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JUVENILE NASOPHARYNGEAL ANGIOFIBROMA: A DIAGNOSTIC AND THERAPEUTIC APPROACH IN TERTIARY CARE CENTRE OF CENTRAL INDIA.

Dr. Harsh Agrawal	Associate Professor, Department of Otorhinolaryngology, Amaltas Institute of Medical Sciences
Dr. M. S. Hora	Associate Professor, Department of Pathology, Amaltas Institute of Medical Sciences, Dewas
Dr. Rajesh Golash	Associate Professor, Department of Pathology, R. D. Gardi Medical College, Ujjain
Dr. Vivek Singh	PG 3rd Year Student, Department of Otorhinolaryngology, Amaltas Institute of Medical Sciences, Dewas
Dr. Anushree Pradhan	PG 2nd Year Student, Department of Ophthalmology, Amaltas Institute of Medical Sciences, Dewas
Dr. Abhishek Arya	PG 1st year Student, Department of Otorhinolaryngology, Amaltas Institute of Medical Sciences, Dewas
Dr. Amarnath Yadav*	Assistant Professor, Department of Pathology Amaltas Institute of Medical Sciences, Dewas *Corresponding Author
Dr. Abhiraj Ramchandani	Professor, Department of Pathology, Amaltas Institute of Medical Sciences, Dewas
Dr. Kirti Hemwani	Associate Professor, Department of Microbiology R. D. Gardi Medical College, Ujjain

ABSTRACT Juvenile nasopharyngeal angiofibroma is the common tumor of nasopharynx.The prognosis for this disease is extremely good if diagnosed well in time and if the tumor has not extended intracranially. Preoperative selective arterial embolization has decreased intraoperative blood loss and facilitated resection of larger tumors. Transnasal endoscopic resection preserves both the anatomy and physiology of the nose, requires less rehabilitation days after surgery,and is highly successful for selected patients. Radiation therapy is generally reserved for larger and/or unresectable tumors but has severe complications..Radio surgery has several advantages over surgery or classic radiation therapy. However, further experiences and studies are required to confirm the usefulness of radiosurgery on JNA.

KEYWORDS : Juvenile Nasopharyngeal Angiofibroma, Histopathology,embolization, Treatment, Radiosurgery

INTRODUCTION

Juvenile nasopharyngeal angiofibroma (JNA) represents 0.05% to 0.5% of all head and neck tumors but is the common tumor of nasopharynx (1,2). It affects almost exclusively male adolescents. The median age at diagnosis is 15years (2-4).Histological benign appearance is often counter balanced by a potentially malignant clinical course,due to severe epistaxis, involvement of endocranial structures with occasional surgical difficulties and complications, and a high incidence of recurrence. Patients usually present at late stage of the disease that arises from either the lateral wall or the roof of the nasopharynx especially the spheno palatine foramen (1,3,5) Theblood supply is most commonly from the internal maxillary artery.Histologically tumor blood vessels typically lack smooth muscle and elastic fibres(3,5,6). Severe epistaxes accompanied by progressive nasal obstruction are the classical symptoms of juvenile angiofibromas at the time of presentation. Its classical treatment is surgery; however there are cases in which may indicate radiotherapy or even hormone therapy and gamma knife surgery (GKS)(2).Currently it is advisable to perform a selective tumoral embolization prior to proceeding with any of the techniques known in order to facilitate surgical access (5,7-10).

Histopathology

Histologically angiofibroma is always composed of an intricate mixture of blood vessels and fibrous stroma(3-4,11-13).Microscopically, there are plump fibro blasts, ovoid tospindle shaped, with a generous amount of connective tissue.In the compact stroma are blood vessels of different sizes and shapes lined by plump endothelial cells but with little or no smooth muscle or elastic fibers (5-6,11) (Figure 1). This lack of muscle undoubtedly contributes to the tumor's capacity for massive hemorrhage following minimal manipulation.

MATERIAL AND METHODS

The study was conducted on 40 newly diagnosed patients of JNA Selected from AIMS dewas. Ethical clearance had been taken from the appropriate institutional authority. JNA is an important disease in otolaryngo-logical practice. It is a rare, vascular, benign tumor with high recurrence, and typically affects adolescent boys. The prognosis for this disease is extremely good if diagnosed well in time and if has not extended intracranially. Preoperative angiography and embolization minimize the intraoperative blood loss and the current shift in the treatment to endoscopic excision in selected lesions reduces perioperative morbidity. Additionally, endoscopic technique can be used in conjunction with open approaches to improve visualization. In view of the very high incidence of recurrent disease, prolonged clinical and radiological monitoring is necessary for all these patients. Primary radiation should be used when the morbidity of surgical resection is unacceptable.With regard to delayed complications, radiosurgery has less chance of risk than conventional radiation therapy theoretically. Further experiences and studies are required to confirm the use fulness of radio surgery on JNA.

