

JUVENILE NASOPHARYNGEAL ANGIOFIBROMA - CASE REPORT

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Abstract

Juvenile nasopharyngeal angiofibroma is a rare highly vascular, benign tumor, very aggressive locally, affecting male adolescents or those at puberty. The occurrence in other age groups or in women is exceptional. The agresiveness and high vascularization make surgery very difficult, and surgical biopsy is not recommended. Surgery is the main method of treatment used, especially for early stages, while for advanced stage cases and having intracranial extension, radiation remains the chosen therapeutic option. In order to establish a preoperative diagnosis and staging, we use CTscan imaging or MRI with and without contrast material as well as angiography. In this study we will present a 20-year-old patient suffering from juvenile nasopharyngeal angiofibroma who was diagnosed using a CT scan. The tumor was successfully removed surgically and the HP examination confirmed the diagnosis. The 7 months post-surgery evolution was favorable.

Key words: Angiofibroma, nasopharyngeal, paralatero-nasal, CTscan

Introduction

Juvenile nasopharyngeal angiofibroma is a rare, histologically benign tumor of the nasopharynx, affecting almost exclusively male teenagers or at puberty, being diagnosed in 14-25 old boys. It is a highly vascularized tumor with high tendency towards local and loco-regional invasion, with extension within the submucosa. For this reason it's a destructive tumor. From a histological point of view, it's a mezenchymal tumor, vascular, having high cells, composed of fibrous connective tissue and abundant vascular endotelium lined spaces (1). There are various theories regarding the formation of angiofibroma, but the most accepted theory is the theory of angiogenesis and histogenesis. This describes angiofibroma as a pure vascular tumor which proliferates in a hemangiomatous manner and in which all other components, including fibrous connective tissue, are derived from non-differentiated vasoformator mezenchymal (2). The tumor origin can be found on the posterolateral wall of the nasopharynx at the upper edge of the sphenopalatine foramen, which is also a way for the sphenopalatine artery, a branch of the internal maxillary artery (3,4). More exactly, it is located where the sphenoid

process of the maxilar bone meets the horizontal wing of the vomer and pterygoid foot of the sphenoid (5). The juvenile nasopharyngeal angiofibroma represents 0.5% of head and neck neoplasms and is considered the most common benign neoplasm of the nasopharynx (2, 5, 6, 7). Despite its benign nature, the local growth and destruction method can cause bone remodeling. Although it does not produce bone erosion, the tumor has a high potential of life threatening complications such as: epistaxis, intracranial extension and massive intraoperative bleeding (8-10).

Classic signs of juvenile nasopharyngeal angiofibroma are progressive nasal obstruction and / or epistaxis and rhinorrhea, together with other symptoms that are dependent on the direction and extent of the tumor (6). The tumour extension in adjacent areas can cause "swelling facial" type deformities as well as proptosis.

Diagnosis is based on history, clinical examination and radiological and imaging results, knowing that biopsy is not recommended.

Angiography helps demarcate the tumor's blood supply and identify the vessels that feed it. However its real use is still questionable. Surgical excision was the main angiofibroma treatment and the approach normally depends on the tumor extension. An important role in this regard is played by the CT scan and appropriate staging.

Case Report

In February 2012, a 20-year-old male residing in a rural area arrives at the ENT Clinic in Timisoara. From history we learn that the young man suffers from left side unilateral nasal obstruction, headaches, oral breathing, repeated epistaxis from the left nasal fossa which have been persisting for 2 years. From history we learn that the young man was operated in 2001 for nasal polyps.

The ENT clinical examination reveals while performing anterior rhinoscopy that the posterior half of the left nasal fossa is occupied by a white tumoral formation, smooth, firm, without other local changes. While performing posterior rhinoscopy we observe a nasopharynx occupied entirely by the tumor. The external facial aspect shows no phisionomy or left / right symmetry feature changes.

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A CT scan with bone contrast and MRI are performed, which show a well defined left nasopharyngeal tumor mass, containing soft tissue, which reaches the soft palate on the left and enters the left nasal fossa and bulges the medial wall of the maxillary sinus by volume compression, without causing bone erosion (left maxillary sinus integral medial

wall bone contour), left maxillary sinusitis with nasal septum deviation to the right due to tumor volume in the left nasal fossa, without changing its bone contour, and extension in the pterygopalatine fossa and left infratemporal fossa (fig. 1 and 2).



