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CLINICAL RECORD

Bilateral, independent juvenile nasopharyngeal angiofibroma: case report

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Abstract

Background: Juvenile nasopharyngeal angiofibroma is a benign, vascular tumour that primarily occurs in adolescent males. Despite its benign nature, aggressive growth patterns can cause potential life-threatening complications. Juvenile nasopharyngeal angiofibroma is normally unilateral, originating from the sphenopalatine artery, but bilateral symptoms can occur if a large tumour extends to the contralateral side of the nasopharynx. This paper presents the first reported case of true bilateral extensive juvenile nasopharyngeal angiofibroma involving clinically challenging pre-surgical planning and surgical strategy.

Case report: A 21-year-old male presented with increasing bilateral nasal obstruction and discharge. Examination revealed tumours bilaterally and imaging demonstrated non-contiguous tumours. Pre-operative angiography showed strictly ipsilateral vascular supplies requiring bilateral embolisation. Radical removal performed as one-step, computer-assisted functional endoscopic sinus surgery was performed. The follow-up period was uncomplicated.

Conclusion: This case illustrates the importance of suspecting bilateral juvenile nasopharyngeal angiofibroma in patients presenting with bilateral symptoms. Our management, including successful pre-operative planning, enabled one-step total removal of both tumours and rapid patient recovery.

Key words: Bilateral Nasal Obstruction; Nasopharynx; Nasopharyngeal Neoplasms; Angiofibroma; Head And Neck Neoplasms; Therapeutic Embolization

Introduction

Juvenile nasopharyngeal angiofibroma is a benign, highly vascular neoplasm.[1] The lesion is described as primarily occurring in young males aged 10–24 years,[2] with initial unilateral nasal obstruction, recurrent recalcitrant epistaxis and nasal discharge symptoms. The reported incidence of juvenile nasopharyngeal angiofibroma is 0.4 per million inhabitants per year.[2]

The tumour typically originates from the superior margin of the sphenopalatine foramen and spreads into the submucosal plane.[1] Despite its benign nature, juvenile nasopharyngeal angiofibroma shows an aggressive growth pattern, especially among the youngest patients, causing the potentially life-threatening complications of haemorrhage and intracranial involvement.[3] Thus, it should be treated as radically as possible.

Here we present a unique case of bilateral juvenile nasopharyngeal angiofibroma. (Written informed consent was obtained from the patient for publication of this case report and any accompanying images.) It is the first reported case with clinical challenges and, to our knowledge, is the most extensive case of bilateral juvenile nasopharyngeal angiofibroma described in the English literature,[4] and only the third case reported in the last 10 years.[5]

Case report

A 21-year-old male presented with a 6-month history of gradually increasing right-sided nasal stenosis and a 2-month history of increasing left-sided nasal stenosis that was not relieved by nasal decongestant spray. The patient experienced frequent bloody nasal secretions, but only one episode of epistaxis. He had no history of weight loss, fatigue or night sweats.

On physical examination, a closed rhinolalia was noted, and anterior rhinoscopy revealed complete obstruction on the left side by a mass. Nasal fibre-optic endoscopy revealed an obstructive reddish tumour on the right side occluding the nasal cavity from the mid-portion of the middle turbinate. The left-sided tumour was impassable.

The left-sided tumour was biopsied under local anaesthesia as juvenile nasopharyngeal angiofibroma was not initially clinically suspected. The biopsy showed polypoid tissue with central necrosis. Based on this unspecific histology, a freeze-drying biopsy was performed under local anaesthesia; the results indicated the diagnosis of juvenile nasopharyngeal angiofibroma. In both cases, temporary bleeding was managed by nasal packing. No biopsy was taken on the right side because of the suspicion of juvenile nasopharyngeal angiofibroma and risk of subsequent bleeding.

