# Endoscopic surgery for juvenile angiofibroma: A critical review of indications after 46 cases

Piero Nicolai, M.D.,[1] Andrea Bolzoni Villaret, M.D.,[1] Davide Farina, M.D.,[2] Sylvie Nadeau, M.D.,[3] Arkadi Yakirevitch, M.D.,[4] Marco Berlucchi, M.D.,[1] and Cristina Galtelli, M.D.[1]

## ABSTRACT

**Background:** *At present, transnasal endoscopic surgery is considered a viable option in the management of small–intermediate size juvenile angiofibromas (JAs). The authors critically review their 14-year experience in the management of this lesion to refine selection criteria for an endoscopic approach.*

**Methods:** *From January 1994 to May 2008, 46 patients were treated by a pure endoscopic resection after vascular embolization (87%). The lesions were classified according to Andrews (Andrews JC, et al., The surgical management of extensive nasopharyngeal angiofibromas with the infratemporal fossa approach, Laryngoscope 99:429–437, 1989) and Önerci (Önerci M, et al. Juvenile nasopharyngeal angiofibroma: A revised staging system, Rhinology 44:39–45, 2006) staging systems. All patients were followed by regular endoscopic and magnetic resonance imaging (MRI) examinations.*

**Results:** *Lesions were classified as follows: stage I, n = 5; stage II, n = 24; stage IIIa, n = 14; stage IIIb, n = 3 according to Andrews classification system; stage 1, n = 9; stage II, n = 12; stage III, n = 26 according to Önerci's system. Unilateral blood supply was detected in 39 (85%) cases. Feeding vessels from the internal carotid artery (ICA) were also reported in 14 (30%) patients. Intraoperative blood loss ranged from 250 to 1300 mL (mean, 580 mL). In four (8.7%) cases, suspicious residual disease was detected by MRI. In one patient, a 1-cm persistent lesion was endoscopically removed because septoplasty was required and a slight increase in size was noticed. The other three lesions, all located in the root of the pterygoid plate, are nearly stable in size and are currently under MRI follow-up.*

**Conclusion:** *The improvement of surgical instrumentation and the experience acquired during a 14-year period have contributed to expanding the indications for endoscopic surgery in the management of JAs. Even stage III lesions may be successfully managed, unless the ICA is encased or if it provides an extensive blood supply. An external approach may be required when critical structures such as the ICA, cavernous sinus, or optic nerve are involved by lesions that are persistent after previous treatment; such a situation may prevent safe and radical dissection with a pure endoscopic approach. Better understanding of the factors influencing the growth of residual lesions is needed to differentiate those requiring re-treatment from those which can be simply observed.*

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**Key words:** Classification, embolization, Endoscopic surgery, juvenile angiofibroma, nasopharyngeal tumor

Juvenile angiofibroma (JA) is a benign lesion that originates from the pterygopalatine fossa and shows characteristic epidemiological features and growth patterns. The typical patient is an adolescent male subject with an age ranging from 12 to 18 years, presenting with mono- or bilateral nasal obstruction and epistaxis. The advent of endoscopes and the refinement of imaging studies have offered the otolaryngologist the possibility to achieve early diagnosis. In fact, endoscopic examination can easily identify a mass with smooth surface and clear signs of hypervascularization growing behind the posterior insertion of the middle turbinate. Computed tomography (CT), or even magnetic resonance imaging (MRI), which can provide greater detail, generally shows a lesion vividly enhancing after contrast medium administration, which has its epicenter of growth at the level of the pterygopalatine fossa and frequently involves the nasopharynx, the nasal cavity, the sphenoid sinus, and the infratemporal fossa.

There is general agreement that surgery plays a key role in the management of JA. The fact that many approaches (suprahyoid, transpalatal, transmaxillary through a lateral rhinotomy or a midfacial degloving, facial translocation, Le Fort I, infratemporal, and craniofacial resection) have been described clearly indicates that there is no "ideal" technique, but rather that the surgeon should have several different techniques available that can be used according to the extent of disease. Recently, transnasal endoscopic surgery has been extensively used as an excellent alternative to external approaches in the management of small–intermediate size JAs.[1–8] Our experience with the endoscopic technique for this specific disease began in 1994, and in 2003 we reported the results in a group of 15 patients.[9] This study, which updates our previous experience, is now based on a series of 46 patients with JA treated by endoscopic surgery and was planned with the intent to reassess the indications and limitations of this approach and to identify factors that influence the outcome.

