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Case Report *Volume 6 Issue 1*

A Rare Case Report of Pleomorphic Adenoma Arising from Nasal Septum: As Scarce as Hen's Teeth

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Abstract

Pleomorphic adenomas (PAs) are the commonest benign tumors of major salivary glands. Interestingly, they can arise at any site of salivary gland tissue. Intranasal PAs are an extremely unaccustomed entity. Till 1981, only 6 Bonafide cases arising from the nasal septum had been documented. In such scenario of its rarity, available literature tending to be in the form of isolated case reports and a high potential of this tumor getting misdiagnosed, every case needs to be reported to facilitate a valid statistical analysis. We present a case of a 36 years old male who presented to us with complaints of left sided nasal obstruction of 6 months duration and was diagnosed to have PA arising from the nasal septum. We also present review of literature of intranasal PAs with emphasis on its clinical presentation, gross and microscopic appearance and surgical management.

Keywords: Pleomorphic Adenoma; Nasal Septum; Unilateral Nasal Mass; Intranasal Mixed Salivary Gland Tumors

Abbreviations

PA: Parathyroid Adenoma; PNSs: Para Nasal Sinuses; ORL: Otorhinolaryngological; DNE: Diagnostic Nasal Endoscopy; CECT: Contrast Enhanced Computed Tomography; HPE: Histopathological Examination.

Introduction

Salivary gland tumors account for approximately 3 % of all head and neck neoplasms [1]. 21.7% of all salivary gland neoplasms are malignant while majority of them being benign [2]. As high as 70% of the benign salivary gland tumors are reported to be pleomorphic adenomas (PAs) Eneroth CM [1]. PAs most commonly originate from major salivary glands (parotid and submandibular). However, in about 8% of cases, they have also been stated to affect minor salivary glands located in oral cavity, pharynx, hypopharynx, larynx, trachea, lacrimal glands, paranasal sinuses (PNS). Sporadic affection of nasal cavity has also been mentioned anecdotally [3]. We present an interesting case of PA arising from the nasal septum.

Case Report

36 years old male presented to our center with complaint of left sided progressive nasal obstruction of 6 months duration. No history of rhinorrhoea, epistaxis, hyposmia, nasal trauma or previous nasal surgery. No other otorhinolaryngological (ORL) complaints. On evaluation, there was no gross

external abnormality. Diagnostic Nasal Endoscopy (DNE) revealed midline septum with a solitary, smooth, greyish pink, fleshy mass ~ 2 cm X 1.5 cm arising from left side of anterior cartilaginous nasal septum completely obstructing the left vestibule (Figure 1a). It did not shrink in size after decongestion. Probe could be passed all around except medially suggesting attachment to septum. It did not bleed on touch except at its stalk. Posteriorly, it extended up to anterior third of inferior turbinate. Both osteo-meatal complexes and nasopharynx were clear. Rest ORL examination was within normal limits. Contrast enhanced computed tomography (CECT) of nose and PNS revealed a well-defined soft tissue attenuation mass lesion in left anterior nasal cavity likely arising from nasal septum measuring 21 X 14 X 17 mm without significant post contrast enhancement (Figure 1b).

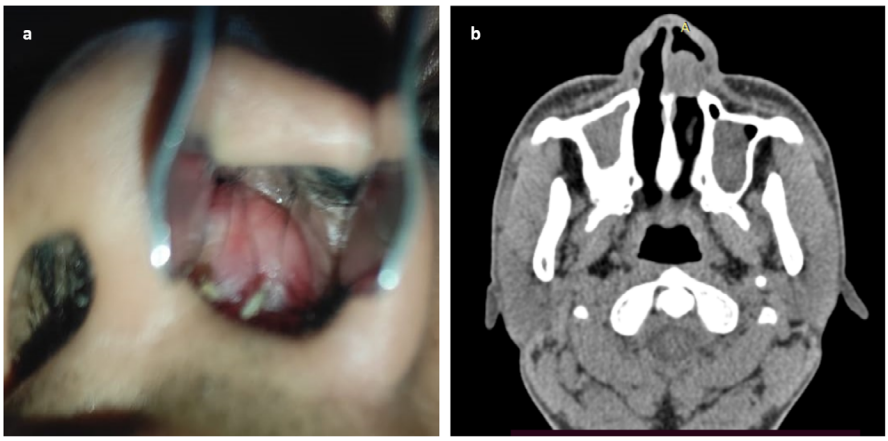

Figure 1a: Smooth, greyish pink, fleshy mass ~ 2 cm X 1.5 cm arising from left side of anterior cartilaginous nasal septum completely obstructing the left vestibule.
Figure 1b: HRCT Nose and PNS: Well-defined soft tissue attenuation mass lesion in left anterior nasal cavity likely arising from nasal septum measuring 21 X 14 X 17 mm without significant post contrast enhancement.

