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Juvenile Angiofibroma Protruding from the Nasal Cavity

ABSTRACT

Objective: To describe a case of juvenile angiofibroma with unusual protrusion out of the nasal cavity, and its management with surgery and radiotherapy.

Methods:

Design: Case Report

Setting: Tertiary Public Referral Centre

Patient: One

Results: A 17-year-old gentleman presented with a huge tumor protruding from his left nostril, diagnosed with juvenile angiofibroma stage IIIA by MRI and angiography. Following successful pre-operative embolization, the protruding mass was ligated and truncated, followed by surgical resection via external approach. Post-operative residual tumor was treated with adjuvant radiotherapy. There was no evidence of recurrence after nine months.

Conclusion: A high index of suspicion is of paramount importance in the diagnosis of JA and avoids the possibility of an unwarranted biopsy which could spell disaster. The most useful tools for diagnosis are MRI and arterial angiography. Treatment is primarily surgical. Irradiation therapy has been reported to achieve satisfactory outcomes, especially for unresectable residual disease and/or intracranial extension, where total surgical resection is unlikely to be attained without unacceptable morbidity.

Keywords: Juvenile Angiofibroma, Juvenile Nasopharyngeal Angiofibroma, head and neck tumor

Juvenile Angiofibroma (JA) is a benign, highly vascular neoplasm affecting primarily male adolescents¹ and accounts for 0.5% of all head and neck tumors.² It often originates from the superior margin of the sphenopalatine foramen.³ The tumor may extend backwards into the nasal cavity and nasopharynx, laterally to the pterygopalatine fossa, involving the paranasal sinuses and infratemporal fossa. It may even invade the skull base and extend to cavernous sinus and pituitary fossa.⁴ The common clinical features include recurrent epistaxis and persistent nasal obstruction.⁵ We report a case of JA with unusual protrusion out of the nasal cavity.

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CASE REPORT

A 17-year-old Sabahan man presented with a one-year history of left nasal obstruction with recurrent epistaxis. Examination revealed a brownish well defined mass measuring 6x5 cm protruding from his left nostril (*Figure 1*). The rest of the physical examination was non-contributory.



Figure 1. Tumor protruding from the left nostril.

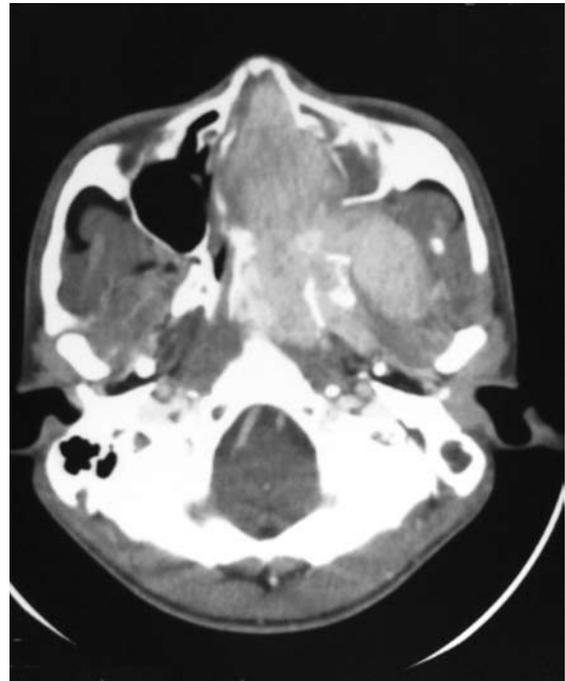


Figure 2. Contrast CT scan (axial view) revealing the extent of the disease.

Contrast-enhanced computed tomography (CT) scan showed a heterogeneous mass occupying the left pterygopalatine fossa with anterior extension through the left nasal cavity protruding through the nose, lateral extension to the left infratemporal fossa, superior extension to the left ethmoidal and sphenoidal sinuses as well as the posterior part of the left orbit. (*Figure 2*) Magnetic resonance imaging (MRI) further characterized the tumor as an enhancing mass with small intracranial extension to left middle cranial fossa and dural involvement. (*Figure 3*) The radiographic features were suggestive of JA.

Angiography of both internal (ICA) and external carotid artery (ECA) revealed the tumor supplied by branches of both ICA and maxillary arteries from both ECA. Seventy percent (70%) of the tumor-feeding vessels were successfully embolized. *Figure 4A and B* shows the arteriogram before and after embolization.