*From the Departments of [1]Otorhinolaryngology, and [2]Radiology, University of Brescia, Brescia, Italy, [3]Department of Otorhinolaryngology, Enfant Jesus Hospital, University Laval, Quebec City, Canada, and [4]Department of Otorhinolaryngology, Sheba Medical Center, Tel Hashomer, Israel*

*Address correspondence and reprint requests to Piero Nicolai, M.D., Department of Otorhinolaryngology, University of Brescia, Piazza Spedali Civili 1, 25123 Brescia, Italy*

*E-mail address: pieronicolai@virgilio.it*



## PATIENTS AND METHODS

### Patient Selection

A total of 46 patients who underwent endoscopic resection for JA were identified from a database of 238 patients treated for benign tumors of the sinonasal tract and nasopharynx at the Department of Otorhinolaryngology of the University of Brescia (Italy) between January 1994 and May 2008. Epidemiological and clinical data, surgical reports, pre- and postoperative images, details concerning complications, and follow-up information were retrieved and analyzed.

### Diagnostic Workup

All of the lesions were prospectively staged according to the classification described by Andrews *et al.*[10] and retrospectively based on the Önerci *et al.*[11] staging system. Physical and endoscopic examination was combined with imaging studies such as MRI, CT and intraarterial diagnostic angiography. Angiographic study was performed 24–72 hours before surgery in the majority of patients with special focus on contralateral blood supply and afferent vessels arising from the internal carotid artery (ICA). Vascular embolization was performed at the same time with polyvinyl alcohol particles.

### Inclusion Criteria and Surgical Techniques

Our selection policy for endoscopic treatment has evolved during the years with our increasing expertise. Basically, all patients with

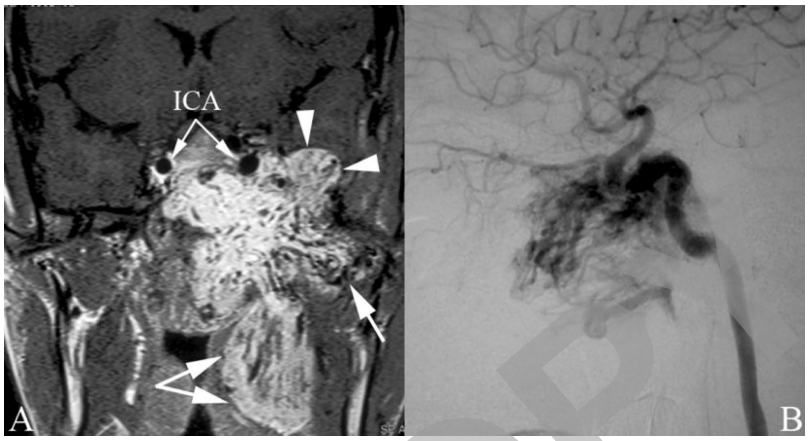


*Figure 1. Contrast enhanced magnetic resonance imaging (MRI) acquired in the (A) coronal plane and (B) digital subtraction angiography. The large JA invades the parapharyngeal space (double arrows), the greater sphenoid wing and masticator space (single arrow), and the middle cranial fossa (arrowheads). Within the sphenoid sinus, the lesion contacts both internal carotid arteries (ICAs); note hypertrophy of the left artery. Digital subtraction angiography shows multiple feeders from the internal carotid, thus contraindicating endoscopic resection.*

lesions involving the nasopharynx, pterygopalatine fossa, nasal fossa(e), sphenoid sinus, maxillary sinus, ethmoid, infratemporal fossa, basisphenoid, and parasellar region were considered eligible for endoscopic surgery. An extensive vascular supply from ICA, its encasement, or an intracranial extension lateral to the paraclival segment were considered contraindications, which dictated the need for combining an endoscopic approach with a subtemporal craniotomy exposing the horizontal portion of the ICA (Fig. 1). Moreover, endoscopic surgery was deemed contraindicated in residual lesions involving critical areas (ICA, optic nerve, cavernous sinus, and dura), where adhesions increase the risk of severe uncontrolled complications during dissection of the lesion.

