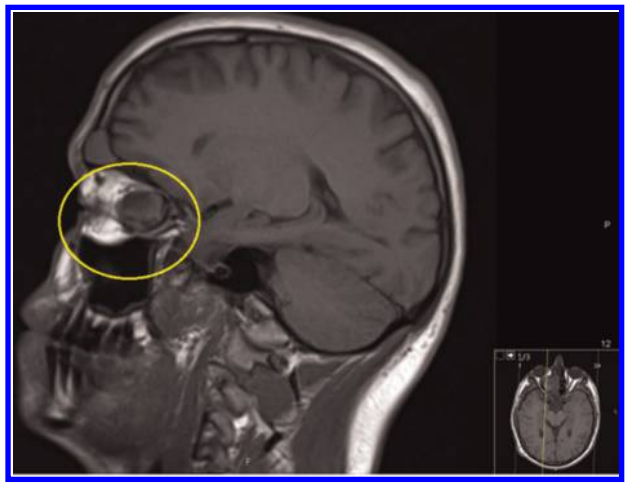

Figure 1 Preoperative sagittal magnetic resonance images.

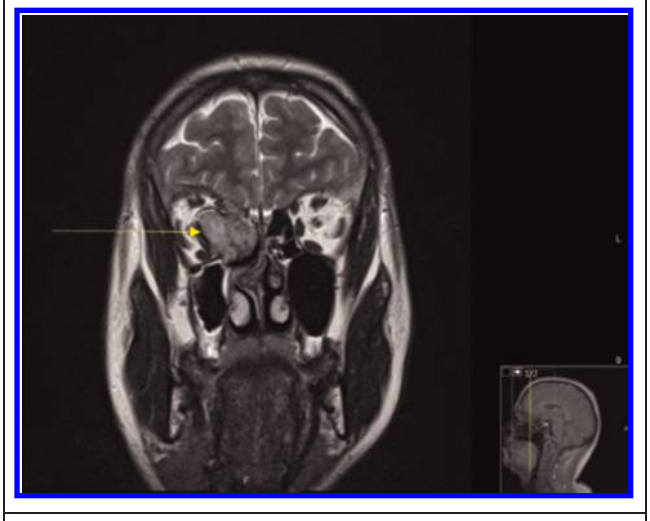

Figure 2 Preoperative coronal magnetic resonance images.

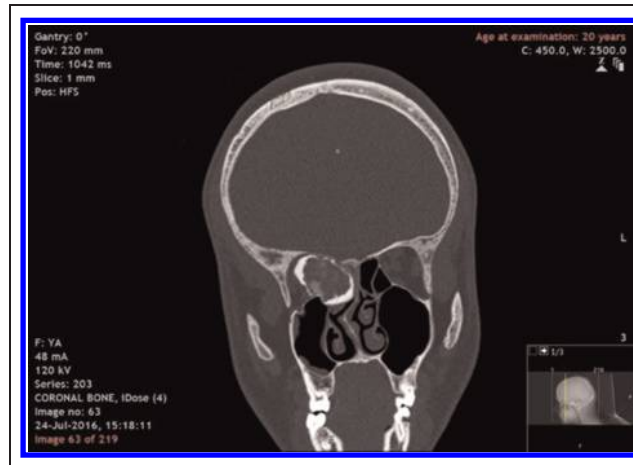

Figure 3 Preoperative coronal computed tomography.

