# Juvenile Nasopharyngeal Angiofibroma

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**KEYWORDS**

• Juvenile nasopharyngeal angiofibroma • Endoscopic surgery • Embolization • Sinus surgery • Epistaxis • Sinonasal tumor • Skull base tumor • Nasopharyngeal tumor

| EBM Question | Level of Evidence | Grade of Recommendation |
|---|---|---|
| Is morbidity less with endoscopic resection (blood loss, cosmesis, CN injury, etc) compared to open surgery? | 3b | C |

## SYMPTOMS OF JNA

The most common presenting symptoms of patients with JNA are unilateral nasal obstruction occurring in 91% and epistaxis occurring in 63% of patients.[2] Other related symptoms include nasal discharge; pain; sinusitis; facial deformity; otologic symptoms, such as hearing impairment and otitis media; and ocular symptoms of proptosis and diplopia.[2] Symptoms are generally present for 6 months to a year before the patient is diagnosed. JNAs are typically found in the male population between the ages of 10 and 24 years, with a median age at diagnosis of 15 years.[2]

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**Key Points: Juvenile Nasopharyngeal Angiofibroma**

- Endoscopic resection of early-stage juvenile nasopharyngeal angiofibromas (JNAs) is a safe and surgically sound treatment that has multiple advantages over traditional open approaches, including better cosmesis, decreased blood loss, shortened hospital stay, and equivalent or improved recurrence rates.
- Magnetic resonance imaging (MRI) is the imaging standard for postoperative surveillance and should be performed within the first postoperative year to detect recurrent/residual disease.
- First described by Chaveau in 1906,[1] JNAs are highly vascular benign tumors that primarily affect the young male population. These neoplasms are rare, accounting for approximately 0.05% of head and neck tumors. Although benign and slow growing, these tumors are locally aggressive and can cause extensive bone destruction, intracranial hemorrhage, facial deformity, severe epistaxis, and blindness. JNAs derive from the superior border of the sphenopalatine foramen. As the tumors enlarge, they extend through well-defined pathways into the infratemporal fossa, cavernous sinus, sphenoid sinus, middle cranial fossa, and, rarely, anterior cranial fossa. Surgical resection has been the mainstay of treatment of JNAs, and multiple open surgical approaches have been proposed depending on the location and extent of the tumor. Radiation therapy has been mostly reserved for unresectable tumors, residual disease after surgical resection, or recurrences occurring in anatomically critical areas. However, over the past 2 decades, endoscopic techniques and technology have improved, and an increasing number of JNAs are being removed endoscopically or in combination with traditional open approaches. Endoscopic techniques offer many advantages over open approaches, with better cosmesis (no external surgical scar), reduced intraoperative blood loss (usually combined with preoperative embolization), decreased hospital stays, and improved or equivalent recurrence rates.

## PATHOLOGY OF JNA

JNAs are characterized on gross pathology as well-defined, mucosalized, red to purple masses found in the nasal cavity and nasopharynx (**Fig. 1**). Histologically, the tumor is composed of 2 main components: spindle- or stellate-shaped cells embedded in a rich collagen matrix and a complex vascular arrangement of blood vessels that vary in size from capillaries to large venous channels. Characteristically, these vessels lack elastic laminae and elastic fibers and have vascular walls that vary in thickness. These features account for the tendency of these tumors to hemorrhage easily.[3] JNAs occur exclusively in men and are thought to be partially androgen dependent. JNAs possess multiple hormone receptors, including testosterone, dihydrotestosterone, and androgen.[4] However, investigation into hormone therapy for these tumors has been disappointing. JNAs may originate from a residual vascular plexus left behind after the involution of the first branchial artery,[5] but the cause of JNAs is still under debate.

## IMAGING

Computed tomography (CT) and MRI are both critical to the proper evaluation of angiofibromas. CT better delineates bony details of the skull base, including bony erosion, in particular, the depth of invasion into the bone of the sphenoid sinus, a main predictor of recurrence (**Fig. 2**). The extent of invasion into the cancellous bone of the sphenoid is difficult to determine intraoperatively, and this leads to a high likelihood of residual tumor and recurrence.[6,7] CT scans are also commonly used for intraoperative stereotactic surgical navigation systems to confirm the extent and resection of tumor. On the other hand, MRI is crucial for highlighting soft tissue