All of the procedures were performed under controlled hypotension by the senior author (P.N.). The surgical technique has already been described in detail.[9] The main steps can be summarized as follows, highlighting some of the technical nuances more recently introduced:

1. After proper decongestion of both nasal cavities was obtained, partial or total resection of the middle turbinate, uncinectomy, middle antrostomy, anterior and posterior ethmoidectomy were first performed to expose the nasal/nasopharyngeal component of the lesion and to gain space for easy movement of endoscopes and instruments.
2. Independently of adhesion of the lesion with the interchoanal septum, the posterior third of the nasal septum was resected to enhance exposure of the nasopharynx and to have the possibility to work through both nostrils with the two-handed technique. This required the cooperation of an assistant with some expertise in endoscopic sinus surgery who kept the lesion or normal tissue under traction during dissection and at the same time suctioned blood to keep the surgical field as clear as possible.
3. The posterior wall of the maxillary sinus was resected as far lateral as dictated by the lateral extent of the lesion to expose the pterygopalatine or even the infratemporal fossa according to individual needs. After incising the periosteum, which is wrapping the pterygopalatine fossa, its content was exposed and the sphenopalatine or the internal maxillary artery was identified, occluded with clips, and transected.
4. If the lesion involved the sphenoid sinus, a wide sphenoidotomy with resection of the anterior and inferior wall was performed. This was essential to better control the extension of the lesion along the vidian canal and to clearly define its relationship with critical structures such as the optic nerve, ICA, and middle cranial fossa. Large-volume lesions, especially those with massive lateral extension into the infratemporal fossa (Fig. 2) needed to be removed in two to three blocks by dividing the different components using a diode laser (Dornier Medizin-Laser Gmbh, Germering, Germany) set on pulse mode (18 W, 0.1 second).

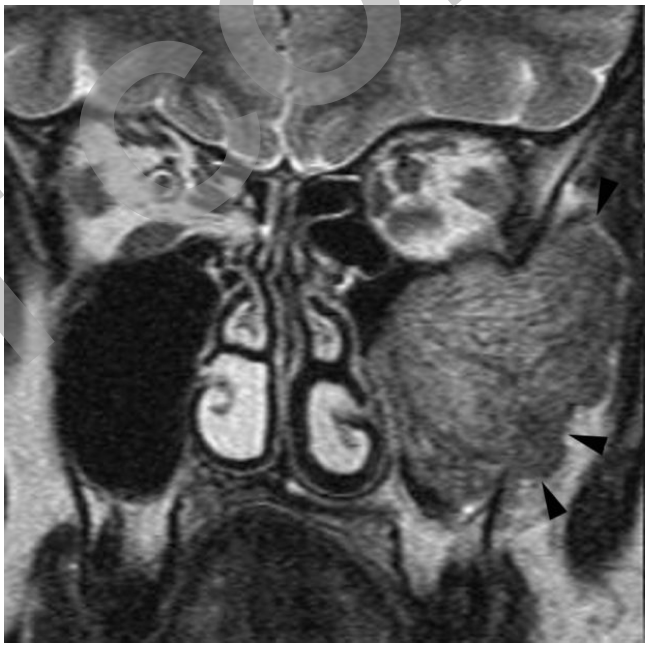

*Figure 2. TSE T2 sequence acquired in the coronal plane. Magnetic resonance imaging (MRI) shows lateral extension of JA through the pterygomaxilary fissure to reach the infratemporal fossa (arrowheads).*

5. More typically, the nasal/nasopharyngeal/sphenoidal part was removed first, the infratemporal part was removed second, and the retropterygoidal part was removed last.
6. Whenever there were clear radiological and intraoperative findings of involvement of the cancellous bone of the basisphenoid, tumor digitations were meticulously identified and the adjacent skull base was extensively drilled, with special reference to the vidian canal and the pterygoid root.

## Follow-Up

All patients were prospectively followed according to a schedule including endoscopic examination and MRI every 4 months during the 1st postoperative year and subsequently every 6 months for 4 years. Since 2006, this policy was slightly modified by performing the first MRI the same day the patient had the nasal packing removed or the day after. Subsequent endoscopic and MRI examination followed every 6 months.

