Unusual presentation of more common disease/injury

Nasal chondromesenchymal hamartoma with no nasal symptoms

Vincent Uzomefuna,¹ Fergal Glynn,² John Russell,² Michael McDermott³¹Department of Otolaryngology, South Infirmary Victoria University Hospital, Cork, Ireland²Department of Otolaryngology, Our Lady's Children Hospital, Dublin, Ireland³Department of Pathology, Our Lady's Children Hospital, Dublin, Ireland

Correspondence to Vincent Uzomefuna, uzomefuna@live.ie, uzomefuna@yahoo.co.uk

Summary

The authors present a case of nasal chondromesenchymal hamartoma (NCMH) in an 8-year-old boy with a 4-month history of frontal headache and no symptoms of nasal obstruction, rhinorrhoea or postnasal drip. An ENT examination as well as ophthalmology assessment presented normal results. CT scan showed a lesion involving the sphenoid and ethmoid sinuses. The patient had an endoscopic resection of the lesion that was confirmed histologically to be a NCMH. Though NCMH is known to present usually in infants with obstructing nasal mass, an unusual presentation of a patient with throbbing headache without any nasal symptoms is reported here.

BACKGROUND

This case is an unusual presentation of an uncommon medical condition.

CASE PRESENTATION

An 8-year-old boy was referred to our service following investigations for a 4-month history of frontal headache that was intermittent and throbbing in nature. There was no history of nasal obstruction, rhinorrhoea, loss of smell or postnasal drip. However, the patient complained of intermittent pain in his right eye. The result of a general clinical examination was essentially normal and ophthalmology assessment showed no visual field defect. An ENT examination presented normal results with no obvious intranasal mass. An MRI scan was obtained and this revealed a mass in the right sphenoid sinus; therefore, a high-resolution CT of

the para-nasal sinuses was obtained. This contrast CT demonstrated a large, lobulated, soft-tissue mass involving the right posterior ethmoid sinus and the anterior two-thirds of the sphenoid on both sides (figure 1). It also extended up to the cribriform plate and abutted the anterior loop of the carotid siphon on both sides, but there was no evidence of intracranial or intra-orbital extension. The patient was listed for an image-guided endoscopic resection of the tumour.

INVESTIGATIONS

A CT scan of the brain and an MRI of the head and neck were carried out.

TREATMENT

The patient underwent an image-guided endoscopic resection of the tumour. At surgery, a fleshy mass with bony

Figure 1 T1 weighted sagittal MRI and a coronal CT images showing a large soft-tissue mass involving the posterior ethmoid sinus and the anterior two-thirds of the sphenoid sinus with significant thinning of the wall of sphenoid.

Figure 2 Section of respiratory mucosa with lobules of variably cellular hyaline cartilage and diffuse irregular calcification accompanied by a predominantly benign chondro-osseous proliferation.

elements was uncapped and the contents removed in full, exposing the back wall of the sphenoid, but leaving the lamina papyracea intact.

OUTCOME AND FOLLOW-UP

The histopathological analysis showed respiratory mucosa with pronounced chronic inflammation, with lobules of variably cellular hyaline cartilage accompanied by diffuse irregular mineralisation and calcification but no frank cytological atypia. The histological findings were in keeping with a benign chondro-osseous proliferation, consistent with a nasal chondromesenchymal hamartoma (NCMH; figure 2). The patient did well postoperatively and was discharged on the second postoperative day. On follow-up at 6 months, he remained asymptomatic and there was no recurrence of tumour.

DISCUSSION

NCMH is a benign mass lesion of the nasal cavity predominantly composed of mesenchymal and cartilaginous elements.¹ It is thought to be caused by an underlying genetic predisposition in combination with possible hormonal or other environmental stimulation.² Most documented cases have occurred in infants and neonates, and typically present as an intranasal mass usually obvious on routine ENT examination.

In its original description in a study of seven children, McDermott *et al* defined NCMH as a tumefactive process of the nasal passages and contiguous paranasal sinuses with a detectable mass in the nose.³ A Pubmed search for NCMH revealed 20 cases in the literature but no record of chondromesenchymal hamartoma of the sphenoid. Frequent symptoms include nasal obstruction, stertor, rhinorrhoea, facial asymmetry or epistaxis. Intra-ocular involvements have been reported with resulting signs of proptosis, enophthalmos, diplopia or ophthalmoplegia.^{4 5}

Our patient presented with a 4-month history of frontal headache and had none of the classical intranasal symptoms. The location of our patient's tumour in the sphenoid sinus as well as the absence of nasal symptoms are unusual and emphasise the need for a high index of suspicion and importance of timely radiological assessment if appropriate diagnosis is to be made. Radiological assessment with CT or

MRI will rarely give a conclusive picture of the nature of the mass, but should confirm its presence and its relationship and impact on surrounding structures including pressure remodelling of adjacent bones, bony destruction, orbital and intracranial extension status. In our patient, a CT scan did show the presence of a large lobulated expansile mass with extensive coarse calcifications that was causing some bowing of the posterior medial wall of the right orbit, with associated narrowing of the right optic foramen. There was a significant thinning of the wall of the sphenoid. The initial radiological impression was that of ossifying fibroma.

Complete excision is usually curative, but is not always technically possible, especially in cases with intra-cranial extension. Incomplete excision opens up the theoretical possibility of continued growth and progressive symptoms, especially in younger patients but this was not always observed in cases with incomplete excision in the original series, so a definitive therapeutic recommendation on such cases remains elusive.²

One of the most intriguing features of NCMH is the recent recognition of its association with pleuropulmonary blastoma (PPB). PPB is a primitive malignant neoplasm of the chest that typically arises in early childhood. PPB has now been recognised as a component of a syndrome of familial neoplastic disorders with many patients demonstrating other tumours at extra-thoracic sites or having close relatives with a history of tumours.⁶ Tumours that have been previously reported in patients with PPB include malignant germ-cell tumour, Sertoli-Leydig tumour, medulloblastoma, cystic nephroma, and benign and malignant thyroid lesions.⁷

The original description of NCMH included a patient with PPB and Johnson *et al* also reported another case of NCMH in association with a PPB and a testosterone-secreting left ovarian stromal tumour.^{2 6}

The Pleuropulmonary Blastoma Registry has now identified further examples of the occurrence of these rare tumours in the same patient, arguing that NCMH should be regarded as a potential marker for the PPB family tumour syndrome.⁸ Indeed, because NCMH tends to occur early in life, often in infancy, it may be the earliest warning sign of such a predisposition in the patient and in close relatives, so further investigation of the patient and relatives is warranted to exclude presence of associated tumours.

Learning points

- ▶ Nasal chondromesenchymal hamartoma (NCMH) is an uncommon but locally invasive neoplasm of mesenchymal and cartilaginous origin.
- ▶ Whereas this lesion has been defined as a tumefactive process of the nasal passages and contiguous paranasal sinuses with a detectable mass in the nose,² we report an unusual case of NCMH which, instead of the usual inferior extension to produce nasal obstruction, has grown postero-superiorly to invade the sphenoid sinus, thereby causing deep-seated headache without nasal symptoms, and leading to initial medical referral and follow-up with a 4-month delay in diagnosis. This is to highlight the importance of a high index of suspicion because certain cases of NCMH that could pose as the most significant threat to life may actually cause neither nasal obstruction nor a detectable mass in the nose.

Competing interests None.

Patient consent Obtained.

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