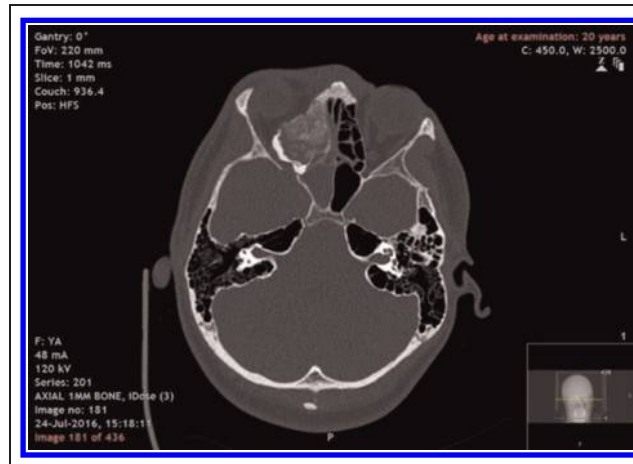

Figure 4 Preoperative axial computed tomography.

and there was mass effect on the optic nerve that was compressed and displaced to the right. There was also mass effect on the extraocular muscles. Additionally, the mass was seen to cause obstruction of the right sphenoidal and the right frontal sinuses with mucus retention. The lesion extended superiorly to the skull base.

Since it was difficult to comment on whether the mass was a primary bony tumour or a sinus mucosal tumour involving the orbit based on the MRI scans, computed tomography (CT) was indicated to characterise the bony anatomy further (Figures 3 and 4). The scans confirmed that this was an osseous lesion with a distinct bony capsule, suggesting benign pathology.

In light of the CT findings, an endoscopic image-guided tissue biopsy was taken of the lesion, which showed fragments of respiratory-type mucosa with a fibro-osseous lesion composed of hypocellular fibrovascular stroma containing curvilinear trabeculae of mineralised bone. There was also focal osteoblast rimming around the fibro-osseous lesion.

Owing to the combination of the locally aggressive nature of the ossifying fibroma, the case was discussed at the skull base multidisciplinary team and a surgical resection was deemed necessary to decompress the optic nerve. A rhinologist and an ophthalmologist undertook an image-guided combined endoscopic endonasal and external approach to ensure adequate debulking of the right orbit and relieve pressure on the optic nerve, thus avoiding unnecessary morbidity while preserving vision.

First, the endoscopic portion of the tumour was resected. A right septal spur, the anterior middle turbinate and the uncinate were resected for better access to the anterior

aspect of the sinus portion of the tumour. The hard bony shell of the tumour was removed inferiorly and medially with a drill, up to the face of the sphenoid. A sphenoidotomy was performed to drain all inspissated mucus and access the back of the tumour. The soft central portion of the tumour was resected towards the orbit and skull base leaving a small rim superiorly. Finally, a frontal sinusotomy was conducted to drain thick inspissated mucus and a T-tube frontal sinus stent was inserted.

The orbital approach was performed via a joint subcutaneous and Lynch incision. The orbital rim was identified and the periosteum incised. Exploration of the orbital roof and medial wall was then possible. The tumour was uncovered at the medial wall of the orbit. The exposure of the entire orbital roof from anterior to posterior length was required to remove all of the tumour. The resection of the tumour was performed in small fragments with image-guided control and some portions were removed endoscopically. Afterwards, a medial wall polytetrafluoroethylene barrier implant (1 mm) was inserted with EpiFilm® (Medtronic) placed against the medial orbital wall graft in the ethmoid sinus cavity. Finally, the medial canthal ligament and periorbitum were repaired with 5.0 vicryl sutures and skin closure was achieved with 6.0 nylon sutures.

All specimens removed during surgery showed the features of a benign fibro-osseous process, consisting of numerous curved, branching trabeculae comprising osteoid and woven bone showing osteoblastic rimming. The trabeculae are surrounded by highly cellular stroma containing several osteoclasts. The appearances are in keeping with the initial clinical suspicion of a juvenile ossifying fibroma.

The patient was discharged the next day with little postoperative morbidity reported. At the 10-day postoperative follow-up, he was found to have minimal residual ocular symptoms and an acceptable visual acuity with settling diplopia. The three-month postoperative CT (Fig 5) showed displacement of medial rectus and right optic nerve.