Fig.1. Skull MRI axial section highlighting left nasal fossa tumoral extension, nasopharynx, pterygopalatine fossa and infratemporal left fossa.



Fig.2. Skull CT scan axial section visualizing the lump in the left nasal cavity, nasopharynx, and pterygopalatine fossa and infratemporal left fossa.

The patient is hospitalized in our ENT clinic in Timisoara on 03/07/2012 when after clinical examination and imaging he is diagnosed with juvenile nasopharyngeal angiofibroma and is proposed to have surgery, which is accepted by the patient. We begin preoperative preparations with biological and interdisciplinary investigations appropriate for this type of surgery, according to agreed protocol, which includes preanesthetic consultation. The surgical intervention takes place on 03/13/2012 in AG with IOT, intraoperative bleeding control technique by controlled hypotension.

Surgery is performed in two stages:
 - stage 1 - left laterocervical horizontal incision, 4 cm of the mandibular arch, removing the subcutaneous tissues, highlighting the left sternocleidomastoid muscle, highlighting the hypoglossal nerve, and the tiro-glossal-facial venous trunk - anteroinferior (Farabeuf triangle); with highlighting the common carotid artery and its bifurcation. Necessity ligation of the venous trunk is performed and a waiting thread is placed on the left external carotid artery. (fig. 3).



Fig. 3. Ligation of the venous trunk and waiting thread on the left external carotid artery.

- stage 2 - the chosen approach was a left paralateronasal incision ranging from the nasal vestibule to 1 cm of the internal edge, removing and delimiting a firm, hard tumoral formation, with very fixed insertion on the maxillar, on the ethmoid bones and sphenoid horn. Total

tumor ablation is performed as well as hemostasis by anterior and posterior compressive dabbing (fig. 4). A sample is taken from the extracted piece for histopatological examination. On the 16.03.2012 dabbing is done with no active bleeding.

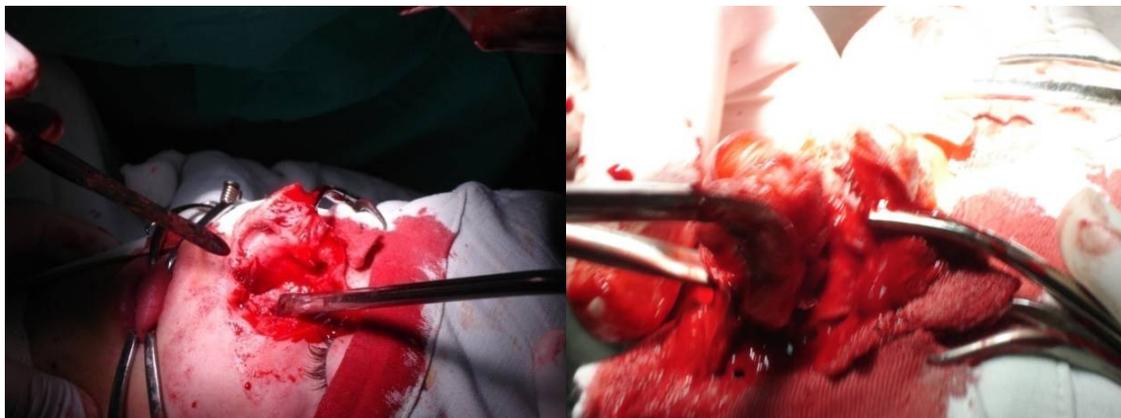


Fig. 4. Paratransnasal approach and extraction of the lump.

Positive diagnosis was based on history, clinical ENT examination imaging laboratory tests, intraoperative findings and HP (fig. 5). The histopathological examination describes a proliferating tumor consisting of connective-fibrous tissue, with hyalinization areas including vascular thin-walled spaces and blood vessels of different sizes (small and large), some with thin walls, some with thickened

walls or deformed by compression exerted by connective tissue. Myxoid areas focally observed. Stromal cells present nuclei with moderate pleomorphism. These histological aspects advocate for nasopharyngeal angiofibroma. Thus the diagnosis of juvenile nasopharyngeal angiofibroma clinical stage IIC, according to the Radkowski classification of 1996.

