Imaging in Juvenile Nasopharyngeal Angiofibroma: Clinical Significance of Ramharan and Chopstick Sign

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Abstract Imaging plays an important role in the diagnosis, staging and prognosis of JNA. Certain radiological changes as seen on CECT were observed to be consistent in our case series. This study analysed preoperative and postoperative CECT of large series of JNA patients to evaluate the sites and pattern of spread of tumor. We evaluated the clinical significance of pterygoid wedge in preoperative and postoperative imaging and thus elucidating two new radiological signs. Retrospective analysis of the pre operative and post operative imaging data of 242 patients with JNA. The findings in the scan were clinically correlated with the endoscopic intraoperative findings. Preoperative evaluation of the pterygoid wedge revealed widening on the involved side in 99.1% of our cases which is 1.8 times greater compared to the uninvolved side. The possibility of residual/recurrent tumor was found to be significantly higher in those where the pterygoid wedge was not

removed by drilling ($p < 0.001$) Drilling of the pterygoid wedge intra operatively, reduced the rate of residual/recurrence from 31.9 to 3.07%. Widening of the pterygoid wedge seen in the preoperative CECT, referred as RAM HARAN sign occurs in JNA. It has a significant diagnostic value as a radiological sign in JNA. Drilling of the pterygoid wedge intraoperatively reduces the rate of recurrence of JNA. Appearance of the two pterygoid plates on post-operative CECT, as two parallel lines, referred as Chopstick sign, has a remarkable prognostic value as a radiological sign in JNA.

Keywords Juvenile nasopharyngeal angiofibroma · JNA · Pterygoid wedge · Ramharan sign · Chopstick sign

Introduction

Technological advancements have created refined diagnostic techniques, providing insights in tumor behaviour which has revolutionized management. The abnormalities can be detected before signs and symptoms appear. And early detection of lesions helps to understand sites of origin. Increased use of radiology has led to increase in therapeutic effectiveness in otorhinolaryngology. Owing to vascular nature and possible intracranial extension, imaging is the primary modality of investigation for many lesions of the skullbase.

JNA is a formidable, locally aggressive, benign tumor of nasopharynx that spreads submucosally. It occurs almost exclusively in adolescent males and forms 0.05% of all head and neck neoplasms [1, 2]. This decade has witnessed a new armamentarium of diagnostic techniques that have revolutionized the management of JNA. Not only are advances in imaging changing the perspectives on the

Availability of data and material master chart uploaded as additional supporting material.

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behaviour of disease, but also on treatment strategies in routine practice. Preoperative radiological evaluation of JNA plays a crucial role in its diagnosis and management.

Computed tomography Scan with contrast enables accurate diagnosis without resorting to the dangers of biopsy. CECT is reliable for mapping tumor extensions into skull base and bony erosion. All staging systems for JNA rely upon tumor extensions on preoperative imaging for its classification. This emphasizes the importance of imaging in staging and thereby determining prognosis.

Imaging provides certain unique differential diagnostic feature that indicates presence of specific pathologies. The signs add confidence to the diagnosis of certain conditions [3, 4]. Implementation of a radiological sign in diagnosis is only possible after knowing precise cause and exact appearance. We propose two such radiological signs pertinent to JNA as they appear on pre and post operative CECT scan.

We retrospectively analysed pre and post-operative imaging data of 242 primary cases of JNA for sites involved and pathway of spread. Based on observations pertinent to tumor extensions and spread we propose a novel site of inception for JNA.

Detailed evaluation of specific bony area at the junction of the medial and lateral pterygoid plates, also known as the pterygoid wedge was done. This paper evaluates significance of pterygoid wedge widening as an objective tool to establish diagnosis of JNA. The postoperative scans after drilling of pterygoid wedge in all cases of JNA were also evaluated for residual tumor.