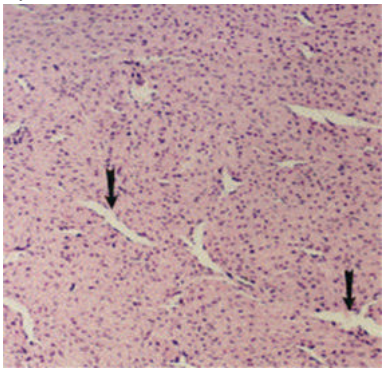

Figure1. Angiofibroma;staghorn-shaped Blood Vessels (arrows) With Endothelial Cells But No Smooth Muscle Are Compressed By Fibrous Tissue.

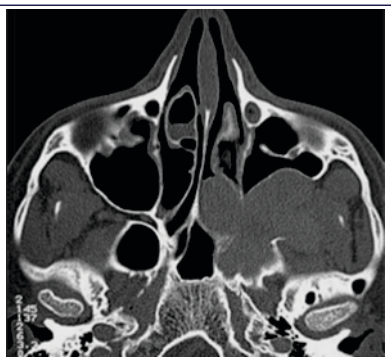

Figure2. Axialct; The Posterior Wall Of The Maxillary Sinus Has Been Bowed Anteriorly (arrowed)

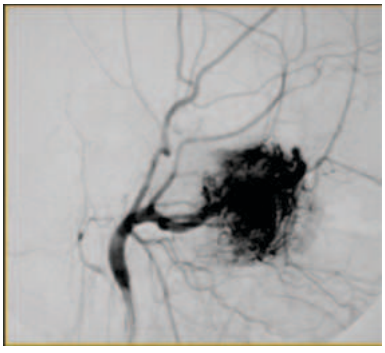

Figure 3. Angiogram; Blood Supply Of The Tumor From The Internal Maxillary Artery. Before Embolization.

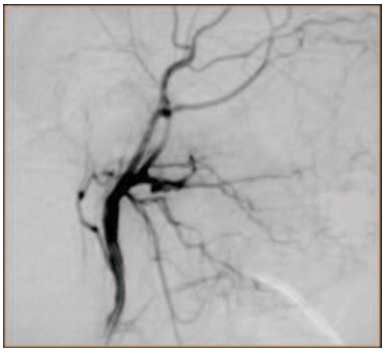

Figure 4. After Embolization Of The Internal Maxillary Artery.

RESULTS

Surgery is the gold standard of treatment for JNA. Treatment options for JNA include chemotherapy ,hormone administration, radiation therapy, embolization.Today, these modalities are used only as occasional complementary treatment (1,4,7-9,18-19).

Hormone Therapy

Hormonal stimulation appears to play an important role with regards to growth of JNAs (11). It has been proposed due to the androgen receptors associated with JNAs in an attempt to decrease tumor size and vascularity (21).Antiandrogenic agents like flutamide may reduce the growth rate of the JNA in vitro3,and in a small series of patients given flutamide, tumor shrinkage of upto 44 percent was reported by Gates et al (20). Estrogen has been shown to decrease size and vascularity of the tumor, but has feminizing side effects, variable response ,and risk of cardiovascular complications (11). Androgen receptor,other steroid receptors have already been detected (3,21) however, neither changes are found in the sexual hormones serum levels nor changes in the JNA patients sexual maturity.

Radiotherapy

External beam radiation is generally reserved for larger and/or unresectable tumors and tumors that are life-threatening due to their location (2,11,19). Studies by Briant et al. and Cummings et al. both reported an 80% control rate with moderate dose primary radio therapy (30 to 35 Gy given in 15 fractions over a 3 week period) (5,23). Longterm severe complications of radiation therapy are encountered including growth re-tardation, panhypopituitarism, temporal lobe necrosis, cataract, and radiation keratopathy (2,5,23). Also others reported remote secondary malignancy of the head and neck such as thyroid carcinoma and sarcomas of bones of tissue (23) a serious adverse effect of radiation therapy on JNA.Dare etal. (26) recently reported two cases of successful GKS application in JNA as a booster treatment for residual tumor after surgical resection. It is not as invasive as to surgery that is almost free of acute complications in surgery including massive hemorrhage (2).

Embolization

JNA consists of proliferating, irregular vascular channels with in a fibrous stroma. Tumor blood vessels typically lack smooth muscle and elastic fibers, this feature contributing to its reputation for sustained bleeding. Blood supply is most commonly from the internal maxillary artery, but may also be supplied by the external carotid artery, the internal carotid artery, the common carotid artery, or the ascending pharyngeal artery. Preoperative selective arterial embolization of feeding vessels from the external carotid artery has significantly decreased intra operative blood loss and facilitated resection of larger tumors (Figure 3, 4) and rendered intra operative or post operative blood transfusions unnecessary (18).

