Juvenile nasopharyngeal angiofibroma

Nasopharyngeal angiofibroma (also called juvenile nasopharyngeal angiofibroma) is a histologically benign but locally aggressive vascular tumor that grows in the back of the nasal cavity. It most commonly affects adolescent males.

Patients with nasopharyngeal angiofibroma usually present with one-sided nasal obstruction and recurrent bleeding.

Clinical features

Frequent chronic epistaxis or blood-tinged nasal discharge

Nasal obstruction and rhinorrhea

facial dysmorphism (when locally invasive)

Conductive hearing loss from eustachian-tube obstruction

Diplopia, which occurs secondary to erosion into superior orbital fissure and due to third and sixth nerve palsy, proptosis when having intraorbital extension.

Rarely anosmia, recurrent otitis media, and eye pain.

Diagnosis

If nasopharyngeal angiofibroma is suspected based on physical examination (a smooth vascular submucosal mass in the posterior nasal cavity of an adolescent male), imaging studies such as CT or MRI should be performed. Biopsy should be avoided as to avoid extensive bleeding since the tumor is composed of blood vessels without a muscular coat.

Antral sign or Holman-Miller sign (forward bowing of posterior wall of maxilla) is pathognomic of angiofibroma.

Differential diagnosis

Antro-choanal polyp (benign neoplasm)

Rhinosporidiosis (as bleeding point is here too)

Malignancy—nasopharyngeal carcinoma, lymphoma, plasmacytoma, rhabdomyosarcoma

Chordoma

Nasopharyngeal cyst

Pyogenic granuloma

Treatment

Treatment for Nasopharyngeal angiofibroma (JNA) is primarily surgical. The tumor is primarily excised by external or endoscopic approach. Medical treatment and radiation therapy are only of historical interest.

External approaches:

transpalatine approach

transpalatine + sublabial (Sardana's) Approach

infratemporal Approach

nasal endoscopic Approach

Transmaxillary Approach

Endoscopic approach is an excellent tool in primary and recurrent JNA, it allows visualisation and precise removal of the lesion. Preoperative embolisation of tumour may be of some use in reducing intraoperative bleeding.

Outcome

Prognosis for nasopharyngeal angiofibroma is favorable. Because these tumors are benign, metastasis to distal sites does not occur. However, these tumors are highly vascularized and grow rapidly. Removal is important in preventing nasal obstruction and recurrent epistaxis. Mortality is not associated with nasopharyngeal angiofibroma.

Case series

A report describes a graduated multiangle, multicorridor, endoscopic approach to JNAs that is illustrated in 4 patients, each with a different tumor location and extent. Four different surgical corridors in varying combinations were used to resect JNAs, based on tumor size and location, including an ipsilateral endonasal approach (uninostril); a contralateral, transseptal approach (binostril); a sublabial, transmaxillary Caldwell-Luc approach; and an orbitozygomatic, extradural, transcavernous, infratemporal fossa approach (transcranial). One patient underwent resection via an ipsilateral endonasal uninostril approach (Corridor 1) only. One patient underwent a binostril approach that included an additional contralateral transseptal approach (Corridors 1 and 2). One patient underwent a binostril approach with an additional sublabial Caldwell-Luc approach for lateral extension in the infratemporal fossa (Corridors 1-3). One patient underwent a combined transcranial and endoscopic endonasal/sublabial Caldwell-Luc approach (Corridors 1-4) for an extensive JNA involving both the lateral infratemporal fossa and cavernous sinus.

A graduated multiangle, multicorridor approach was used in a stepwise fashion to allow for maximal surgical exposure and maneuverability for resection of JNAs. Gross-total resection was achieved in all 4 patients. One patient had a postoperative CSF leak that was successfully repaired endoscopically. One patient had a delayed local recurrence that was successfully resected endoscopically. There were

no vascular complications.

An individualized, multiangle, multicorridor approach allows for safe and effective surgical customization of access for resection of JNAs depending on the size and exact location of the tumor. Combining the endoscopic endonasal approach with a transcranial approach via an orbitozygomatic, extradural, transcavernous approach may be considered in giant extensive JNAs that have intracranial extension and intimate involvement of the cavernous sinus [1].

Case reports

A 21 year-old male with a history of left proptosis and diplopia of two weeks of onset. The MRI showed an ethmoid-orbital vascular lesion with anterior skull base invasion and orbital extension. Biopsy of the ethmoid confirmed fibrovascular tissue, which supported the diagnosis of angiofibroma.

It is a benign neoplasm with local characteristics of malignancy due to its ability to invade adjacent areas. In this case, the debut presented with manifestations of orbital extension. A broad and multidisciplinary approach is needed in order to improve prognosis [2].

1)
Liu JK, Husain Q, Kanumuri V, Khan MN, Mendelson ZS, Eloy JA. Endoscopic graduated multiangle, multicorridor resection of juvenile nasopharyngeal angiofibroma: an individualized, tailored, multicorridor skull base approach. J Neurosurg. 2015 Nov 13:1-11. [Epub ahead of print] PubMed PMID: 26566205.
2)
Hervás Ontiveros A, España Gregori E, Climent Vallano L, Rivas Rodero S, Alamar Velázquez A, Simal Julián JA. [Juvenile nasopharyngeal angiofibroma with orbital extension]. Arch Soc Esp Oftalmol. 2015 Jan;90(1):22-5. doi: 10.1016/j.oftal.2014.02.010. Epub 2014 Nov 20. Spanish. PubMed PMID: 25443183.