Case report

An extremely rare and atypical paediatric presentation of a maxillary sinus haemangioma in the UK

Aphrodite Iacovidou,1 Vikas Acharya,2 Devavrata Joshi,1 Ali Taghi3

SUMMARY
We present a rare and unusual case of a 16-year-old girl, with no significant medical history, presenting with right nasal obstruction and suspected sinusitis with occasional epistaxis and haemoptysis. On examination, she had a mass lesion in the right nasal cavity, with no evidence of other pathology on assessment of the ears, nose, throat or head and neck. A CT scan revealed an opacified right maxillary sinus with polypoidal mucosa, extending and passing through the accessory ostium into the right nasal cavity. Examination under anaesthesia with functional endoscopic sinus surgery and excision of the lesion was subsequently undertaken. Histological analysis confirmed the mass lesion as a haemangioma. This case report is the first to present a maxillary haemangioma presenting as nasal obstruction with intermittent sinusitis symptoms in a child. The authors discuss the incidence, presentation and management of maxillary haemangiomas in the paediatric population.

BACKGROUND
Haemangiomas are benign, vascular neoplasms which can arise from most tissue cells in the human body. They very rarely arise from the paranasal sinuses. Vascular tumours/malformations are uncommon findings in the nasal cavity, and if are found, are very unlikely to be haemangiomas.1 Evidence suggests that haemangiomas of the paranasal sinus, specifically the maxillary sinus, are almost unheard of, due to them being so rare in presentation.2

Due to their sporadic nature, lesions of the maxillary sinus are rarely considered to be haemangiomas. This can be detrimental to the patients’ care pathway, due to the risk of excessive bleeding on excision or tissue sampling of this highly vascular tumour.3 They are often difficult to differentiate from other more common lesions or tumours on clinical examination, cavernous haemangiomas can often grow laterally, eroding adjacent structures and bones and thus even causing difficulty in differentiating from other tumours radiologically.4–7 Typical features of these on CT and MRI are expanse, well-enhanced heterogeneous masses, associated with bone remodelling, erosive changes and internal calcification due to haemorrhage.8

This paper reports the unusual case of a 16-year-old girl who presented with a right maxillary haemangioma, a rare presentation in itself, but the first in the literature of a paediatric case in the UK with this case presentation.

CASE PRESENTATION
This otherwise fit and well, 16-year-old girl was referred to the paediatric ear, nose and throat (ENT) team by her general practitioner because of ongoing right nasal obstruction, with associated suspected recurrent right-sided sinusitis. She complained of right frontal headaches with fullness, postnasal drip, mucous and discharge from the right nostril which was often foul smelling, hyposmia and intermittent right-sided epistaxis. These symptoms had been persisting for just under 1 year prior to being seen in the ENT clinic. This has been managed by her general practitioner with multiple courses of antibiotics and a nasal spray, with no improvement in her symptoms.

There was no relevant antenatal, birth or childhood, including medical history to note. She is on no regular medications and is up to date with her immunisations. There is no history suggestive of atopy, asthma, eczema or food intolerances or sensitivity.

On initial assessment in clinic, she had left nasal rhinitis and the right nasal vestibule was full of mucous and yellow discharge. She was started on chronic rhinosinusitis treatments including nasal douching, antibiotics and a steroid-based nasal spray. A non-contrast CT scan of the sinuses was requested (figures 3 and 4) as opposed to an MRI modality as a mass lesion was not visualised or suspected. At her next review, the symptoms other than nasal obstruction and blockage, had slightly improved. Flexible nasoendoscopy revealed a large polyp arising from the right maxillary sinus and occupying a significant proportion of the nasal cavity. She still had ongoing evidence of rhinitis. The scan revealed a polypoidal mass extending from the right maxillary sinus and occupying most of the right nasal cavity, suspected to be an antrochoanal polyp.

After explaining the clinical and radiological findings, she was listed for functional endoscopic sinus surgery. This included an examination under anaesthesia, tissue sampling for histopathology and excision.

INVESTIGATIONS
Preoperatively planning was already completed with the non-contrast CT scan, allowing for delineation...
of relevant anatomy (figures 1 and 2). As it was initially thought to be a benign polyp or antrochoanal polyp, no further imaging was undertaken prior to surgery. As a haemangioma was not suspected, and on clinical examination a lesion was not initially visualised, an MRI scan was not undertaken; however, one should acknowledge its benefit in order to assist with differential diagnoses and consider embolisation if relevant.

Endoscopic examination of the sinuses revealed a polypoidal mass arising from the maxillary antrum. Functional endoscopic surgery was performed with antegrade uncinectomy, and middle meatal antrostomy. During biopsy of the lesion, it was noted that this was very vascular and bleeding. Preoperative embolisation may have been able to assist in reducing bleeding, had an MRI scan been undertaken and a haemangioma been considered, this may have assisted with the definitive surgery. Complete excision of the lesion was attempted endoscopically but the origin of the lesion from the medial wall of the maxillary sinus was not feasible to be removed. Partial excision for tissue sampling was undertaken, as per the initial surgical plan with the patient. An intraoperative frozen section specimen is an alternative method for histological analysis, but not the most common practiced for sinus surgery in the UK.

