Juvenile ‘Perinasal’ Angiofibroma

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Abstract The extranasopharyngeal angiofibroma is a separate clinical entity but those involving infratemporal fossa and cheek resemble juvenile nasopharyngeal angiofibroma (JNA) and hence have been labelled as juvenile perinasal angiofibroma (JPA) in this paper. This paper presents a 7th case of JPA and attempts to review the world literature on JPA, along with a proposal of staging the disease. A 16 year male presented with a painless compressible facial swelling since 7 months without any epistaxis or nasal obstruction. Initially a vascular lesion was suspected but JNA without nasal extension was strongly suspected on imaging. A deep trucut biopsy confirmed the histopathology. The vascular enhancement was significant and the tumour was excised through open approach (Weber Fergusson). JPA that can be regarded as a variant of JNA that fails to extend medially. Imaging demonstrates classical JNA findings with a clear nose/nasopharynx. A deep trucut biopsy under control in inpatient settings may sometimes help. JPA presents most commonly in Stage II where an open facial approach preferably following selective preoperative embolization is indicated. Hence with painless compressible (or non-compressible) cheek swelling suspected to be of a vascular etiology, a high degree of clinical suspicion for JPA needs to be maintained in order to prevent a misdiagnosis.

Keywords Extranasopharyngeal · Angiofibroma · Infratemporal fossa

The occurrence of extranasopharyngeal angiofibroma (ENA) is extremely rare and till 2013 only 73 cases have been reported. No specific classification exists for this heterogeneous group owing to a wide variation in origin, presentation, management and prognosis. The behaviour of angiofibroma involving the nasopharynx (JNA) is quite different from the majority that occur at the extranasopharyngeal sites (ENA). Although bleeding is supposedly less in ENA but the other parameters such as rapidity of growth, recurrence, morbidity and mortality have not yet been standardised owing to very small number of cases reported till date at different sites. There is a specific subset of ENA that presents exclusively along the lateral extension pathways of classical juvenile nasopharyngeal angiofibroma (JNA) without involving the nose or nasopharynx and hence do not present with either nasal obstruction or epistaxis. Only 6 such cases have been reported in the literature so far. We present the 7th such case with some other peculiarities. This subset may better be labelled as the Juvenile Perinasal Angiofibroma (JPA) since it behaves almost similar to JNA in terms of vascularity, intraoperative bleeding, surgical management, recurrence potential, morbidity and mortality. Although many authors regard ENA as a separate clinical entity from JNA, we suggest JPA as a possible variant of JNA that has failed to extend medially. This paper presents a unique such case and attempts to review the world literature on JPA, along with a proposal of staging the disease.

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Case Report

A 16 year old male patient presented with a cheek mass causing disfigurement of face since 7 months. The history was characterised by the absence of nasal obstruction and epistaxis. The physical examination revealed cheek fullness with mass protruding into the upper gingivobuccal sulcus (Fig. 1). The patient was operated for cleft lip at the age of 8 years and cleft palate at the age of 12 years. There was overcrowding of the anterior teeth while the hard palate showed evidences of malformation. It appeared that the premaxilla did not fuse with the main palate on the right side while a slight fissuring on the right side of the upper lip corresponded to the earlier cleft deformity. In addition the patient also had polydactyly in the right hand.

A small degree of squint was also appreciated on cover-uncover testing. The hearing functions were within normal limits while nasal apertures appeared somewhat narrow. The posterior rhinoscopy revealed adenoids while indirect laryngoscopy was normal. Noteworthy was the fact that the cheek swelling was compressible (Fig. 2) without any bruit on auscultation. Aspiration of the swelling with a wide bore needle revealed frank blood only. A diagnosis of some vascular mass/malformation was suspected clinically.

The angiography and imaging (Fig. 3) revealed a highly vascularised lesion predominantly supplied by the internal maxillary artery, involving sphenopalatine foramina on the

right side extending well laterally across the pterygopalatine fossa, infratemporal fossa and cheek, indenting the posterior wall of maxilla (Hollman Millar sign). None of the paranasal sinuses were involved and nose/nasopharynx were clear.