## Statistical Analysis

Statistical analysis was performed using a commercially available computer software package (SPSS for Windows, Version 10.0.11999; SPSS, Inc., Chicago, IL). The influence on blood loss and blood transfusion of different parameters (uni-/bilateral vascular supply, presence of ICA vascular supply, and stage of the disease) was analyzed by Mann-Whitney and Pearson chi-square tests, for continuous and categorical variables, respectively. Furthermore, the impact of different factors (previous surgery, different sites of extension, and vascular pattern) on the presence of persistent disease was analyzed by a Fisher's exact test. Finally, the ability of different classification systems[10,11] to predict the presence of persistent disease was also evaluated by Pearson's chi-square test. All value of $p < 0.05$ were considered statistically significant.

## RESULTS

All patients were white male subjects ranging in age from 10 to 35 years (mean, 17 years). Five patients (12.5%) had been previously treated at other centers. Four patients had undergone surgery through a lateral rhinotomy or a transpalatal approach, and one had received radiotherapy (RT) and estrogen therapy.

Nasal obstruction and recurrent epistaxis were the most common presenting symptom and sign, occurring in >97% of patients, followed by nasal discharge (7%), headache (4%), pain (4%), and cheek swelling (2%). Clinical staging of the lesions according to Andrews *et al.*[10] and Önerci *et al.*[11] are summarized in Tables 1 and 2, respectively.

For each patient, CT and/or MRI with contrast enhancement were obtained. MRI was performed in 43 (93.5%) patients and CT was performed in 23 (50%) patients. JA involved the nasopharynx in all but one case in which the residual lesion was limited to the pterygopalatine fossa. Involvement of the pterygopalatine fossa was detected in 38 (82.6%) cases, of the sphenoid sinus in 28 (70%) cases, of the cancellous bone of the pterygoid process in 23 (50%) cases, of the infratemporal fossa in 16 (35%) cases, of the maxillary sinus in 7 (15%) cases, and of the ethmoid sinus in only 2 (4%) cases. Two (4%) patients had intracranial extradural extension. Intraorbital involvement was detected in one case (2%). Limited pushing of the orbital content by the lesion through the inferior orbital fissure was not considered as orbital involvement (Table 3).

With regard to vascularization, eight (17%) patients presented adjunctive blood supply from the contralateral external carotid artery system through the maxillary, sphenopalatine, and/or pharyngeal artery. Moreover, 14 (30%) patients also had a vascular supply from the ICA, through siphon vessels (78%), the mandibular artery (35%), and the ophthalmic artery (7%). Embolization was performed uneventfully in 40 (87%) patients, and in 6 (13%) patients, in view of the limited size of the lesion with no major lateral extension, the procedure was not applied and surgery therefore included as a first step the exposure of the pterygopalatine fossa and bipolar cauterization or clipping of the sphenopalatine artery.

Intraoperative blood loss varied from 250 to 1300 mL (mean, 580 mL). Intraoperative blood transfusion was required in five patients (11%). Moreover, in eight (17%) patients blood transfusion was required during the postoperative course. Nasal packing was removed 1 to 3 days after surgery (mean, 2 days). No minor or major postoperative complications were observed, and the mean hospitalization time was 5 days (range, 2–13 days).

All patients are still under regular follow-up (mean, 73 months; range, 9–172 months). In four (8.6%) patients a persistent lesion was suspected (Table 4). One patient presented a 16-mm lesion involving the floor of the sphenoid sinus 24 months after surgery at MRI examination. This lesion showed an increase in size (from 16 to 21 mm) over a period of 20 months. Because the patient required septoplasty, after proper counseling it was decided to concomitantly plan endoscopic removal of the persistent JA. In two additional patients a nodule suspicious for persistence (respectively, 14 and 9 mm) located in the root of the pterygoid process was detected by MRI examinations performed 9 and 4 months after surgery. The last suspicious persistence appeared like a double nodular lesion, with one component located in the basisphenoid and the other in the cancellous bone of the pterygoid root (Fig. 3). All these lesions are asymptomatic and undetectable during standard endoscopic examination. Two lesions are stable in size over a follow-up of 24 and 39 months, respectively; one lesion located in the pterygoid root showed 2-mm decrease in size over a follow-up of 39 months, possibly because of resolution of perilesional inflammation.