Histopathological analysis of the lesion revealed oedematous mucosa lining with respiratory epithelium showing focal ulceration with underlying granulation tissue and squamous metaplasia. The stroma shows multiple lobules of dilated thin walled blood vessels of variable sizes amidst coagulative necrosis (figure 3). The blood vessel walls are surrounded by hyalinised stroma and are lined with a single layer of bland epithelium and show intraluminal red blood cells (figure 4). There is no atypia, tumour necrosis or invasive malignancy.

Haemangiomas are divided into capillary and cavernous types depending on the dominant vessel size seen on microscopic examination. Capillary haemangiomas are the most common and are layered by capillary sized vessels surrounded by flattened epithelium. Cavernous haemangiomas are composed of large, endothelium-lined vascular spaces.8 Our patient had a capillary haemangioma which is the most common type of paediatric haemangioma in the head and neck region.

**DIFFERENTIAL DIAGNOSIS**

Following initial clinical examination and investigations, the main differential diagnoses were as thought to be; inverted papilloma, antrochoanal polyp, benign inflammatory polyp, organised haematoma or juvenile angiofibroma.

Organised haematoma usually damages the medial maxilla and ethmoid cavity, and angiofibroma usually affects the pterygopalatine fossa extending to the nasal cavity, neopharynx and sphenoid. Fungal balls usually show calcification on an unenhanced CT scan and sinonasal malignancy usually exhibits invasive and
destructive features on CT and MRI which differentiate them from haemangioma.9

It is not until intraoperatively, when the endoscopic examination was undertaken did a benign inflammatory polyp and angiofibroma become less likely. It was still unclear as to the diagnosis and thus tissue sampling was taken prior to excision. The nature of the ongoing bleeding from the tissue did make an atypical diagnosis more likely, but without histopathology one can never truly differentiate and be certain. The differential diagnosis in a key milestone in the care pathway for a patient with a unilateral nasal or sinus mass, and should be taken seriously and managed in an urgent manner.

**TREATMENT**

Treatment options for this condition vary depending on the size of the haemangioma and clinical presentation, clinical assessment and investigations.

Initially as in the case of our patient, unilateral nasal obstruction, with symptoms of sinusitis, both acute and chronic, would be managed according to the approved guidelines for this condition. Treatment with nasal douching, decongestants, steroid-based nasal sprays are the most common first line treatments. In acute sinusitis presentations, the treatment additionally includes antibiotics and analgesia. Supportive treatment, such as being in a humidified room, and steam inhalation may also provide supportive, symptomatic relief to some.

Epistaxis is usually managed conservatively by giving advice on correct application of local pressure to the nose, using ice packs to promote vasoconstriction to the nose and facial area and by leaning forward. There is then a stepwise approach to managing epistaxis depending on its clinical manifestation and the patients history. Assessment of the nose, benchmarked against the frequency, duration of nose bleeds and blood volume are considered when planning further management.

Treatment of the maxillary haemangioma itself is determined by the patient symptoms. If there is ongoing nasal obstruction, facial fullness or pain which is not being managed medically, or epistaxis which is not feasible to be treated in the outpatient clinic setting, then surgery may be considered. However, Kim et al10 have shown there is a linear regression between symptom duration and size of haemangioma. A delay in diagnosis even by 1 month can increase the size of the tumour by 0.38 mm. Therefore, if there is suspicion of haemangioma or any sinister pathology, early surgery is advisable.

**OUTCOME AND FOLLOW-UP**

Following the hospital admission for urgent examination under anaesthetic with tissue sampling, the patient stayed in hospital for one night for observation and was subsequently discharged home with nasal drops and sprays.

She had been seen 2 weeks postoperatively, in the outpatient clinic, and was symptomatically much improved. The histology results were explained to the family and plans for further management and investigations agreed. This will include an MRI scan, further review in the clinic, and consideration of further surgery. The patient remains stable, well, and has reduction in all her symptoms since the operation.

**DISCUSSION**

Sinonasal haemangiomas are rare entities and there have been no case reports of maxillary haemangiomas in the paediatric population.

Surgical resection is the main treatment in the literature, based on tumour extent. Techniques include lateral rhinotomy, Caldwell Luc operations or endoscopic sinus surgery. In the largest series of 37 patients with sinonasal haemangioma by Kim et al,10 all were managed with endoscopic surgery, while the most common site of presentation was nasal septum (40.5%), inferior turbinate (29.7%), maxillary sinus (8.1%) and uncinate process (8%). In a similar large case series with 31 patients, Takaishi et al11 identified only two patients who required preoperative arterial embolisation, while the remaining patients had endoscopic excision under local or general anaesthesia. The reasons behind this decision were due to the size of lesion being bigger than 20 mm and an inability to identify the origin on CT or MRI. Both had preoperative embolisation, followed by surgery the following day.