Methods

A retrospective analysis was conducted on pre and post-operative CECT scans of 242 primary cases of JNA evaluated and operated at our centre from January 2005 to January 2014. Written informed consent was obtained for participation from all the patients and from the guardians in cases of minors. All patients were evaluated with CECT preoperatively. Intravenous iodinated contrast medium was used. Pre and post contrast studies were done with 0.625 mm thick intervals, coronal and axial sections were studied in two different settings (windows):

1. Soft tissue intermediate window to see for avid contrast enhancement of Nasopharyngeal Angiofibroma in axial and coronal planes.
2. Bone window to assess the expansion and erosion of bone in axial and coronal planes.

MRI was performed only when CECT showed intracranial extension of tumour or engulfment of the internal carotid artery.

CECT was repeated post-operatively after 36 h to assess for residual tumour. The patients who received alternative therapy, recurrent/residual tumours and those who underwent radiotherapy due to faulty diagnosis of malignancy were excluded from study.

Results

Demographics

242 JNA patients were evaluated and operated in our department from January 2005 to January 2014. All patients were males. The mean age was 17.33 years (10–25 years). All the cases were histopathologically confirmed post operatively.

Preoperative Imaging

Preoperative imaging data for 242 JNA cases was retrospectively studied. Involvement and widening of the pterygoid wedge on the diseased side was seen in almost all the primary cases (99.1%). Pterygoid wedge is defined as anterior junction of the medial and lateral pterygoid plates [5]. The thickness of the pterygoid wedge was calculated as the distance between the medial border of the medial pterygoid plate to lateral pterygoid plate. This characteristic widening of the pterygoid wedge was observed in our cases consistently and was coined as “RAM- HARAN” sign. (named after the authors Dr. Janakiram and Dr. Bavaharan).

New Observation

In the analysis we observed that in three of our cases, the only involved area by the tumor was the pterygoid wedge with or without extension to the sphenoid sinus. Here other areas like the sphenopalatine foramen, nasopharynx or the vidian canal remained uninvolved. This was confirmed endoscopically during the surgery.

It was observed that, the width of pterygoid wedge of the involved side was greater than the uninvolved opposite side. The average width of the involved side was about 1.8 times greater than that of uninvolved side (Fig. 1).

Furthermore, scans were analysed for tumor extension following the pterygoid wedge and floor of sphenoid sinus was the other most commonly involved site. The vidian canal was seen separate in 19% of the cases.

The pterygopalatine fossa and nasopharynx were involved in 95.8 and 98.4% of cases respectively. Orbital extension through inferior orbital fissure was seen in 25.6% cases. Pterygoid fossa was involved in 21.9% of patients. The greater wing of sphenoid was involved in 45.8% of

Fig. 1 Coronal CECT scan images, showing the contrast enhancing mass seen widening the pterygoid wedge compared to the uninvolved side (RAM HARAN sign)

patients. In 2.1% of patients the tumor extended from the pterygoid wedge along the greater wing of sphenoid into the middle cranial fossa. The quadrangular space was involved in 30.1% cases. The tumours extending into quadrangular space were lying anterior to the anterolateral wall of cavernous sinus. Involvement of cavernous sinus was rare and seen only in six cases. The most common part of internal carotid artery involved was the paraclival ICA. The involvement of various sites is given in Table 1.

Post Operative Imaging

Early postoperative evaluation included a CECT scan done after 36 h of surgery to identify any residual tumor. No contrast enhancement was seen in the sinonasal cavity in 92.2% of cases suggesting complete removal of tumor. Residual tumor was observed in 19 cases, the most common site being pterygoid wedge (57.8%; Table 2). In case

of residual tumor patient was taken up for second stage revision surgery as soon as the patient was deemed fit for general anesthesia.

Recurrent disease was defined as tumor seen subsequently after a negative postoperative scan. Post-operatively, a CT scan was done every 6 months for the next 2 years. The most common site of recurrence was the pterygoid wedge (45.4%) followed by the lateral aspect of Infratemporal fossa (18.1%), Cavernous sinus (9%), intracranial (4%). Observations in the initial cases revealed pterygoid wedge to be the most common site of residual/recurrent tumor. The observation was followed up in CT and addressed endoscopically during surgery, tumor was identified in pterygoid wedge and drilling of the wedge was done in rest all cases. This practice to drill the cancellous bone in the pterygoid wedge resulted in the appearance of the two pterygoid plates as parallel lines on postoperative CECT, which resemble CHOPSTICKS. This was a