The advanced stage of the disease was statistically related to both blood loss ($p = 0.02$) and the need for blood transfusion ($p = 0.02$). Moreover, blood loss was significantly increased in case of bilateral vascular supply ($p = 0.02$) and the presence of feeding vessels from ICA ($p = 0.005$). In contrast, none of the factors considered was significantly related to the detection of persistent disease. Finally, none of the classification systems used[10,11] was able to predict the presence of persistent disease.

Table 1 **Andrews *et al.*[10] staging system for juvenile angiofibroma**

| | |
|---|---|
| Type I | Tumor limited to the nasopharyngeal cavity; bone destruction negligible or limited to the sphenopalatine foramen |
| Type II | Tumor invading the pterygopalatine fossa or the maxillary, ethmoid, or sphenoid sinus with bone destruction |
| Type III | Tumor invading the infratemporal fossa or orbital region without intracranial involvement (a) or with intracranial extradural (parasellar) involvement (b) |
| Type IV | Intracranial intradural tumor without (a) or with (b) infiltration of the cavernous sinus, pituitary fossa, or optic chiasm |

Table 2 **Önerci *et al.*[11] staging system for juvenile angiofibroma**

| | |
|---|---|
| Type I | Nose, nasopharyngeal vault, ethmoidal-sphenoidal sinuses or minimal extension to PMF |
| Type II | Maxillary sinus, full occupation of PMF, extension to the anterior cranial fossa, and limited extension to the ITF |
| Type III | Deep extension into the cancellous bone at the base of the pterygoid or the body and the greater wing of sphenoid, significant lateral extension to the ITF or to the pterygoid plates posteriorly or orbital region, cavernous sinus obliteration |
| Type IV | Intracranial extension between the pituitary gland and internal carotid artery, tumor localization lateral to ICA, middle fossa extension, and extensive intracranial extension |

*ICA = internal carotid artery; ITF = infratemporal fossa; PMF = pterygomaxillary fossa.*

Table 3 **Staging of patients according to Andrew's *et al.*[10] and Önerci's *et al.*[11] classification**

| Andrew's Classification | No. of Patients (%) | Önerci's Classification | No. of Patients (%) |
|---|---|---|---|
| I | 5 (11%) | I | 9 (20%) |
| II | 24 (52%) | II | 12 (26%) |
| IIIa | 14 (30%) | III | 25 (54%) |
| IIIb | 3 (7%) | | |

Table 4 **Previous treatment, staging, preoperative extension, and vascular supply of lesions considered suspicious for persistence after endoscopic resection**

| Patient No. | Age (yr) | Previous Treatment | Stage (Andrews) | Stage (Önerci) | Involved Anatomical Sites | Contralateral ECA Vascular Supply | ICA Vascular Supply |
|---|---|---|---|---|---|---|---|
| 1 | 14 | — | I | I | NP | Absent | Absent |
| 2 | 18 | — | IIIa | II | NP, PPF, ITF | Absent | Absent |
| 3 | 16 | Biopsy* | IIIa | III | NP, PPF, MS, SS, ITF | Present | Present |
| 4 | 19 | — | IIIb | III | NP, PPF, SS, ITF, O, PP, SB, IC ED | Absent | Present |

**Biopsy was performed at another Institution.*
*NP = nasopharynx; PPF = pterygopalatine fossa; MS = maxillary sinus; SS = sphenoid sinus; ITF = infratemporal fossa; O = orbit; PP = pterygoid plates; SB = skull base; IC ED = intracranial extradural extension; ECA = external carotid artery; ICA = internal carotid artery.*

## DISCUSSION

The pathogenesis of JA is still a matter of debate. Recent immunohistochemical and electron microscopy studies have suggested that the lesion may be considered a vascular malformation (or hamartoma) rather than a tumor.[12] These observations led Schick *et al.*[13] to postulate that JA might be caused by incomplete regression of a branchial artery, which arises in embryogenesis between days 22 and 24 and forms a temporary connection between the ventral and dorsal aorta. This artery commonly regresses and forms a vascular plexus that either involutes or may leave remnants potentially leading to JA development. This theory is supported by the finding that JA vessels express laminin $\alpha 2$, which is considered to be a marker for early angiogenesis.[14]

JA has a characteristic propensity to develop within the pterygopalatine fossa and to grow medially through the sphenopalatine