A radiological diagnosis of JNA was evident but dilemma continued owing to its fully compressible nature as well as its extranasopharyngeal location with nasal cavity being spared. Hence a trucut deep biopsy was undertaken sublabially that revealed a histopathologic picture of angiofibroma. In the absence of preoperative selective embolization facility at our centre the tumour was excised elsewhere via an open facial transmaxillary approach (Fig. 1). No recurrence was seen till 2 years.

Discussion

ENAs are considered to be a separate entity from JNA and most commonly originate in the maxillary sinus. The other reported sites include ethmoid and sphenoid sinuses, nasal septum, middle and inferior turbinates, conjunctiva, molar and retromolar region, and larynx. Till date only 7 cases of ENAs (involving pterygopalatine fossa, ITF, cheek) have been reported and they seem to have a close resemblance with JNA. Such JPAs are extremely rare and are reported only in males with age 13–23 years. In contrast other

Fig. 1 Preoperative picture showing fullness of cheek and upper gingivobuccal sulcus (*left*). Postoperative picture 6 months after surgery (*right*). Note the evidences of incision line scar corresponding to the open facial approach

Fig. 2 Compressibility of cheek swelling. The *left figure* shows compressibility and the *right figure* shows expansile nature after 2 s of removal of compression

Fig. 3 Imaging (*left*) and angiography (*right*) showing a highly vascularised lesion predominantly supplied by the internal maxillary artery, involving sphenopalatine foramina on the right side extending well laterally across the pterygopalatine fossa, infratemporal fossa and cheek, with Hollman Millar sign. Note the clear nose and nasopharynx

Table 1 Global summary of 7 cases of ENAs presenting exclusively in infratemporal fossa (ITF) and cheek

References	Age	Sex	Sites	Surgery	History
Irby [5]	M	21	Right cheek, parapharyngeal space, infratemporal fossa (ITF)	Transoral biopsy and resection; bleeding requiring 2 blood transfusions	Premaxillary swelling 1 year
Hora and Weller [6]	M	21	Right cheek, infratemporal fossa, temporal fossa, pterygomaxillary and parapharyngeal space	Open approach	Swelling of the cheek, pain 7 months
Whitlock [7]	M	16	Left cheek, ITF	Open surgery	Cheek swelling
Isherwood [8]	M	13	Right pterygomaxillary fissure, infratemporal fossa	Transzygomatic resection with/ligation of the maxillary artery	Painless facial swelling 3 weeks Decayed tooth extracted; biopsy due to prolonged healing from granulation tissue; repeated biopsy (transmaxillary); no bleeding
Gupta et al. [9]	M	13	Right ITF, pterygomaxillary fossa. Also invading the right orbit, sphenoid sinus and middle cranial fossa on the right side	Open approach by Weber-Fergusson extended sublabial incision up to maxillary tuberosity. Ipsilateral external carotid artery ligation	Facial swelling since 5 m, proptosis for 2 m and headache since 1 m. No history of epistaxis or nasal obstruction
Szymańska [10]	M	23	Left ITF, cheek and pterygopalatine fossa	Cervical approach with temporary mandible split	Painless cheek swelling since 12 m with extensive blush on CT scan
Present (2015)	M	16	Right sphenopalatine foramina, ITF, Cheek	Open surgery with Weber fergusson extended sublabial	Collapsible painless facial swelling since 7 m with past history of operated cleft lip and palate

ENAs in general develop in all the age groups and are more common in females. Table 1 summarises the reported cases in the literature. While ENAs in general lack typical vascularity and radiological features of JNA, JPA shows more or less a similar behaviour and clinicoradiological picture except for nasal involvement or related symptoms. Therefore JPA may be considered as variant of JNA.

The etiopathogenesis of JNA has been rather quite controversial. Handoussa et al. [1] felt that it is a true neoplasm arising from basisphenoid periosteum. Osborn [2] suggested that it either developed from remnants of fetal erectile tissue or alternatively was a true hamartoma. JNA has been proposed to be a hemangioma [3] a vascular

malformation [4]. The compressible nature with a frank blood on aspiration defends the theory of origin of JNA related to either incomplete regression of the first brachial artery [3] or being hemangiomatous entity or even as a result of abnormal differentiation of the nonchromaffin paraganglionic cells of the terminal end of the maxillary artery. With its origin in the pterygopalatine foramen/fossa the JPA may extend laterally in ITF and the cheek particularly when the coexisting bony ‘obstructions’ prevent its spread medially. Ours is the only case with the associated congenital deformity (maldevelopment of maxilla-palatine complex) and hence might have contributed to limitations in spread.