The definitive surgical procedure was performed the next day. The external nasal mass was first ligated and truncated from the nose. The internal portion of the tumor was resected via a Weber Ferguson

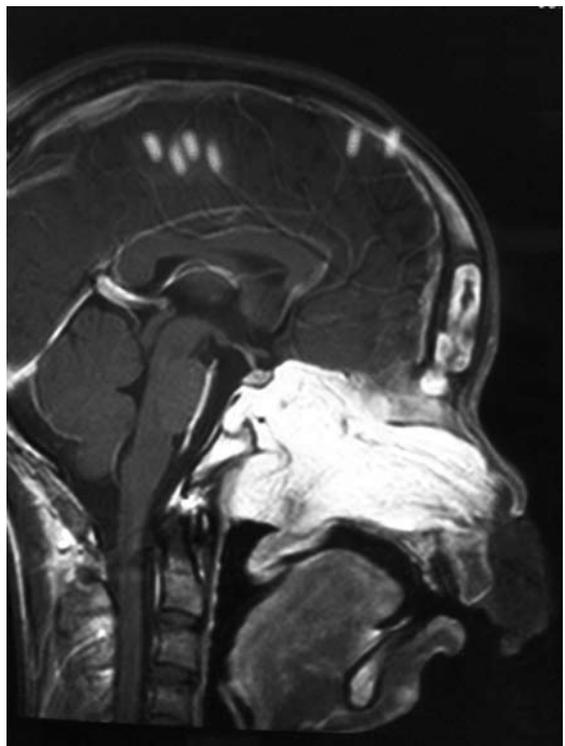


Figure 3. MRI scan characterizing the tumor and showing intracranial involvement

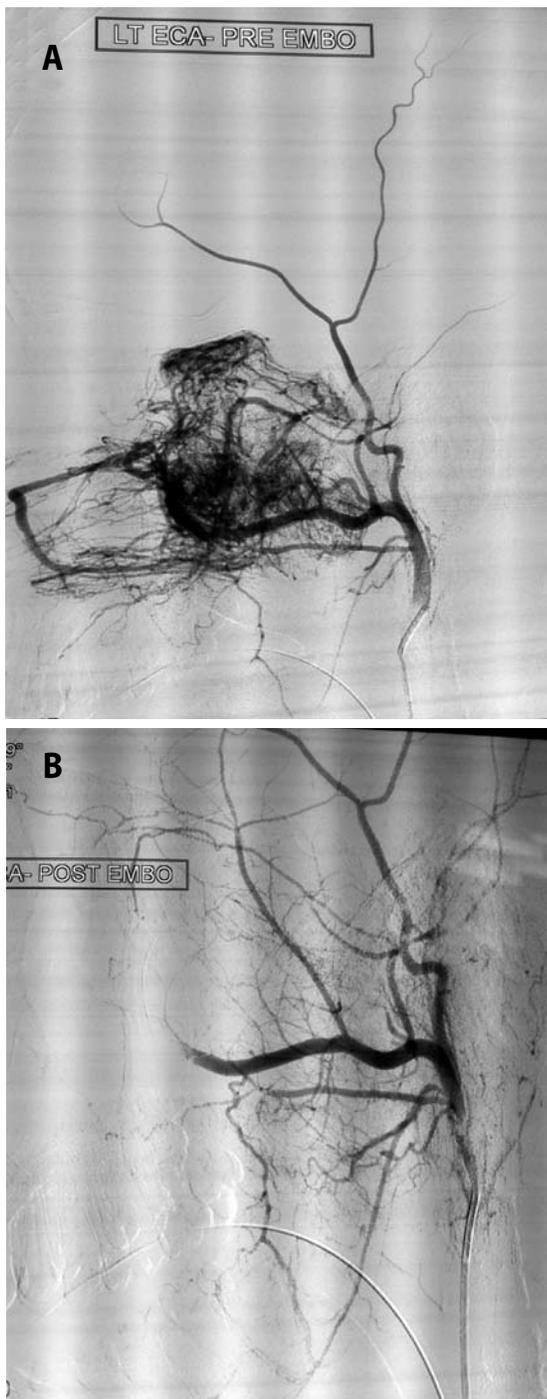


Figure 4. Angiograms- A, Pre-embolization; B, Post-embolization.

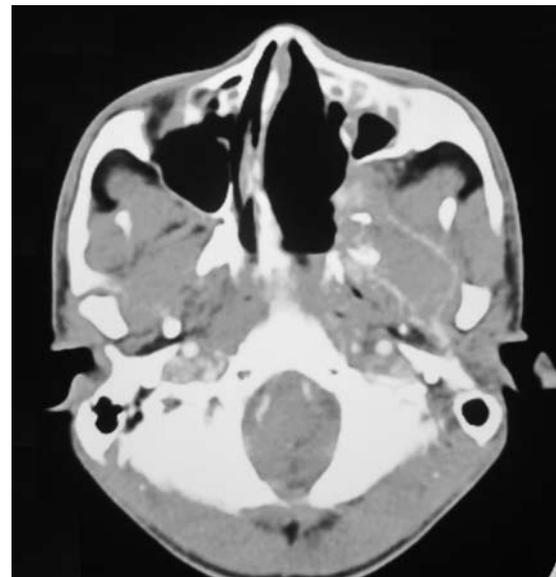


Figure 5. Post-radiotherapy CT scan (axial view) revealing no evidence of recurrence.

received 50Gy radiotherapy given in 25 fractions over five weeks which eradicated the residual lesion. At nine months post-irradiation, nasal endoscopy and CT scan revealed no recurrence. (Figure 5)