The ideal time between tumor embolization and the surgical procedure should be around 24 to 72 hours (27). Many papers also point out the benefits of preoperative arterial embolization in controlling intraoperative bleeding (1,4,5,9,10,27), reducing it by as much as fifty percent, thus allowing the full excision of the tumor (9,10) ,lower relapse rates and neurological complications (28) ,and reduced surgery duration. However some authors consider preoperative embolization to provide no benefit ,or even to increase the risk of recurrence (22). Complication of preoperative embolization rates may be as high as 20% (29). Such complications may include occlusion of the central retinal artery and consequent temporary blindness ,oronasal fistula due to tissue necrosis, occlusion of the middle cerebral artery followed by stroke,and occlusion of the ophthalmic artery (10,27,29).

Surgery

Open Surgical Approach

Open surgical approaches are trans oral, trans facial, and combined craniofacial approaches (5) (more specifically transpalatal, transantral, transnasal, lateral rhinotomy, midfacial degloving, Le Fort 1 osteotomy, and infratemporal fossa approach). Lateral rhinotomy approach has the advantage of access to the nose, maxillary antrum, ethmoids and nasopharynx, pterygopalatine fossa (8).However it leaves an external scar and removal of nasal and facial bones in prepubertal boys could lead to facial asymmetry (8). Persistent nasal crusting, facial paresthesia, lacrimal apparatus injuries are also seen in this approach. Midfacial degloving approach provides good exposure to the maxillary antrum,nose,pterygopalatine fossa and infratemporal fossa. There will be no deforming scar on face because of the use of a sub labial incision,but needs extensive removal of bones from the anterior , posterior, medial and lateral walls of maxillary antrum (8) .Transpalatine approach provides access to the nasopharynx, sphenoid, sphenopalatine foramen and posterior nares.It avoid external scar and does not effect the facial growth but oronasal fistula is a more common side effect (8). This approach was selected in the majority of patients with advanced disease by Cansiz etal (30) . Open approaches are favored by some surgeons and remain indicated for larger JNAs (11). Additional all open approaches can be supplemented by the use of endoscopes.

DISCUSSION

Improvements in endo nasal technique and knowledge of the intranasal anatomy enable the use of nasal endoscopic surgery to remove some JNA tumors.The decision to perform JNA resection endoscopically should be based on the experience and skill of the surgeon as well as the extent of the tumor (7-10,17) . Endoscopic sino nasal surgery and embolize these tumors preoperatively is to allow atraumatic dissection, minimizing bleeding and more precise tumor resection, thus minimizing the possibility of residual tumor (9,17,28). Patient selection for endoscopic resection is of paramount importance for a successful outcome. It has been suggested that tumors involving the ethmoid, maxillary, or sphenoid sinus, the sphenopalatine foramen, nasopharynx ,pterygomaxillary fossa and have limited extension into the infratemporal fossa are amenable to endoscopic resection (9-10,17,27-28).Onerci etal. reported no recurrence for tumors extension in to infratemporal fossa(7,31).

Endoscopic surgery has a great advantage because it preserves both the anatomy and physiology of the nose,requires less rehabilitation days after surgery, requiring less days of hospitalization and is less subject to hospital infections (8). The"push-pull"technique involves an incision of about 1 inch in the upper buccogingival sulcus,one more step in the evolution of endoscopic surgery for JNA (17,32). It is now possible to excise tumors with large lateral extensions in the infratemporal and parapharyngeal regions without resorting to an open approach.

Despite technological and surgical advances the recurrence rates have been reported between 30 and 50%(5,11), most symptomatic recurrences occurring during the first 12 months after primary treatment (11).Unfortunately, recurrent tumors are often reactivated residual tumors (5,24-25). The primary factor affecting recurrence was found to be the tumor stage at the time of diagnosis (11). The younger the patient the more likely that future recurrence will develop. Notsurprisingly, recurrence is more likely in patients with advanced disease.

Immunohistochemistry

SMA, HHF35 / h-caldesmon are positive in smooth muscle wall of large vasculature and CD31, CD34 and ERG highlight endothelial cells of vasculature whereas AR is positive in stromal cells in 40 - 75% cases; the expression may be focal and weak. Reported positive rate of ER is highly variable, ranging from 0% to over 90% partially depending on the subunits stained Beta catenin: nuclear staining in stromal cells (fibroblasts) in 70 - 90% of cases, membranous / cytoplasmic staining in endothelial cells

CONCLUSION

During this study we were able to conclude that case based approach in Juvenile nasopharyngeal angiofibroma is required in diagnostic and therapeutic conundrum. We thank the staff of departments of Otorhinolaryngology, Opthalmology and Pathology of AIMS Dewas and departments of Pathology and Microbiology of RDGMC, Ujjain. Whose role in this diagnostic and therapeutic study was impeccable.

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