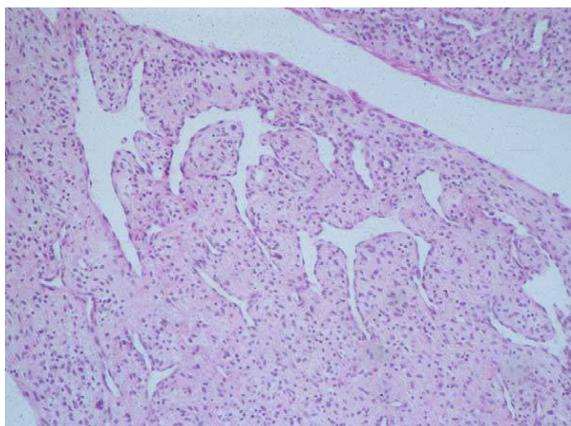


Fig. 5. Cells of origin are myofibroblast.

Differential diagnosis is made: sinuso-choanal polyp, osteoma, craniopharyngioma, neuroblastoma, chordoma, chondrosarcoma, rhabdomyosarcoma, nasopharyngeal carcinoma, hemangiopericytoma.

Evolution is expected to be good, with no further relapse, given that the patient underwent radical surgery and that he is at an adult age. The patient will be clinically and imaginatively cared for in a dispensary (nasal and nasopharyngeal endoscopy). The prognosis is positive.

Currently, 7 months after the surgery, the patient has a good evolution without recurrence of clinical changes; the imaging assessment being performed at 24 months postoperatively.

Postoperative complications that can occur are: infraorbital nerve injury, atrophic rhinitis crust, anosmia, serious medium otitis, local dental complications, sinusitis.

Discussions

Tumor incidence is rare, being found predominantly in the developing males. The occurrence age varies between 11-20 years with an average of 14 years (3). The occurrence exclusively in boys is probably related to androgen receptors (5). Diagnosis is made mainly on clinical manifestations of the tumor. Most of the times, the presence of the triad: progressive nasal obstruction, epistaxis and nasopharynx tumor in young men suggests the diagnosis of juvenile nasopharyngeal angiofibroma. In some studies the appearance of anterior radiological bulging of the posterior wall maxillary sinus (antral sign) is a pathognomonic sign. An essential role in preoperative diagnosis of the angiofibroma is played by endoscopic evaluation of the patient, and imaging: CT scan and MRI, which apply to all patients evaluated in our clinic. It is well known that

preoperative biopsy should be avoided because of the risk of subsequent massive hemorrhage. Therefore preoperative diagnosis is a stage diagnosis, because the safety diagnosis is established only postoperatively by histopathological examination from the surgically extracted piece.

The choice of surgical technique is still debatable, but surgery remains the main treatment that completely removes the tumor. Traditional techniques used in surgical treatment of juvenile nasopharyngeal angiofibroma are: transpalatal technique, transpharyngeal, Denker-Rouge, medial maxillectomy, lateral transfacial rhinotomy, mediofacial degloving and Le Fort I osteotomy, as well as infratemporal or subtemporal lateral approach. In this case the technique used was the paralateronasal approach (12).

The preoperative hormonal therapy with estrogen and progesterone is without noticeable effects (5).

External radiation radiotherapy treatment is made with doses of 3600-4600 cGy/23Fr/5Fr/week/31 days after CT simulation. Complications that can occur after or during radiation therapy are: epistaxis, panhypopituitarism, temporal lobe necrosis, cataracts, post-irradiation keratitis (6).

Regional selective embolization proves to be effective only for stage I patients. It can be used preoperatively to reduce the vascular intake in case of endoscopy approach. It is burdened by complications such as soft tissue and skin necrosis, facial paralysis or endocranial complications.

For unresectable tumors, chemotherapy may be used as an alternative to radiotherapy; the most commonly used chemotherapy drugs are Adriamycin and Decarbazine (8).

Conclusions

Juvenile nasopharyngeal angiofibroma is a benign, rare tumor of the nasopharynx, which, due to its strategical anatomical position can affect several vital structures of the skull base.

The tumor almost exclusively affects male teenagers. The triad: progressive nasal obstruction, epistaxis and tumor mass in the nasopharynx leads to the clinical diagnosis of juvenile nasopharyngeal angiofibroma. The nasopharynx tumor size is not necessarily in accordance with the real tumor extension and size, it can be only the "tip of the iceberg". Extranasopharyngeal extension is very common in all cases - the most common being the nasal extension.

A CTscan with contrast material is pathognomonic for the diagnosis of juvenile nasopharyngeal angiofibroma and allows accurate staging of the tumor, which is very necessary for choosing the surgical technique, estimating prognosis and reporting results.

Radio therapy must be reserved for inoperable cases, for recurrences or for patients who refuse surgery because of various reasons.

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