Particular aspects in endoscopic surgery for juvenile nasopharyngeal angiofibromas. Case reports and review of literature

V. BUDU1), I. BULESCU1), CARMEN AURELIA MOGOANTĂ2)

1)"Prof. Dr. Dorin Hociotă” Institute for Phono-Audiology and Functional ENT Surgery, Bucharest
2)ENT Department, University of Medicine and Pharmacy of Craiova

Abstract
Juvenile nasopharyngeal angiofibromas (JNA) are rare, benign, highly vascular tumors which appear in proximity to the sphenopalatine foramen. The tumor arises most commonly in adolescent males suggesting that it could be hormonally responsive. Although it is histologically benign, it has a high destructive potential and a high grade of recurrence. It is a fibrous vascular tumor with vascular sources from branches of the external or internal carotid arteries. Modern treatment of JNA includes surgery and also radiotherapy, chemotherapy and hormone therapy. This paper presents two cases from the eight of our clinic’s experience, with morphological features which made their treatment challenging.

Keywords: juvenile angiofibroma, nasopharynx, endoscopic surgery, arteriography.

Introduction
Juvenile nasopharyngeal angiofibromas represent between 0.05% and 0.5% of head and neck neoplasms [1–3]. It is a highly vascular tumor, which affects primarily adolescent males [4]. Usually, JNA patients present with unilateral nasal obstruction, recurrent epistaxis and nasopharyngeal mass in adolescent males [1]. The histological appearance of the tumor is benign, although some theories suggest that it could possibly be a vascular malformation [4–7]. The tumor usually arises near the sphenopalatine foramen and extends towards nearby structures [8, 9]. Histopathologically, JNAs are non-encapsulated tumors composed of a mixture of blood vessels and fibrous stroma [1, 3, 10]. Microscopic examination of the tumor reveals plump fibroblasts of ovoid to spindle shape and a high amount of connective tissue. The stroma contains blood vessels of different sizes and shapes lined by endothelial cells but with little or no smooth muscle or elastic fibers (Figure 1).

This structure, lacking muscles fibers, contributes to the capacity of JNAs to bleed excessively after minimal manipulation [1, 11, 12], thus making its surgical treatment a challenge.

Because of the rich vascularity of this type of tumor, the vascular sources that supply it have been long studied and observed. Usually, the blood supply of JNA comes from branches of the external carotid arteries, but sometimes also from the internal carotid system [9, 13]. According to the study of Tang IP et al. [9] the tumors were supplied by branches from both left and right maxillary arteries in 69.2% of cases and 30.8% were supplied by branches from the ipsilateral maxillary artery. These results were similar to the ones presented by Pryor SG et al. [14], where bilateral and unilateral contributions to the vascular supply of the tumor were almost in numbers. Knowledge of the vascular supplies of JNA is of great importance prior to any surgical procedure for this tumor. For that reason, pre-operative angiography is usually a standard procedure. Treatment for JNA is usually surgical. Other treatment methods have been emphasized in the past such as chemotherapy, radiotherapy or hormone therapy. These methods are nowadays only used as occasional complementary treatments [1, 8, 10, 15]. Surgical approaches for JNA include classic approaches such as medial maxillotomy, transpalatine, lateral rhinotomy, mid-facial degloving, and more recently video-assisted nasal endoscopic surgery [8, 14, 16–18]. The type of surgical approach for JNA, classical or endoscopic is limited by the extension of the tumor in surrounding areas such as pterygopalatine fossa or infratemporal fossa [19]. However, Mitskavich MT et al. [17] concluded that the tumor may be treated exclusively via nasal endoscopy even when it is extended to the pterygopalatine of infratemporal fossae [8]. The endoscopic
or classic approaches are chosen in relation to the tumoral stage. Staging of JNA has been heavily disputed among authors according to certain criteria like behavior of the tumor [20], routes of spread, difficult areas for surgical exposure and possible sites of recurrence [4]. According to Andrade NA et al. [8], the current staging used for JNA is that of Andrews JC et al., which classifies local and regional spreading of the tumor, as well as bone destruction [22]. Whatever the stage, or the surgical technique used, most authors consider that preoperative selective arterial embolization of feeding vessels of the tumor has significantly decreased intraoperative bleeding and facilitated resection of larger tumors [1, 23].