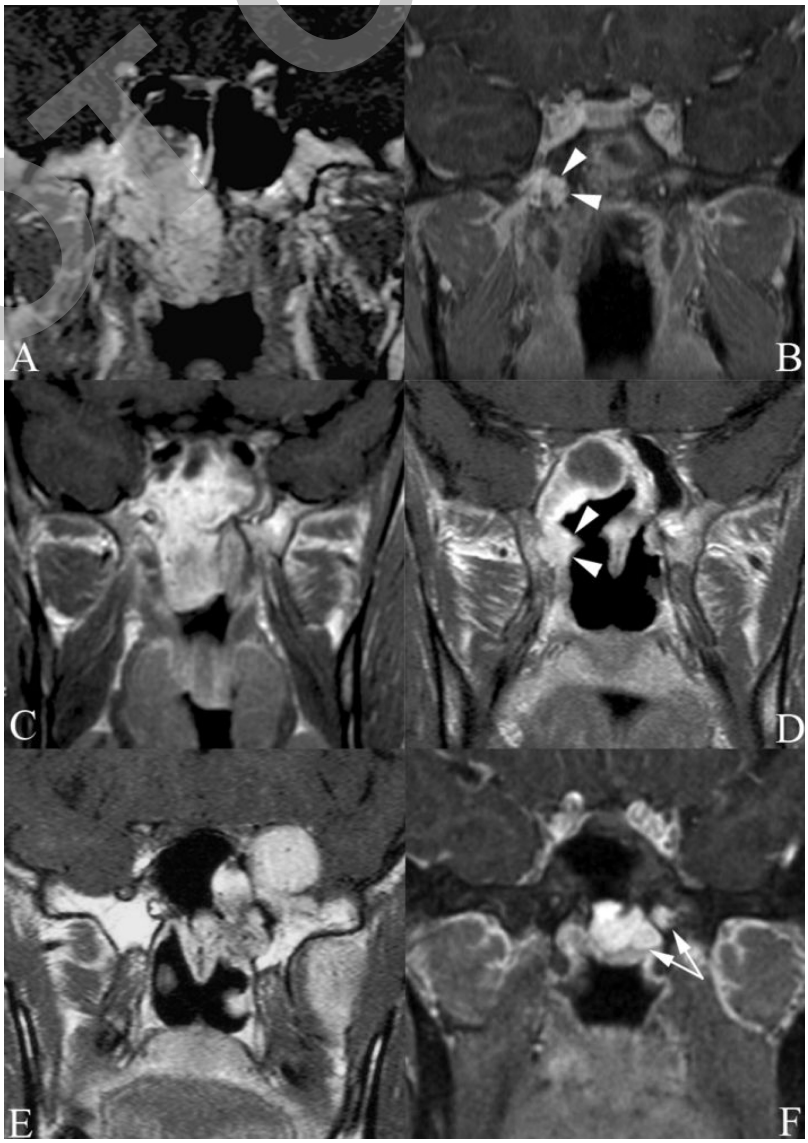


***Figure 3.*** *Suspected persistent juvenile angiofibromas (JAs). (A, C, and E) Pretreatment and (B, D, and F) follow-up contrast-enhanced magnetic resonance imaging (MRI) examinations are compared in three different patients; all displayed images were acquired in the coronal plane through the sphenoid bone. Suspected persistent JA is shown in panels B and D as a brightly enhancing nodule located in the right pterygoid root (arrowheads in panels B and D). The third lesion is composed of two nodules located in the basisphenoid and left pterygoid root, respectively (arrows in panel F).*

foramen along a submucosal plane as well as in other multiple directions crossing foramina and fissures of the adjacent skull base. Through these low resistance pathways, the lesion may extend into the middle cranial fossa *via* the foramen lacerum, along the maxillary nerve, or through the superior orbital fissure. Bone may be involved through two mechanisms: resorption by pressure coming from subperiosteal growth or invasion of the cancellous component initially at the level of the root of the pterygoid process along the vidian canal, with subsequent extension posteriorly toward the upper middle clivus and laterally within the greater wing of the sphenoid with erosion of the floor of the middle cranial fossa and possible intracranial growth.

