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Juvenile Nasopharyngeal Angiofibroma - A Case Report

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Introduction:

Juvenile nasopharyngeal angiofibroma is an uncommon benign tumor of the nasopharynx which arises in the region of the sphenopalatine foramen and it grows insidiously invading surrounding structures. It is the commonest benign tumor of the nasopharynx. It is a vascular tumor and believed to affect males exclusively. It presents commonly with nasal obstruction and recurrent epistaxis in young adolescent males. The nasopharynx is not easily accessible but has a high capacity and as such, tumors in this region tend to present late. Making a diagnosis of this condition requires a high index of suspicion as it mimics other sinonasal conditions and approaching it as such may result in fatal outcome.

Epidemiology:

Nasopharyngeal angiofibroma is seen almost exclusively in adolescent males, accounting for approximately 0.05 to 0.5% of head and neck tumors, and a reported incidence ranging from 1 in 150000 to 1 in 1500000. There are also reports that individuals from India and the Middle East appear to have an increased incidence when compared to those of European descent.The typical age range of JNA is 9 to 25 years, and though there have been case reports of NA diagnoses in older males, this is still considered a rare occurrence

Etiopathogenesis:

Nasopharyngeal angiofibroma characteristically demonstrates angiogenesis and vascular proliferation situated within the posterior nasal cavity, sphenopalatine foramen, and nasopharynx. There are suggestions that hormonal influences, chromosomal abnormalities, and overexpression of vascular growth factor receptors play a role, but much of this is still open to debate, and the exact mechanism remains unknown. Given the extensive vascularity of this tumor, recruitment of adjacent arterial supply and aggressive growth can cause osseous erosion and extension into the orbits, skull base, frontal and middle cranial fossae, and other high-value territories that can make treatment difficult.

Clinical presentation:

The presentation is typical with obstructive symptoms, epistaxis, and chronic otomastoiditis due to obstruction of the Eustachian tube. Patients may present with life-threatening epistaxis. On examination, it may be seen as a pale reddish-blue mass. It is, as the name suggests, very vascular and a biopsy can sometimes be fatal.

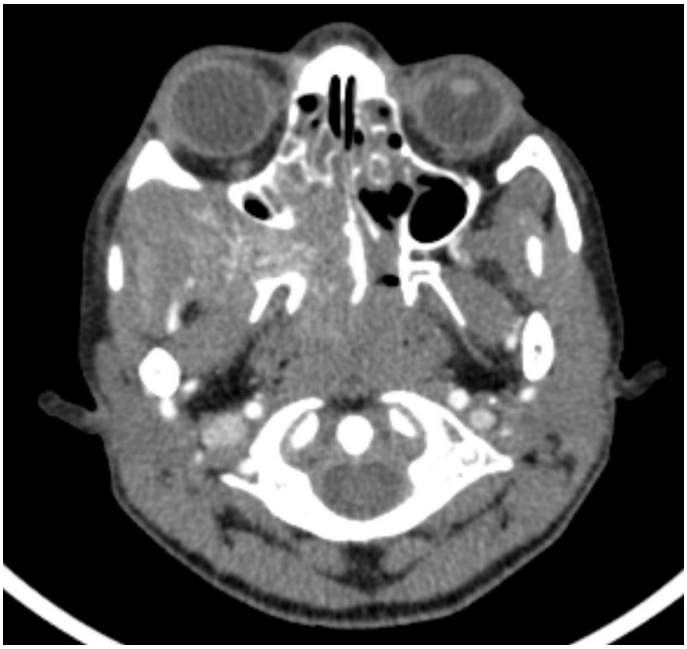
Case report:

15 year old male who presented with a 6 month history of nasal obstruction and recurrent nasal bleeding. He had had about ten episodes of epistaxis in the 3 months. Examination revealed that vital signs were essentially within normal limits.