**Patients, Methods and Results**

This paper presents two cases from the eight of our clinic’s experience, with morphological features which made them challenging from a surgical point of view. Both patients underwent preoperative computed-tomography scans and angiography. Both cases were resolved using an endoscopic endonasal surgical technique.

**Case No. 1**

The first case was an 18-year-old male, which presented with unilateral nasal obstruction and recurrent epistaxis following surgery for JNA with ligation of the left external carotid artery three years before presentation. The CT-scan revealed a tumoral mass, which completely blocked the rhinopharynx and the posterior third of both nasal fossae. Preoperative angiography revealed a vascular-type tumor with feeding vessels from the left internal carotid artery (Figure 2). The protocol used for this case was endoscopic endonasal surgical excision with prior dissection and exposure of the left common carotid artery.

**Case No. 2**

The second case was a 14-year-old male, which presented with bilateral nasal obstruction, epistaxis and rhinophonia following previous adenoidectomy. The CT-scan revealed a mass obstructing the rhinopharynx extending to the left pterygopalatine fossa and the left sphenoid sinus. Preoperative angiography showed a highly vascular tumor with feeding vessels from both left and right maxillary arteries (Figure 3). The protocol in this case was preoperative angiography with selective arterial embolization and complete endoscopic endonasal resection of the tumor and its expansions.

**Figure 2** – Case No. 1: CT-scan showing the tumoral mass obstructing the rhinopharynx. Pre-operative angiography revealing vascular sources from the internal carotid artery.

**Figure 3** – Case No. 2: Pre-operative angiography revealing the tumor with vascular sources from both left and right maxillary arteries.
For both cases, the tumors were examined in the pathology compartment revealing areas with numerous anastomotic blood vessels, with relatively thin walls, teleangiectatic appearance and heavily developed fibrous stroma. Also, immunohistochemical analysis showed receptors for estrogen, progesterone and androgen hormones in the stromal cells, CK7, CK8/18 positive in surface epithelium, SMA, CD31, CD34 positive for the blood vessels and S100 positive for the stromal cells and nervous fibers. The histological diagnosis was nasopharyngeal angiofibroma.

Discussion

The highly vascular nature and the structure of JNA with numerous blood vessels lacking muscular fibers makes this particular tumor a never ending challenge as it is for head and neck surgeons. Nowadays improvements of endoscopic endonasal surgical techniques enable the use of endoscopic surgery for complete removal of JNA [1]. However, the decision to perform JNA resection in head and neck surgeons. Various treatment options have been emphasized for its treatment (surgery, radiotherapy, chemotheraphy, hormone therapy). The gold standard treatment method in selected cases is endoscopic surgery combined with pre-operative selective embolization of the main blood vessels feeding the tumor, although special cases such as recurrences and vascular variations should be considered separately. Endoscopic endonasal surgery in JNA is only limited by the extension of the tumor in surrounding anatomical regions, and the experience of the surgical team.

Conclusions

JNA is a highly vascular tumor with aggressive local behavior that affects primarily adolescent males. It has a high persistence and recurrence rate and represents a challenge to head and neck surgeons. Various treatment options have been emphasized for its treatment (surgery, radiotherapy, chemotheraphy, hormone therapy). The gold standard treatment method in selected cases is endoscopic surgery combined with pre-operative selective embolization of the main blood vessels feeding the tumor, although special cases such as recurrences and vascular variations should be considered separately. Endoscopic endonasal surgery in JNA is only limited by the extension of the tumor in surrounding anatomical regions, and the experience of the surgical team.

References


Corresponding author
Vlad Budu, MD, PhD, Department of ENT Microsurgery, “Prof. Dr. Dorin Hociotă” Institute for Phono-Audiology and Functional ENT Surgery, 21 Mihai Cioranu Street, 050751 Bucharest, Romania; Phone +4021–410 21 70, e-mail: vladbudu@yahoo.com

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