Computed tomography (CT) and magnetic resonance imaging (MRI) scans of the nose and sinuses (Figures 1 and 2) demonstrated a $5 \times 3 \times 2$ cm tumour on the left side and a $4 \times 4 \times 3$ cm tumour on the right side, corresponding to Radkowski *et al*. stages Ia and IIIa respectively (Table I).[6] On CT and MRI, the right-sided tumour was in the proximity of the cavernous sinus, and enlargement of

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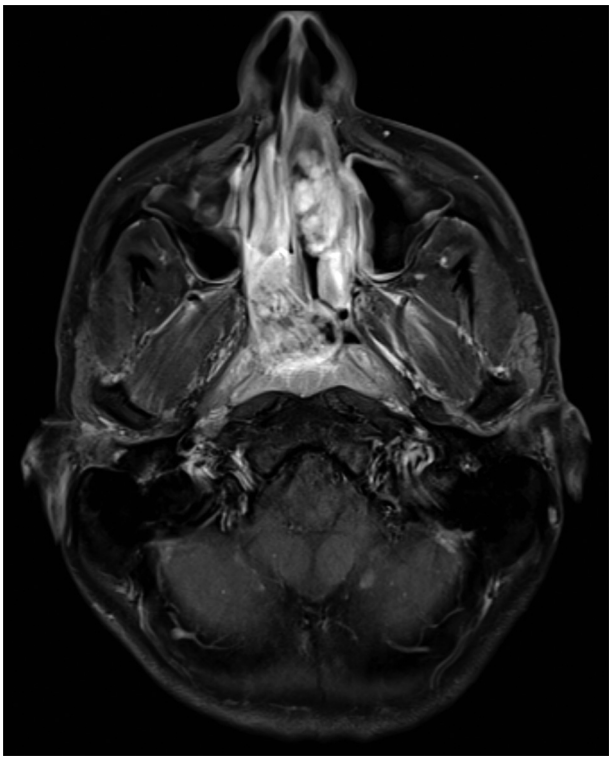


FIG. 1

Pre-operative axial, contrast-enhanced, T1-weighted magnetic resonance imaging scan showing the two independent tumours, sized 4 × 4 × 3 cm on the right side and 5 × 3 × 2 cm on the left side.

the sphenopalatine foramen was pronounced. There were no signs of nasal septum perforation. All laboratory test results were within normal levels.

Pre-operative diagnostic digital subtraction angiography was performed bilaterally in the common carotid artery, internal carotid arteries and the external carotid artery to determine the vascular supply of the tumours and to embolise them. Both tumours were strictly ipsilaterally supplied by their respective sphenopalatine arteries (Figures 3 and 4). The right-sided tumour had a small additional supply from the ipsilateral Vidian artery, which branches from the internal carotid artery. Embolisation with 150–355 μ polyvinyl alcohol particles was performed bilaterally without complications.

The following day, both tumours were successfully excised using CT and MRI guided functional endoscopic sinus surgery. The tumours from both sides were delivered transorally. The supplying arteries were electro-coagulated; the total peri-operative blood loss was 400 ml.

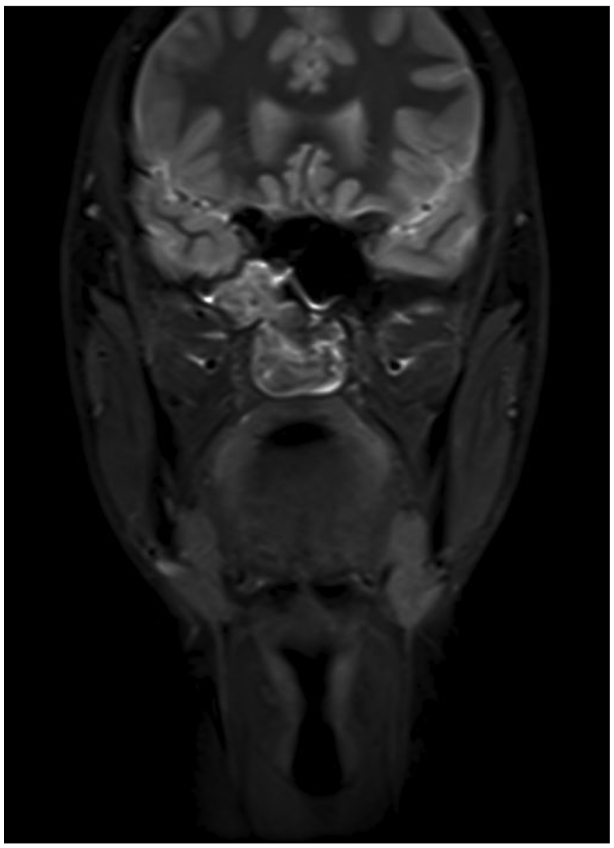
The post-operative course was uncomplicated. The patient was instructed to perform daily nasal irrigations and was discharged on the first post-operative day.