Diagnosis of JA is based on its typical site of origin and its hypervascular appearance after contrast enhancement. Both of these elements can be adequately shown using either multislice CT or MRI. Nonetheless, in our experience the latter is superior in showing the permeative invasion of diploic bone, commonly seen at the pterygoid root and greater sphenoid wing. Furthermore, MRI is superior in delineating the intracranial extent of JA, allowing detailed information on the relationships between the lesion, dura, cavernous sinus, and the nerves running through it. Finally, during follow-up the superb contrast resolution of MRI improves detection of persistent lesions in a background of scar tissue and inflammation.

Surgery has always been considered the ideal treatment for JA. However, in the preembolization era, intraoperative bleeding was so dramatic that the procedure was a true challenge for the surgeon and was associated with high rates of persistent disease and notable morbidity. In the early 1970s, the introduction of this technique,[15] which is commonly performed 48 hours before surgery, has revolutionized the treatment of JA by dramatically decreasing intraoperative bleeding and therefore making the assessment of tumor borders more accurate at dissection. Although the currently available methods of embolization provide excellent devascularization of feeders coming from the internal maxillary artery and its branches as well as from the ascending pharyngeal artery, control of major vascularization from the ICA is demanding. Devascularization by direct tumor puncture and embolization is associated with an unacceptable risk of major neurological complications[16] and has been therefore abandoned. In the event of encasement of the internal artery, which is nowadays a very rare event, balloon occlusion test and sacrifice of the ICA or, as a less invasive procedure, stenting of the intratemporal carotid artery, may be considered.[17]

Even though in our experience we did not observe any major complications after embolization, one should keep in mind that this technique may be potentially associated with severe complications such as neurological deficits and loss of vision due to occlusion of the central retinal artery.[18] Another source of criticism toward embolization is from the experience of Lloyd *et al.*, who considered this procedure as a risk factor for persistence because of the increased difficulty in identifying tumor margins particularly at the interface with the basisphenoid.[19]

Some of the surgical approaches (*i.e.*, transpalatal and suprahyoid) proposed in the past for JA resection should now be considered obsolete because they do not allow good exposure of the area involved by the lesion. Among the transfacial techniques, midfacial degloving has gained popularity in view of the absence of skin incision and wide access to the nasopharynx, maxillary sinus, sphenoid, infratemporal fossa, and skull base. Infratemporal or subtemporal approaches may be reasonably combined with a transnasal or transfacial route when there is the need to have the ICA under control. In this scenario, transnasal endoscopic approach has emerged during the recent years as a viable alternative to external techniques in view of the excellent visualization of the surgical field through multiangled exposure, the low morbidity without any external scar and osteotomy, and short hospitalization times. Based on our previous experience,[9] we concluded that endoscopic surgery was suitable for all stage I–II lesions, but only for small-size stage III JA. In recent years, however, we have successfully managed a small number of JA with extensive involvement of the infratemporal fossa. We found that some technical refinements during dissection may greatly help in appropriately exposing the lesion and removing it without leaving residual tissue behind. The first is related to dissection of the pterygopalatine/infratemporal fossa: once the posterior wall of the maxillary sinus has been removed, the periosteum should be largely incised to expose the tumor. This maneuver is critical to identify a correct cleavage plane between the lesion and adjacent soft tissues and to proceed with the dissection using a two-handed technique, having the assistant gently pull on the lesion while the surgeon separates it from fat and pterygoid muscles until the maxillary artery is detected. A similar technique is used when the lesion is extending to the parasellar area in close contact with the cavernous sinus. The copious venous bleeding that usually follows the removal of an intracranial tumor projection is easily controlled by placing polyanhydroglucuronic acid or hemostatic matrix. Another surgical maneuver we found extremely helpful when dealing with large volume lesions is sectioning it in two or more blocks. This provides a greater area for instruments to move around the lesion and to adequately expose the interface with the adjacent tissues. The use of a diode laser allows improving bleeding control.