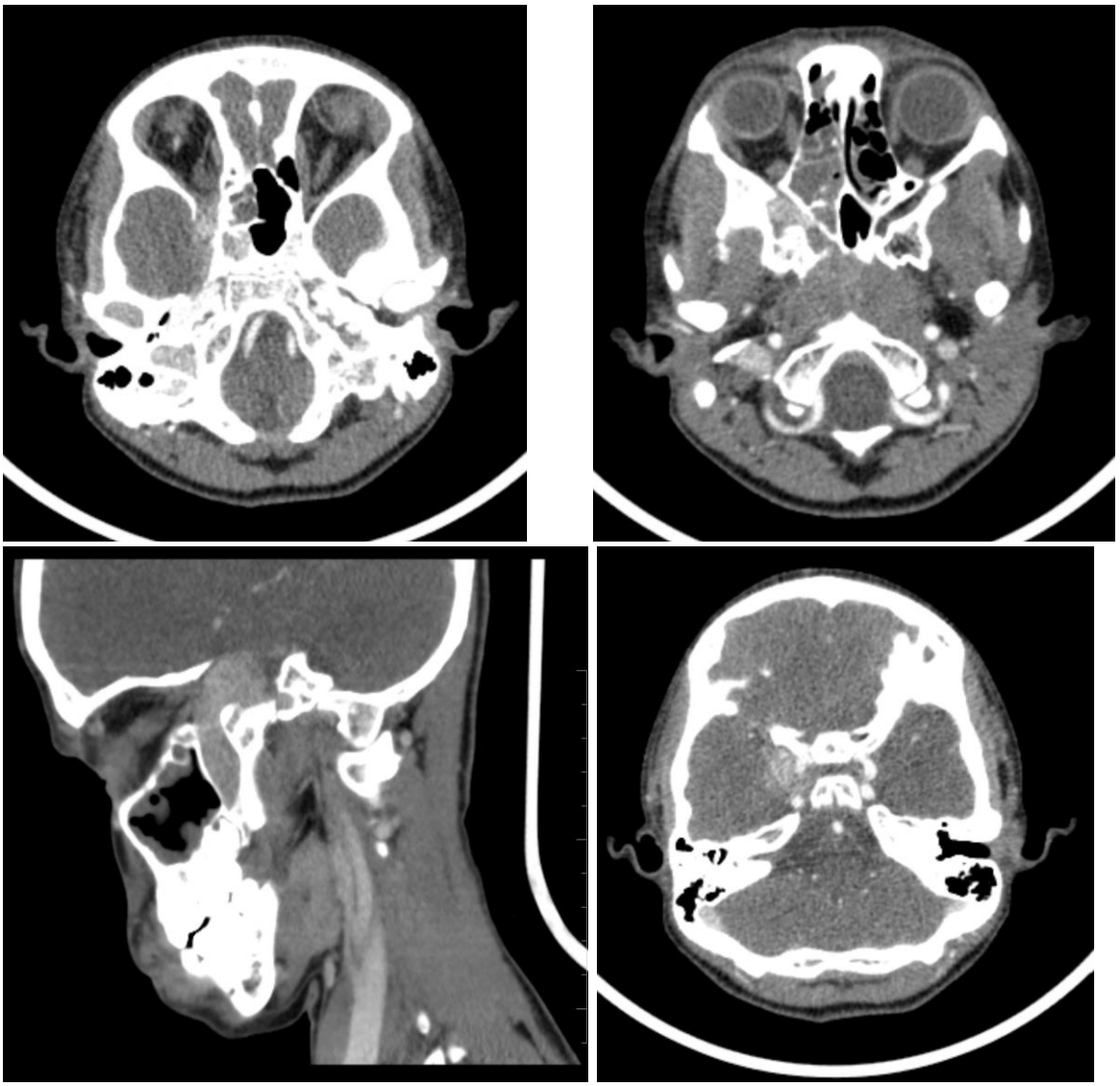
The nasal cavity revealed fresh blood in the right nasal cavity but the bleeding site could not be visualized. There was no nasal secretion and there was no area of tenderness on the face. No obvious mass was visualized in the oropharynx. An impression of epistaxis secondary rhinosinusitis was made.