Upon follow-up examinations at zero, three, five, six and nine months post-operatively, the patient had no complaints, and there were no signs of non-radical surgery or recurrence upon nasal fibre-optic endoscopy. The absence of recurrence was confirmed by MRI nine months post-operatively.

Discussion

Juvenile nasopharyngeal angiofibroma is a rare tumour, with a reported incidence of only four to six new cases in Denmark per year. Juvenile nasopharyngeal angiofibroma may be misdiagnosed initially, especially in cases of bilateral

(a)



(b)

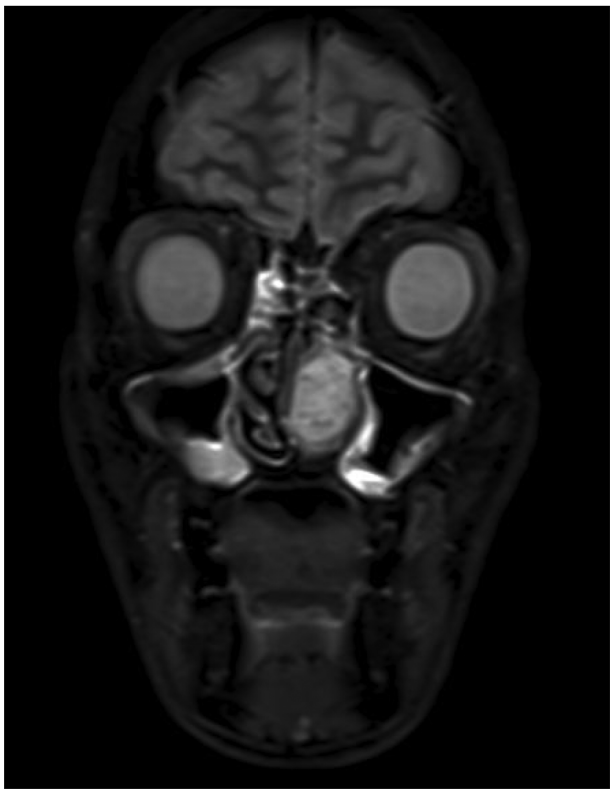


FIG. 2

Pre-operative coronal, turbo inversion recovery magnitude magnetic resonance imaging scans showing: (a) angiofibroma in the right nasal cavity, extending through the right sphenopalatine foramen and fossa to the right infratemporal fossa; and (b) angiofibroma in the left nasal cavity.

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TABLE I
RADKOWSKI AND COLLEAGUES' STAGING SYSTEM[6]

Stage	Description
Ia	Involvement limited to nose &/or nasopharynx
Ib	Extension into 1+ sinuses
IIa	Minimal extension into pterygopalatine fossa
IIb	Occupation of entire pterygopalatine fossa with or without erosion of orbital apex
IIc	Involvement of infratemporal fossa with or without extension to cheek or posterior to pterygoid plates
IIIa	Erosion of skull base (middle cranial fossa or pterygoid base); minimal intracranial extension
IIIb	Erosion of skull base; extensive intracranial extension with or without cavernous sinus invasion

juvenile nasopharyngeal angiofibroma; the current case is only the third reported case of bilateral juvenile nasopharyngeal angiofibroma in the literature.[4,5]

Juvenile nasopharyngeal angiofibromas are composed of a rich, irregular, vascular network, completely devoid of any muscular layer.[7] Early suspicion of juvenile nasopharyngeal angiofibroma is important, as biopsying in juvenile nasopharyngeal angiofibroma cases can cause massive bleeding.

The typical clinical presentation of juvenile nasopharyngeal angiofibroma is the triad of unilateral nasal obstruction, recurrent epistaxis and nasal discharge, primarily occurring in adolescent males,[1] but recognition of the extremely rare case of bilateral juvenile nasopharyngeal angiofibroma based on symptoms is difficult. The bilateral obstruction, as observed in the current case, can be mimicked by a unilateral tumour extending to the contralateral side of the nasopharynx. The possibility of bilateral juvenile nasopharyngeal angiofibroma must, however, be considered. Thorough clinical and radiological investigation is necessary to optimise pre-surgical planning, minimising the risk of massive peri-operative bleeding and ensuring radical removal of the tumours.