Since the mass was anterior and easily accessible, endonasal endoscopic excision biopsy was performed under local anaesthesia. With the help of bipolar cautery, mass was excised in toto along with a cuff of circumferential normal septal mucosa (Figure 2a). Specimen was sent for histopathological examination (HPE). Base of its attachment on the septum was also cauterised and haemostasis achieved. HPR showed stratified squamous epithelium lined tissue with sub-epithelium showing well circumscribed biphasic tumor. The stroma was myxoid and edematous comprising of spindle and stellate cells which was consistent with pleomorphic adenoma (Figure 2b). Patient has been on follow-up for 6 months now. He has been asymptomatic with no evidence of local recurrence.

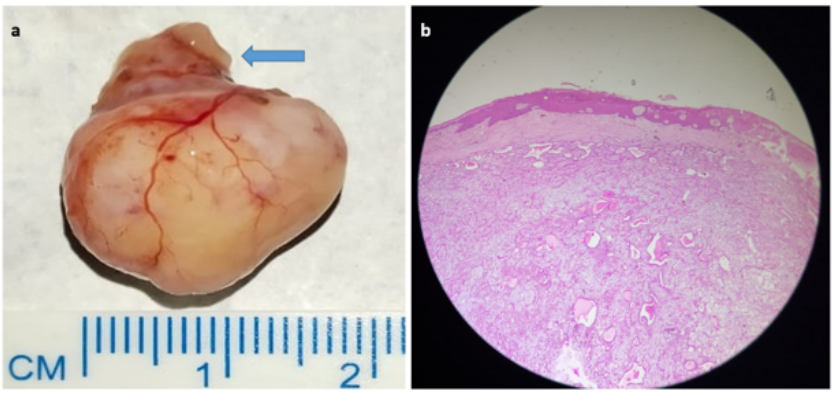

Figure 2a: Totally excised mass along with a cuff of circumferential normal septal mucosa (arrow).
Figure 2b: Low power microphotograph (10X)-Stratified squamous epithelium lined tissue with sub-epithelium showing well circumscribed biphasic tumor, comprising of glands and tubules lined by epithelial cells. The stroma is myxoid and edematous comprising of spindle and stellate cells. The epithelium appears melting into stroma. No atypia/ mitosis or necrosis seen.

Discussion

PAs are the commonest tumors of major salivary glands, predominantly the parotid (60%) [4]. Intranasal PA is extremely rare with its first ever published case dating back to 1929 [5]. Intranasal PA is an extremely unaccustomed and rare entity with available literature tending to be in the form of case series and isolated case reports. Therefore, every case deserves notification to facilitate a valid statistical analysis and prognostication. They are slow growing tumors which originate from the septal mucosa despite the anatomical location of salivary glands in the lateral nasal wall and turbinates [6].

Intranasal PAs predominantly present between 30 to 60 years of age with slight female preponderance. Clinical symptomatology typically includes unilateral nasal obstruction (71%) and epistaxis (56%). Other aberrant symptoms include intermittent mucopurulent rhinorrhoea, epiphora, nasal swelling/ deformity or a visible nasal mass [7]. In our reported case, the patient is a male in his 4th decade of life who presented only with left sided nasal obstruction.

The nasal mass usually varies greatly in size (0.5 cm – 7 cm), has smooth surface with broad-based attachment, does not bleed on probing and does not infiltrate the surrounding tissue [3]. Grossly, it often appears polypoid, soft, fleshy with a blue-gray translucency [3]. In our case, the mass was consistent with these features except that it bled when probed near its stalk and did not have a blue-gray translucency, rather prominent blood vessels were seen on its surface.

Quite a few differential diagnoses can be offered for a mass arising from the nasal septum. These include leiomyoma, osteochondroma, transitional cell papilloma, melanoma, adenoid cystic carcinoma and squamous cell carcinoma. Majority of these tumors originate from the septal mucosa. One must also consider nasoseptal swell body (discrete erectile tissue in submucosa over anterior nasal septum) especially for a small septal mass presenting as a doubtful lesion [8]. We have included a tabular comparison of nasal PA with other nasal tumors like papillomas, schwanommas, adenocarcinoma along with their characteristic imaging features (Table 1).

