

Juvenile Nasopharyngeal Angiofibroma: A Trap for the Unwary

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SUMMARY: Juvenile nasopharyngeal angiofibroma (JNA) is a rare, benign but aggressive tumour of the nasopharynx. It occurs primarily in male adolescents. The usual presentation is either nasal obstruction or epistaxis. We present a case with complete nasal obstruction of short duration. The management is discussed with reference to the literature. Because this tumour tends to occur in men in their second decade it is an important disease to consider in soldiers.

Case Report

A 19 year old craftsman was referred to the Ear, Nose and Throat outpatient department with a six week history of complete nasal obstruction. There was no history of epistaxis, no symptoms related to rhinitis or sinusitis, nor was there any significant past medical or family history.

Clinically there was complete obstruction of his nasal airways. Examination of the anterior nares was normal. Posterior rhinoscopy showed a large, smooth, glistening mass filling the whole posterior choanae and pressing on the soft palate. There were no enlarged lymph nodes in his neck. Lateral X-ray of the soft tissues of his nose and mouth demonstrated a soft-tissue mass filling the whole post-nasal space (Fig 1).

An examination under anaesthetic of the nose, sinuses and post-nasal space was performed. This was to establish the origin and size of the mass.

Examination through the mouth revealed a smooth, pedunculated mass, the size of a large plum. This completely filled the post-nasal space and extended into both nostrils. There was no extension into the skull base. By direct palpation it was found to be arising from the left pterygopalatine fossa and to be attached to the whole length of left medial pterygoid plate. The mass was then avulsed through the mouth with part of the medial pterygoid plate. Bleeding was initially controlled by direct pressure and then by insertion of anterior and posterior nasal packs. Total blood loss was approximately 400 millilitres and blood transfusion was not required. The macroscopic specimen is shown in Figure 2.

The packs were removed after two days with no subsequent bleeding. Post-operative recovery and follow-up was uneventful.

The photomicrograph at Figure 3 shows the tissue to be composed of numerous irregular blood vessels with a prominent tunica media. The intervening stroma is primarily fibrotic with occasional cells. These features are characteristic of JNA.



Fig 1. Lateral soft tissue X-Ray demonstrating a mass in the post nasal space.

Discussion

Complete nasal obstruction of short duration is an uncommon symptom which must have a significant structural cause. Juvenile nasopharyngeal angiofibroma is a tumour with a reported incidence from 1/5000 to 1/50,000 of otorhinolaryngological patients (1). The

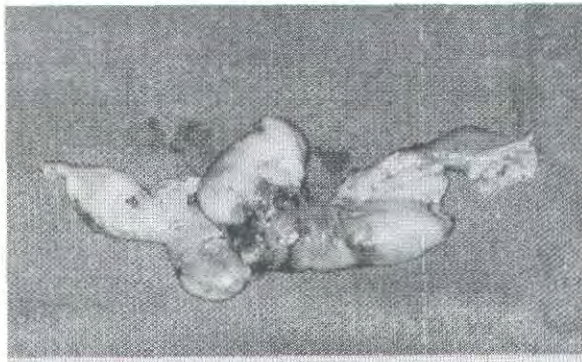


Fig 2. The specimen after removal from the post nasal space.

almost invariably occur in adolescent males (1,2,3,4,5).

The age and sex distribution of JNA supports a theory that angiofibromata are hormone dependent tumours arising as a result of androgen-oestrogen imbalance (1). However the tissue of origin remains unknown. The fibroblastic theory suggests abnormal growth of the embryonic occipital plate prior to its ossification at the age of 25 (6). An alternative theory of origin is from hamartomatous tissue because of the histological similarity between the tumour and nasal erectile tissue (1).

The tumour presents most often with epistaxis and nasal obstruction (4,5,7) though other presentations may occur if there is extension beyond the nasopharynx.

Clinical examination and plain X-rays are sufficient to provide a high level of suspicion of the diagnosis, which is confirmed by histological examination of the tissue. The highly vascular nature of the tumour makes biopsy dangerous. Therefore appropriate pre-operative investigations may be required prior to full assessment under general anaesthesia.

Various imaging techniques have been used in the diagnosis of JNA. Plain X-ray will show a nasopharyngeal soft tissue mass and possibly widening of the pterygopalatine fissure. Tomography may further help define the mass. Juvenile nasopharyngeal angiofibroma may be demonstrated by carotid angiography because it is highly vascular. This may show collateral blood vessels amenable to embolization prior to surgical resection. Computerised tomography scan and magnetic resonance imaging scan may also be used to help delineate the anatomy of the mass (8).

If the tumour is pedunculated, as occurred in our case, complete surgical removal is the treatment of choice (7). For unresectable lesions radiotherapy has been used but may cause sarcomatous change or affect growth of the facial skeleton (7). Androgens or oestrogens, cryosurgery, sclerotherapy and electro-coagulation may be used as adjuncts or to treat small recurrences (1,2,4).

This tumour is important in military practice because

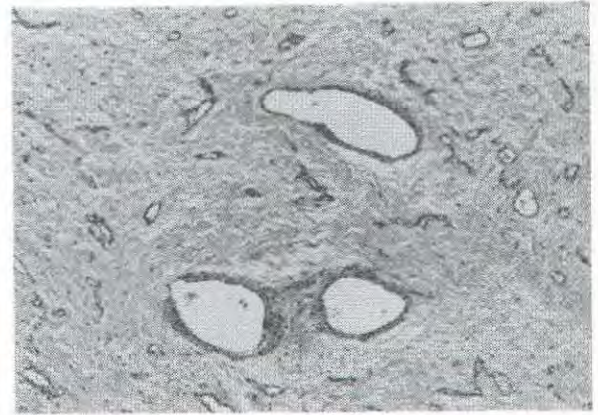


Fig 3. Photomicrograph confirming the diagnosis of juvenile nasopharyngeal angiofibroma.

it occurs within the military age group. Biopsy should be avoided because of the risk of uncontrollable bleeding. Adequate pre-operative and intra-operative assessment will ensure that significant haemorrhage during surgery is avoided.

Acknowledgements

The authors wish to thank Major D K MacIver FRCR (Consultant Radiologist, BMH Rinteln), Captain P Barrett (Army Histopathology Registry) and the Department of Medical Illustration (Royal Army Medical College) for their help in the preparation of the illustrations.

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