In these two studies, with 68 patients, there were only three recurrences of haemangioma. In Kim et al, both patients had cavernous haemangioma and no recurrence was observed in those with capillary haemangioma. Recurrence happened at 4 months and 60 months postsurgery for two patients. Takaishi et al had one patient with recurrence who had a local excision during her last trimester of pregnancy due to repeated bleeding episodes. This recurred at week 6 postsurgery.

Even though sinonasal haemangiomas are rare entities, they should be considered as a differential in unilateral nasal lesions with epistaxis. These should be managed with appropriate investigations and early planning to prevent progression and potential intraoperative complications with excessive bleeding. In paediatric patients, this condition should be managed in a specialist centre where interventional radiology and paediatric intensive care is available.

**Patient’s perspective**

I had been suffering from a right-sided blocked nose and nosebleeds for a long time, and it was starting to affect me at school, home and when I was out with my friends. I would almost always have a headache and pressure in my face and knew something was going on. When I was shown the mass in my sinus on the scan I was very upset and anxious, but having a quick appointment date for surgery made me feel better.

I was relieved finally to have an answer, and know what had been causing these problems with me for so long. I am worried and so is my mum, about it coming back or growing again. I will see my doctors in the hospital regularly to make sure things are ok.

**Learning points**

► Patients with nasal obstruction or atypical sinusitis type symptoms should be referred to ENT.
► Unilateral nasal masses or polyps need urgent investigation with clinical imaging in the form of CT and MRI to delineate relevant anatomy and pathology, prior to considering surgery.
► Tissue sampling in theatre by surgical excision is required to confirm the diagnosis.
► Despite being rare, if epistaxis or haemoptysis is a symptom, haemangiomas should be considered as a differential and preoperative arterial embolisation should considered.

**Acknowledgements** The authors would like to thank all staff members involved in the care pathway of the child and their colleagues in the departments of pathology and radiology for their assistance with the images.

Iacovidou A, et al. BMJ Case Rep 2019;12:e230696. doi:10.1136/bcr-2019-230696
Contributors All authors were involved in the care of the patient, contributed to the creation and completion and proofreading of the manuscript and are happy for submission. VA and AI were involved in writing the case report and proofreading, undertaking and streamlining the literature review and preparing the manuscript for submission including making changes for the revised manuscript. DJ and AT were both involved in the primary operation for the patient, were involved in assisting with the writing of the manuscript and proofreading, in addition to assisting with images and figures and their descriptions for the manuscript. They also reviewed the revised manuscript.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Open access This is an open access article distributed in accordance with the Creative Commons Attribution Non Commercial (CC BY-NC 4.0) license, which permits others to distribute, remix, adapt, build upon this work non-commercially, and license their derivative works on different terms, provided the original work is properly cited and the use is non-commercial. See: http://creativecommons.org/licenses/by-nc/4.0/

REFERENCES
1 Batsakis JG. Tumors of the head and neck: clinical and pathological considerations. Baltimore: Williams & Wilkins, 1979:291–312.
2 Ys F, Perzin KH. Nonepithelial tumours of the nasal cavity, paranasal sinuses and nasopharynx: a clinicopathologic study. Cancer 1974;33:1275–88.
3 Engels T, Schomer W, Felix R, et al. Kavernoses Hamangiom des Sinus maxillaris: HNO, 1990:136–42.
4 Hellquist HB. Pathology of the nose and paranasal sinuses. London: Butterworth and Co. Ltd, 1990:136–42.
5 Raboso E, Rosell A, Plaza G, et al. Haemangioma of the maxillary sinus. J Laryngol Otol 1997;111:638–40.
6 Kim HJ, Kim JH, Hwang E.G. Bone erosion caused by sinonasal cavernous haemangioma: CT findings in two patients. Am J Neuroradiol 1995;16:1176–8.
7 Jammal H, Barakat F, Hadi U. Maxillary sinus cavernous hemangioma: a rare entity. Acta Otolaryngol 2004;124:331–3.
8 Jung WS, Yoo CY, Park YS, et al. Hemangioma of the maxillary sinus presenting as a mass: CT and MR Features. Iran J Radiol 2015;12:e6923.
9 Seo YJ, Kim J, Kim K, Kim J, et al. Radiologic characteristics of sinonasal fungus ball: an analysis of 119 cases. Acta Radiol 2011;52:790–5.
10 Kim JS, Kwon SH. Sinonasal hemangioma: diagnosis, treatment, and follow-up of 37 patients at a single center. J Oral Maxillofac Surg 2017;75:1775–83.
11 Takaishi S, Asaka D, Nakayama T, et al. Features of sinonasal hemangioma: A retrospective study of 31 cases. Auris Nasus Larynx 2017;44:719–23.