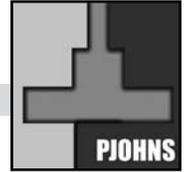
DISCUSSION

The diagnosis of JA was almost certain in view of the patient's gender, age and clinical presentation. A tumor protruding from the nasal cavity can be very tempting for a tissue biopsy, but doing so in an unprepared circumstance could spell disaster. The diagnosis could have been easily missed were it not for the initial evaluation with imaging studies because JA does not usually present as a mass protruding from the nose. Angiography further established the diagnosis and allowed embolization to be done in the same setting,

The Fisch⁶ classification (Table 1) has been commonly used for staging the disease. There are three other commonly employed staging systems (Table 2).⁷⁻⁹ None of these classifications mention tumor extension out of the nostril.

Staging is important to the surgeon to predict the outcome, to ascertain the chance of complete excision as well as to determine the approach to tumor resection. Not counting the extranasal tumor extension, the patient in this case was stage IIIA based on Radkowski classification. He underwent selective angiography to establish a definite diagnosis and to embolize the feeding vessels. This method has been widely recognized to reduce intraoperative blood loss which in turn facilitates the removal of tumor.¹⁰ Ungkanont *et al.* reported in a study with 43 cases that morbidity, recurrence and intraoperative complications decreased with embolization.¹¹ However, there are several risks of embolization including bleeding, allergic reactions and

incision and canine approach. He made an uneventful recovery. Histopathological examination showed that the tumor was made up of variably sized blood vessels set in a delicate fibrocollagenous background, consistent with JA. A post-operative CT scan showed residual enhancing mass in the left pterygopalatine fossa, infratemporal fossa, sphenoid sinus, and superior orbital fissure. He subsequently

**Table 1.** Stages of the Fisch classification⁶

Stage	Description
I	Tumors limited to the nasal cavity nasopharynx with no bony destruction.
II	Tumors invading the pterygomaxillary fossa, paranasal sinuses with bony destruction.
III	Tumors invading the infratemporal fossa, orbit and parasellar region remaining lateral to the cavernous sinus.
IV	Tumors with invasion to the cavernous sinus, optic chiasmal region and pituitary fossa.

Table 2. Other staging systems for juvenile angiofibroma

Chandler et al, 1984 ⁷	
I	Tumor confined to nasopharyngeal vault
II	Tumor extending into nasal cavity or sphenoid sinus
III	Tumor extending into antrum, ethmoid sinus, PMF, orbit, and/or cheek
IV	Intracranial tumor
Sessions et al, 1981 ⁸	
IA	Limited to nose and/or nasopharyngeal vault
IB	Extension into ≥ 1 sinus
IIA	Minimal extension into PMF
IIB	Full occupation of PMF with or without erosion of orbital bones
IIC	Infratemporal fossa with or without cheek erosion
III	Intracranial extension
Radkowski et al, 1996 ⁹	
IA	Limited to nose and/or nasopharyngeal vault
IB	Extension into ≥ 1 sinus
IIA	Minimal extension into PMF
IIB	Full occupation of PMF with or without erosion of orbital bones
IIC	Or posterior to pterygoid plates
IIIA	Erosion of skull base — minimal intracranial extension
IIIB	Erosion of skull base — extensive intracranial extension with or without cavernous sinus invasion

strokes. The patient in this case was free from these complications.

There are several modalities of treatment for JA, with surgical resection and irradiation therapy being the most successful. It is generally accepted that surgery is the treatment of choice while

radiotherapy is best for recurrence post surgery or extensive tumor with significant intracranial extension (where total resection with acceptable morbidity is unlikely). There is also a relatively large series of 55 patients with JA, where 42 underwent radiotherapy as the primary therapy and 80% were successfully treated.¹²

Surgical resection was essential for the patient in this case because he had a huge tumor protruding from his nostril, which was unlikely to regress with other treatments. It is also generally accepted that external approaches are superior in advanced tumors.¹³ In view of the tumor extent, the open approach was adopted. However, resection was incomplete due to intracranial extension. Hence, adjuvant radiotherapy was given for residual disease. Mendenhall *et al.* summarized six studies on the outcome of radiotherapy for JA.⁵ Most patients in these series were those who exhibited recurrence after prior surgery or had intracranial extension. After completing radiotherapy, local control rates ranged from 73% to 100%. Almost all those with recurrence after radiotherapy were successfully treated with another surgery or a second course of radiotherapy. Hence, the ultimate local control was nearly 100%.

Our case shows that a high index of suspicion is of paramount importance in the diagnosis of JA and alleviates the possibility of an unwarranted biopsy, as an indiscriminate biopsy could spell disaster. The most useful tools are MRI and arterial angiography. Once the definite diagnosis is established, the treatment plan has to be mapped out which is primarily surgical. Irradiation therapy has been reported to achieve satisfactory outcome, especially for unresectable residual disease and/or intracranial extension, where total surgical resection is unlikely to be attained without unacceptable morbidity.

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