Table 1 Various sites of involvement of JNA in our study

Site	Number
Pterygoid wedge	241
Sphenoid sinus	240
Nasopharynx	239
Nasal cavity	232
Medial pterygopalatine fossa	5
Complete pterygoalatine fossa	227
Infra temporal fossa	217
Cheek	27
Temporal fossa	2
Pterygoid fossa	53
Inferior orbital fissure	100
Orbit	61
Vidian canal	46
Lateral recess	237
Greater wing of sphenoid	110
Quadrangular space	73
Meckel's cave	11
Middle cranial fossa	28
Superior orbital fissure	39
Cavernous sinus	37
Internal carotid artery	11
Parapharyngeal space above the lower border of mandible	5
Intracranial intradural extension	5

Table 2 Sites of residual and recurrent tumor

Site	Recurrence	Residual
Pterygoid wedge	10	11
Infratemporal fossa	4	
Cavernous sinus	2	2
Superior orbital fissure	2	1
Middle cranial fossa	1	1
Greater wing of sphenoid	1	
Quadrangular space	1	1
Internal carotid artery	1	1
Inferior orbital fissure		2

constant finding in all the cases where pterygoid wedge was drilled (Fig. 2).

Discussion

Multiple modalities have been offered for the management of JNA, however most authorities agree that surgical management is the treatment of choice. Increased

understanding in the endoscopic anatomy, complemented with new observations in imaging and expansion of surgical corridors have made endoscopic excision of JNA as a preferred surgical approach.

Imaging has been an indispensable part in management of JNA. Preoperative imaging is the primary diagnostic as well as prognostic tool for this tumor. It allows for accurate assessment of the tumor extent and appropriate surgical planning. Involvement of various areas as seen on preoperative imaging forms the basis for all the staging systems for JNA. Diagnosis of Juvenile nasopharyngeal angiofibroma is based on history, clinical examination and imaging studies. Nasopharyngeal angiofibroma presents with typical radiological signs that aids in preoperative diagnosis without subjecting the patients to the perils of a biopsy [6]. Lloyd et al. [6] reported two characteristic features suggesting a diagnosis of JNA, a soft tissue mass in the posterior nasal cavity and pterygopalatine fossa with erosion of posterior osseous margin of the sphenopalatine foramen extending to the base of the medial pterygoid plate. The finding of the anterior bowing of the posterior wall of the maxillary sinus on plain X-ray, was described by Holman and Miller as 'antral sign' (Holman Miller sign). Antral sign was found in 87% of JNA patients in Holman-Miller et al. [7] and in 81% of patients with JNA in Lloyd and Phelps et al. [8]. However, Schaffer et al. (1978), described two cases of trigeminal nerve neurofibroma with positive antral sign. Thus any slow growing benign tumor causing long term pressure on the posterior wall of the maxilla is bound to cause this bowing effect [8]. Thereby, antral sign cannot be pathognomonic for JNA.

In retrospective analysis of preoperative CECT of 242 primary JNA cases at our centre, it was observed that the pterygoid wedge, at the anterior junction of the lateral and the medial plates, was consistently involved by nasopharyngeal angiofibroma. The pterygoid wedge was found to be involved in 99.1% (241) of our cases. Another interesting finding noted in these cases was the widening of the pterygoid wedge, approximately 1.8 times greater than that of the uninvolved side. Lloyd and Phelps [8] noted a deformity of the base of the medial pterygoid lamina in CECT of all of their cases. We describe this unique observation of widening of the pterygoid wedge as RAM-HARAN sign (named after the authors Dr. Janakiram and Dr. Bavaharan). However this sign may not be seen in cases where the pterygoid wedge has already been eroded preoperatively by the tumor.