In the present case, the left-sided tumour was biopsied, as juvenile nasopharyngeal angiofibroma was not suspected initially, and this caused some bleeding. The biopsy did not reveal the typical pattern of juvenile nasopharyngeal angiofibroma. Bremer *et al*. reported that a superficial biopsy of a juvenile nasopharyngeal angiofibroma tumour mass might be misleading and that the classical histological appearance of juvenile nasopharyngeal angiofibroma is generally found in the deeper aspects of the tumour.[8] In the present case, the second biopsy and the arteriography revealed the diagnosis of bilateral juvenile nasopharyngeal angiofibroma.

Magnetic resonance imaging was found to be superior to CT in detecting the soft tissue extensions of the juvenile nasopharyngeal angiofibroma.[9] Computed tomography was used to display potential bone changes, including the typical enlargement of the sphenopalatine foramen in juvenile nasopharyngeal angiofibroma cases.[10] In this particular case, it was not possible to compare an affected foramen to an unaffected foramen.

Pre-operative identification of the total blood supply is essential in planning the surgical approach of these highly vascular tumours. This is achieved by angiography, which also enables pre-operative embolisation. The blood supply typically arises from the internal maxillary artery. With the continuing growth of a juvenile nasopharyngeal angiofibroma, an additional blood supply is also induced from the internal carotid system,[2] as was the case for the

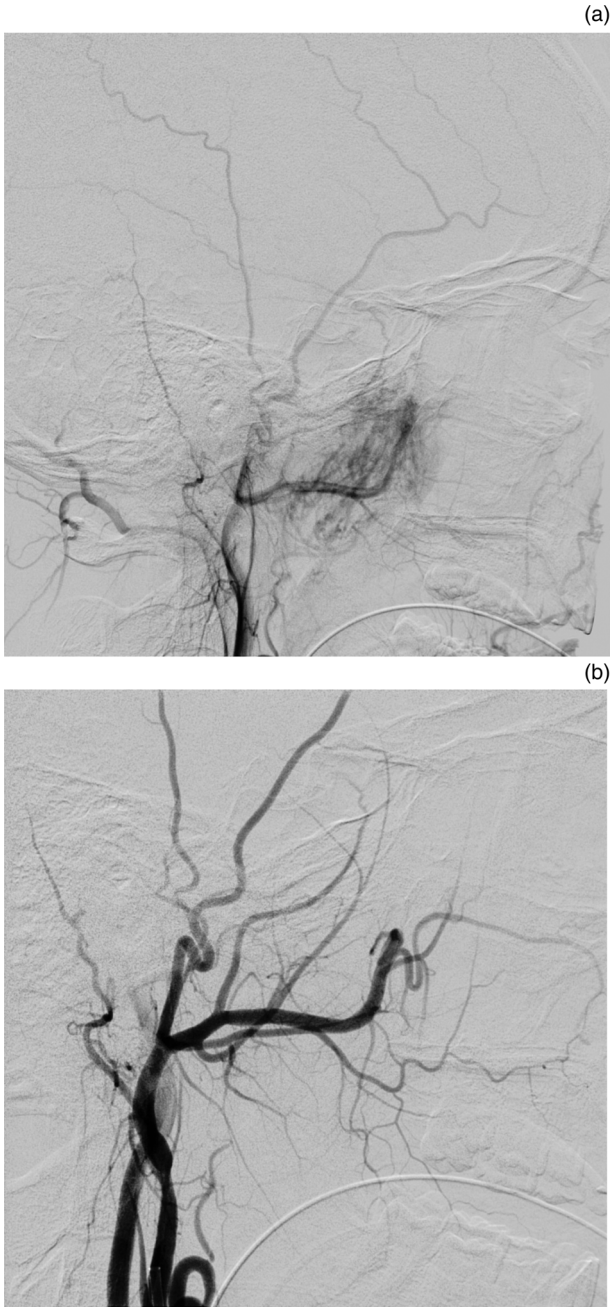

FIG. 3
Digital subtraction angiography pre-embolisation (a) and post-embolisation (b) images of the right external carotid artery. (a) Shows tumour blush from the sphenopalatine artery via the internal maxillary artery; (b) shows no visible tumour blush.

right-sided juvenile nasopharyngeal angiofibroma in our patient.