Imaging:

CECT PNS

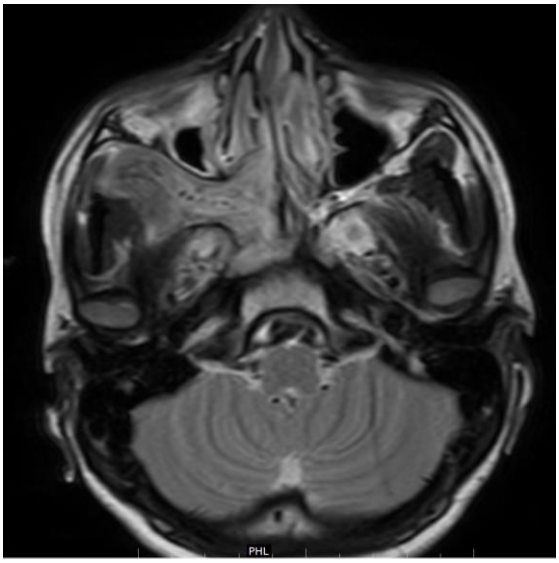


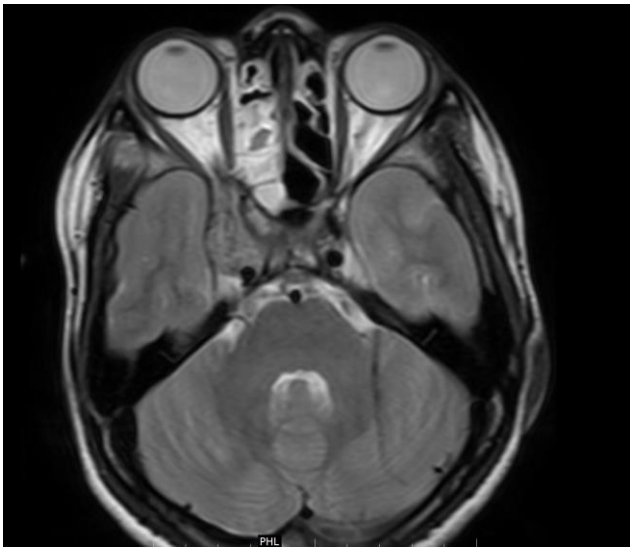
CECT PNS: Demonstrates avidly enhancing soft tissue mass within the posterior nasal cavity near the sphenopalatine foramen with extension to and/or beyond the nasopharynx, pterygopalatine fossa, and adjacent sinuses.



Intracranial extension into the right parasellar region and cavernous sinus, through the superior orbital fissure. Lesion is encasing the right cavernous ICA, without causing luminal narrowing or thrombosis

MRI PNS





T2W image of PNS axial section showing extensive, nasopharyngeal mass lesion with numerous flow voids. There is extension into the left infratemporal fossa, as well as intraorbitally through the right superior orbital fissure and orbital apex. In addition, there is a compression of left cavernous sinus, associated with encasement of the ICA cavernous segment, the later shows however preserved flow void.

Young male patients presenting with recurrent epistaxis JNA should be considered as differential diagnosis. CT and MRI play major role in diagnosing this condition since most patients will have normal clinical examination and laboratory findings although mass can be palpated in few cases.

Final diagnosis: JNA

Treatment:

Surgical resection (either open or, increasingly, endoscopic) is the treatment of choice, usually performed after preoperative embolisation to help with haemostasis. The embolisation may be performed up to five days prior to surgery. Irradiation may be an option if surgery is not possible or only incomplete resection has been achieved.

Teaching points:

CT is particularly useful at delineating bony changes. Findings are similar to those described above. Typically a lobulated non-encapsulated soft tissue mass is demonstrated centered on the sphenopalatine foramen (which is often widened) and usually bowing the posterior wall of the maxillary antrum anteriorly- “Holman Miller sign”

MRI is excellent at evaluating tumour extension into the orbit and intracranial compartments.

References:

1. Tork CA, Simpson DL. Nasopharyngeal Angiofibroma. [Updated 2022 Jun 27]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2023 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK545240/