The exact site of origin of JNA is controversial. While the majority feel it to originate from the upper lip of the sphenopalatine foramen, others claim it to arise from the distal bony vidian canal. The site of origin of JPA may correspond to JNA while an absence of medial extension may be due to restricting anatomy as indicated earlier. It may have been possible for the other reported cases to have some coexisting occult malformation around the sphenopalatine foramina area that went unnoticed and precipitated the occurrence of JPA instead of JNA. It has been suggested that ENAs around maxilla may be caused by unequal development of skull bones. Although not supported by literature our case does reveal a maldevelopment of maxillo-palatine complex as evident by cleft palate. To the best of our knowledge a compressible JNA/JPA has not yet been reported in the literature. It may also be possible to have malformations in the vascular network of ITF as well as abnormal proliferation of arterial system manifesting as a compressible vascular swelling. This particular finding needs to be noted to avoid misdiagnosis for a hemangioma or a vascular malformation subsequently to be treated with prolonged sclerosants. Another interesting fact about JPA is its occurrence on the right side (5 of the 7 cases till date). Whether it is related to a more propensity for occult malformation to occur on the right side or otherwise is not yet known.

It is well established that surface biopsies can be misleading as the typical histological appearance is only seen internally. Our case too revealed only blood on superficial aspiration as the histopathology of JNA was concluded only on a deep trucut biopsy. It is not unsafe to obtain a deep trucut biopsy of the cheek mass sublabially if needed. Such a procedure may be carried out under local anaesthesia but always in inpatient settings and never as an office procedure. Following the biopsy procedure a deep pressure on the site is to be maintained for achieving hemostasis and subsequent icepack to prevent hematoma. Histopathologically the ENAs in general are more heterogenous with the absence of universal predominance of vascular components in the fibrous stroma. JPA however are more consistent to resemble JNA in this regard. Majority of the JPAs reported so far had undergone an open type of surgical approach particularly the Weber Fergusson incision with extension in the sublabium up to maxillary tuberosity depending upon the case. The intraoperative bleeding was comparable with JNA and 2 cases underwent external carotid ligation while a single one preoperative embolization.

With only 7 cases of JPA reported so far in the world literature an attempt is made to stage the disease based on the evidence and the complexities of surgery involved to deal with this extremely rare entity. Our proposed staging scheme for JPA is as follows:

Stage I	Involvement of SPF and ITF.
Stage IIA	Involvement of cheek with or without gingivobuccal sulcus.
Stage IIB	Involvement of temporal fossa and/or parapharynx.
Stage III	Involvement of orbit.
Stage IV	Intracranial extension.

The most common stage of the disease encountered so far is stage IIA while stage I is hardly detectable clinically. The extension in temporal fossa and stages III/IV have been appreciated in a single patient each. The role of an exclusive endoscopic resection for JPA seems to be limited at the stage of advanced presentation, but an endoscopically assisted open technique is likely to be adapted for JPA in future. Being primarily a surgical disease and a likely variant, the indications of radiation therapy and chemotherapy for JPA are expected to be the same as of JNA. With a better understanding and evidence the JPA is likely to evolve fast.

Conclusions

With painless compressible (or non-compressible) cheek swelling suspected to be of a vascular etiology, a high degree of clinical suspicion for JPA needs to be maintained to prevent misdiagnosis. Similarly a radiological diagnosis of JPA should always be considered in case of classical JNA findings with a clear nose/nasopharynx.

Compliance with Ethical Standards

Conflict of interest Anupam Mishra, Veerendra Verma, Subhash Chandra Mishra state that they have no conflict of interest.

Informed Consent Informed consent was obtained from the patient included in the study.

Human and Animal Rights All procedures performed on a single human participant was in accordance with the ethical standards of the institutional and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

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