Pre-operative embolisation has been shown to reduce peri-operative blood loss and the need for blood transfusions,[11] and is now considered standard treatment. This rare case of bilateral juvenile nasopharyngeal angiofibroma emphasises the importance of bilateral embolisation, which reduces the risk of heavy bleeding from two possible sources upon excision. Our pre-operative intervention, with embolisation on both sides conducted in one procedure, was successful. It allowed for uncomplicated, one-step radical removal of both juvenile nasopharyngeal angiofibromas, with minimal blood loss and no subsequent necrosis of the nasal mucosa.

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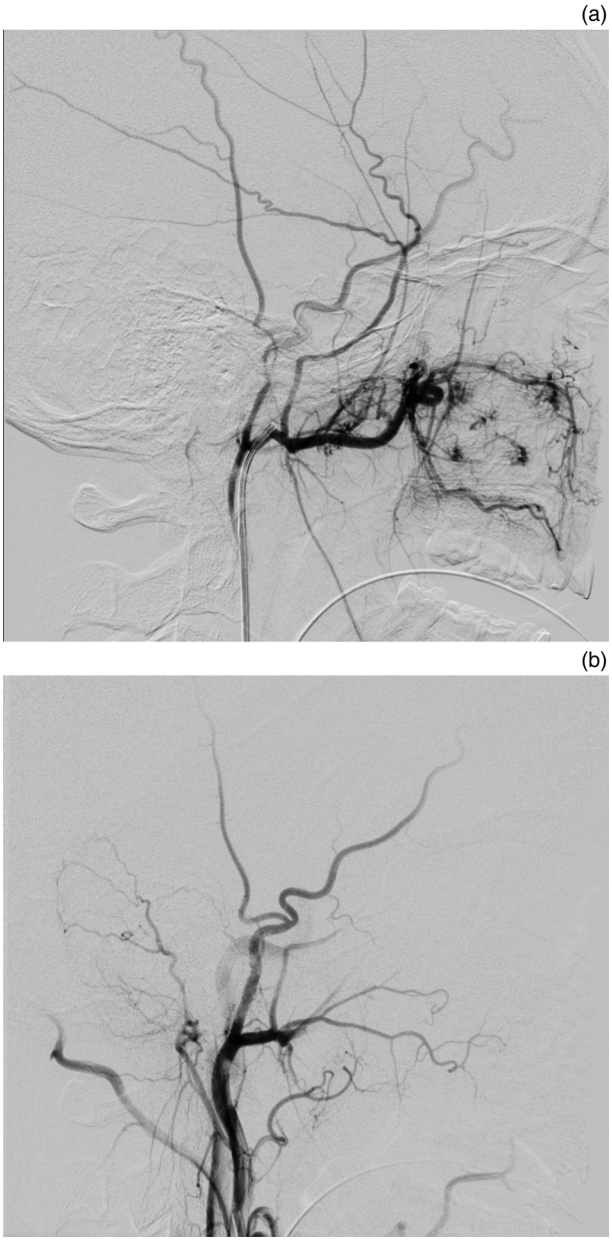

FIG. 4

Digital subtraction angiography pre-embolisation (a) and post-embolisation (b) images of the left external carotid artery. (a) Shows nodular tumour blush from the sphenopalatine artery via the internal maxillary artery; (b) shows no visible tumour blush.

Surgical excision is the 'gold standard' treatment. During the last two decades, endoscopic excision has been shown to be less disfiguring and to have lower recurrence rates than open surgical approaches.[12]

- **This is the first case of clinically challenging bilateral juvenile nasopharyngeal angiofibroma**
- **The complications are potentially life-threatening**
- **Both tumours were pre-operatively embolised**
- **One-step bilateral resection was performed; blood loss was minimal and recovery was rapid**

The overall recurrence rate of juvenile nasopharyngeal angiofibroma is reported to be about 23 per cent.[2] However, endoscopic excision might reduce the recurrence rate to about 10 per cent.[12] This might be a result of technological development within embolisation, imaging and surgery. There are no records of malignant transformation of juvenile nasopharyngeal angiofibromas.

Conclusion

In this report, we describe the first case of clinically challenging bilateral juvenile nasopharyngeal angiofibroma, which warranted thorough pre-operative planning, successful bilateral embolisation and total resection of both juvenile nasopharyngeal angiofibromas. The patient was discharged the day after surgery.

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