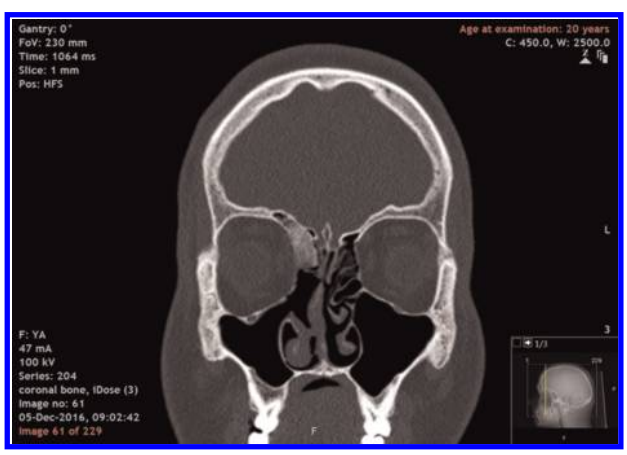

Figure 5 Postoperative coronal computed tomography at 3 months.

At the six-month postoperative follow-up, the patient reported no recurrent symptoms. On endosopic examination of the sinuses, the right frontal sinus cavity was patent and free of discharge; the frontal sinus stent (Fig 6) was subsequently removed.

At the 18-month postoperative follow-up, the right eye visual acuity had returned to normal (6/6) with no recurrent ocular or sinus symptoms. The CT (Fig 7) showed that the right medial rectus was no longer displaced and patency of the right frontal sinus. The patient was happy with a small aesthetically pleasing external scar.

Conclusion

Although there are few cases of ossifying fibroma reported in the sinonasal tract, including the ethmoid sinus and the orbit, the majority have presented with minimal to no intra-orbital involvement.[5,10–12] In one case series, most patients were reported to experience minor symptoms such as nasal obstruction or were asymptomatic.[6] In this case, visual loss as a result of optic nerve compression due

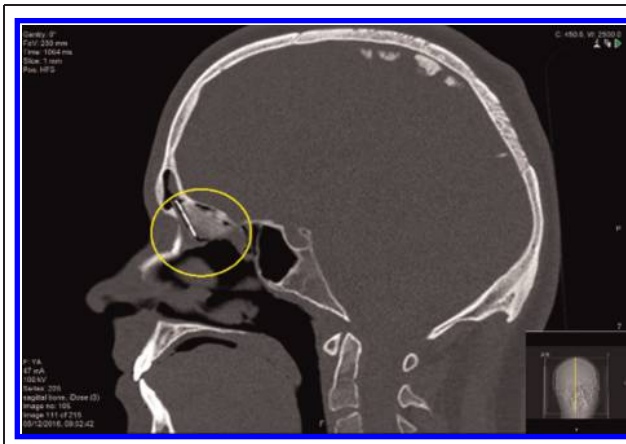

Figure 6 Postoperative sagittal computed tomography at 3 months with frontal sinus stent in situ.

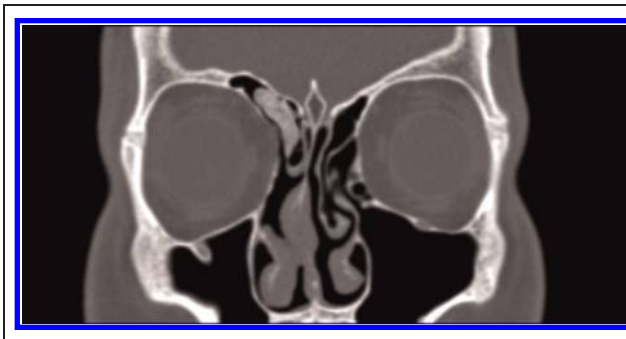

Figure 7 Postoperative computed tomography at 18 months.

to the extent of the tumour was a pivotal concern. There has been no previously published report of sight-preserving surgery to remove an ossifying fibroma in the sinonasal cavity with intra-orbital involvement.

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