Comparison of the results using external and endoscopic techniques is hindered by several biases related to stratification by staging and follow-up methodology. However, the recurrence rate of series with a consistent number of patients reported in the 1990s and treated with an external technique is around 36–40%.[19,20] More recently, Danesi *et al.* showed that a transfacial approach (through lateral rhinotomy or midfacial degloving) can lead to excellent results, with only 13.5 and 18.2% of residual disease in lesions with extracranial and intracranial extent, respectively.[21] As shown by our results, endoscopic resection achieved good results not only in stage I–II but also in stage IIIa–b lesions, according to Andrews, without any major complication and an acceptable overall rate of residual disease (8.6%). This is in accordance with the reports of other authors.[7,8,22] Interestingly enough, despite the fact that we extensively drilled the basisphenoid adjacent to the lesion, all four residual lesions we observed were located at this level. This observation reinforces the recommendation of Howard et al to urge the surgeon to extend drilling well beyond the apparent margin of infiltration.[23] Contraindications for an endoscopic approach are still a matter of debate, but in our opinion lesions with extensive blood supply from or encasing the ICA, extending intracranially lateral to the paraclival segment of ICA, or growing through the dura are better treated with an external approach, which according to different situations may include midfacial degloving or a subtemporal craniotomy with ICA exposure. Even JAs that are persistent after previous surgery and with critical relationships with ICA, optic nerve, or dura should be scheduled, after discussing with the patient the need to intraoperatively shift to an external procedure.

Postoperative surveillance is based on the combination of endoscopic examination and imaging techniques. However, since most of the residual lesions tend to grow submucosally, contrast-enhanced CT or MRI play a key role in their early detection. Based on the experience reported by Kania *et al.*,[24] who recently suggested obtaining a CT in the immediate postoperative course for early identification of any suspicious residual disease, we have started to scan patients with MRI the same day or the day after removal of nasal packing. In this setting, MRI does not show any of the inflammatory changes that, after 3–4 months, frequently make the differentiation between small residual lesions and active scar tissue challenging. To date, not one of the patients who had an early postoperative MRI have shown any residual lesion even at subsequent MRI examination. However, a longer follow-up is needed before drawing any meaningful conclusion about the validity of this surveillance policy.

In regard to therapeutic strategies alternative to surgery, it is still a matter of debate if RT can be considered a safe therapeutic option in young patients who have the likelihood to develop sarcomatoid degeneration of residual JA or radio-induced neoplasms in the following decades. Kuppersmith *et al.*[25] proposed the use of intensity-modulated RT for treatment of extensive and persistent JA to limit irradiation of noble structures. They reported no acute toxicity and only two limited

cases of late toxicity with epistaxis and persistent rhinitis. No patient developed recurrence. Reddy *et al.*[26] treated 15 patients affected by advanced JA (Andrew's type IIIb and IV) with RT. Local control obtained was 85% with two local persistences. Five (33%) patients developed late complications, including cataracts, transient central nervous system syndrome, and basal cell carcinoma of the skin. In our opinion, RT might be limited to selected cases of unresectable disease.

Another topic that needs to be addressed is the best treatment option for patients with residual disease and what should be the most appropriate follow-up protocol. At the time of writing, in our series only one patient with persistent and growing disease has required endoscopic revision. The three other residual lesions are stable in size by MRI examination. Önerci *et al.* suggested that a "wait and see" policy for intracranial residual disease may be preferable to infratemporal craniofacial resection.[11] In addition, one recent interesting case report reintroduced the concept of spontaneous involution of JA. The case described was never treated and was followed for 8 years during which time the lesion constantly decreased in size.[27] In this regard, Reddy *et al.* has noted that regression of a residual lesion may take up to 3 years.[26]

## CONCLUSION

Endoscopic surgery allows better exposure and visualization of JA projections because of angled telescopes and avoids unsightly scars with no interference with craniofacial bone growth while decreasing postoperative pain and hospitalization time. However, extensive training is required especially with the two-handed technique, and appropriate instrumentation is necessary. Endoscopic surgery is undoubtedly a good option for management of JA and can be considered appropriate for early staged lesions and in the case of extension into the infratemporal fossa, orbital cavity, or parasellar region. Refinement in surgical techniques as well as in dedicated instrumentation could make it possible to further expand the indications to lesions with intracranial extradural extension. Particular attention should be paid in selecting patients who had previous surgery for an endoscopic approach, because the adhesions between the tumor and critical structures as ICA, optic nerve, and dura can increase the risks of the operation. More data are needed to improve our knowledge on the biological behavior of residual lesions, on factors affecting their evolution, to consequently better define treatment strategy.

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