The pterygoid wedge widening was consistently seen in 99.1% (241) of our cases, including earlier stages of the disease. This was an unswerving feature seen even in the cases where sphenopalatine foramen was not involved (Fig. 3). It has been hypothesized that JNA grows along the pathway of least resistance from its site of origin in the

Fig. 2 Coronal post operative CECT images, showing the dysjunctioned pterygoid plates as two parallel lines (Chopstick sign)

sphenopalatine foramen [8]. However, we contend this aspect based on our understanding that if there were to be a least resistant pathway in the skull base it should be through the nasopharynx towards the oropharynx. However, extension into tough bony areas studded with fissures and canals was a frequent observation in our study. This has led to our belief that the tumour possibly arises intradiploic within the cancellous bone of the pterygoid wedge. Furthermore it spreads along the perforating arteries in the Haversian canals and extends along vascular paths. Thus pterygoid wedge as a site of origin fits with the known extensions of the tumor and the earliest bone changes visible, as described by RAM- HARAN sign. Hence we postulate that the cancellous bone of the pterygoid wedge could be the site of origin of the tumour. Sites such as sphenopalatine foramen and pterygoid canal have been consistently seen in medical literature as the potential site of origin. On detailed analysis of the imaging studies, it was observed that the epicenter of the tumor was at the

pterygoid wedge in all our cases. Pterygoid wedge was involved in all cases in our series, even in the earliest of stages where other proposed sites of inception remained uninvolved. This finding was confirmed intraoperatively. Based on this observation we propose pterygoid wedge to be the most consistent and probable site of origin.

Addressing the pterygoid wedge is crucial intraoperatively (Fig. 4). A reasonably high residual/recurrence rate (31.9%), was noted in the pterygoid wedge in cases where it was not removed in primary surgery. Complementary to this observation, the residual/recurrence rates in this area dropped significantly (3.07%), when pterygoid wedge was drilled and removed. ($p < 0.001$; Table 3). This statistically significant change in the residual/recurrence rate cannot be attributed to any other factors like the disease stage or age of patient. The possibility of residual/recurrent tumor was significantly higher in those where the pterygoid wedge was not removed by drilling by Fischer's exact test $p < 0.001$. Diligent extirpation of the JNA by drilling the

Fig. 3 Coronal CT image showing widening of the pterygoid wedge on the involved side. Note the normal pterygoid wedge on the uninvolved side

cancellous bone of the pterygoid wedge is pivotal in avoiding recurrence. The absence of pterygoid wedge in the post-operative coronal CECT, is depicted by the appearance of dysjunctioned pterygoid plates, which resemble chopsticks, described by senior authors as Chopstick sign. However this sign may not be seen in situations where the tumor has already eroded the pterygoid plates preoperatively or where they have been drilled to access the tumor that spreads posterior to the plates.

Conclusions

Widening of the pterygoid wedge is caused by its involvement by Juvenile Nasopharyngeal Angiofibroma. Widening of pterygoid wedge as revealed in preoperative imaging, RAM-HARAN sign, is of significant diagnostic value in all cases of JNA. Drilling of the wedge intraoperatively significantly reduces the recurrence and improves the surgical outcome. The senior authors suggested the

Fig. 4 Intraoperative pictures showing the drilling (a, c) and removal of the tumor remnant (b, d) from pterygoid wedge in two separate cases

Table 3 Association between drilling of the pterygoid wedge and residual/recurrent JNA

	Recurrence/ residual in pterygoid wedge	No recurrence/ residual in pterygoid wedge	Total
Non drilled	15	32	47
Drilled	6	189	195
Total	21	221	242

term ‘Chopstick sign’ may be considered to describe this finding in post-operative CT. Owing to most consistent involvement even in earliest stage of disease, Pterygoid wedge is proposed to be the site of origin for JNA.

Author Contributions TNJ is the chief operating surgeon and major contributor to the inception and realization of the two radiological signs. SB has a major role in composing of the manuscript. UCS has a contribution in preparation of manuscript, images and supporting material. OD has analysed and interpreted statistical data and contributed to manuscript preparation. BR is the radiologist and has analysed the radiological imaging data. VS has a role in preparation of manuscript. All authors read and approved the final manuscript.

Compliance with Ethical Standards

Conflict of interest none.

Ethical Approval The article does not contain any studies with human participants or animals performed by any of the authors.

Informed Consent Written informed consent was obtained from all the individual participants included in the study for publishing data.

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