Differential diagnosis of intranasal tumor	Characteristic imaging/ manifestation	Remarks
Inverted Papilloma	Lobulated soft tissue mass along lateral nasal wall and middle meatus region with cerebriform appearance with occasional calcifications	
Squamous cell carcinoma	Soft tissue mass with prominent bone destruction. Smaller lesions are homogenous in signal intensity while larger lesions are more heterogenous with areas of necrosis and hemorrhage	Skull base erosion with invasion into anterior, middle cranial fossae and dural extension is best seen on contrast-enhanced fat suppressed T-1 weighted Magnetic Resonance Imaging (MRI)
Adenocarcinoma	Similar to and indistinguishable from squamous cell carcinoma	
Schwannoma	Subtle hyper-attenuation on CT (Antoni A tissue type- more cellularly dense); MRI-T1 hypointensity, T2 hyperintense with robust post-contrast enhancement	Features of bone remodeling are present and peripheral ring like enhancement is seen if cystic changes are present

Table 1: Tabular comparison of nasal PA.

CECT nose and PNS is the investigation of choice to work up any unilateral nasal mass. It appears as a smooth, localized lesion causing smooth bony expansion implicating a benign and slow etiological process as opposed to bony destruction which is seen in malignancies [10]. In our reported case, imaging revealed a well-defined soft tissue mass lesion arising from nasal septum without significant post contrast enhancement.

HPE clinches the diagnosis of PA. They have a characteristic lobular architecture with chondromyxoid stroma. Although histological architecture of intranasal PAs is similar to those of major salivary glands, they show unusually high cellularity with predominance of myoepithelial elements accounting for its relatively low recurrence (10% compared to 50% recurrence rates of parotid mixed tumors) and biologic aggressiveness. Also intranasal PAs have high epithelial, low stromal components and lack a capsule compared to their major salivary gland counterparts. All these features lend them a resemblance to malignant mixed tumors making the diagnosis even more challenging. Since there exists a tendency to get misdiagnosed especially at early stages, early

correct diagnosis is critical as misdiagnosis could potentially lead to mismanagement [4,7].

Immunohistochemistry is similar to that of parotid gland. Occasionally, PA may behave in a malignant fashion. Commonest such variant showing metastatic potential is carcinoma ex PA which frequently spreads to bone, lungs, regional lymph nodes and liver. Compango, et al. reported a malignant transformation rate between 2.5-10% [7,8]. In our reported case, the HPR findings were consistent with the available literature on intranasal PAs.

Surgery is the primary modality of treatment as it offers instant alleviation of symptoms and clears the tumor which has a chance of malignant transformation. Wide local excision with histologically clear margins is recommended to prevent recurrence. As per the size and location, feasible surgical approaches are endoscopic endonasal (for small tumors), lateral rhinotomy, midfacial degloving approach and partial maxillectomy. En bloc resection is also recommended to rule out microscopic recurrence [11]. In our reported case, the mass was excised in toto with clear margins endoscopically through endonasal route.

The authors believe that the prospects of intranasal mixed tumors are better than those in other ectopic sites, because they produce symptoms early facilitating a timely diagnosis. Another likely reason of early presentation is the tendency to originate anteriorly wherein the mass gets easily identified even on anterior rhinoscopy. The affection of surrounding bone is rare as the lesion has plenty of space to grow inside the nasal cavity. We have collated the data available from two largest case series on intranasal PAs (Table 2) [7,11].

Case series	Symptoms	Endoscopic findings	CT findings	Treatment offered	Follow up
Compogno et al. (40 cases)	Unilateral nasal obstruction (n=15), mass in nasal cavity (n=15), episodic epistaxis (n=6), asymptomatic (n=1)	Exophytic, polypoid, oval, dome-shaped/ irregular, firm gray mass or nodule with overlying translucent membrane/ mucosa	Expanding intranasal soft-tissue lesion causing bony destruction arising from cartilaginous/bony septum (n=25), lateral nasal wall (n=8)	Total wide surgical excision encompassing septectomy, turbinectomy via lateral rhinotomy or Caldwell-Luc procedure	Average follow up period 7.5 years (1-41 years), no recurrence (90%), recurrence (3 cases), 1 patient refused definitive surgery.
Rha M-S et al (2018) (7 new cases with review of 101 patients)	Unilateral nasal obstruction, epistaxis, intranasal mass, cheek swelling, sinusitis	-	-	Endoscopic endonasal, lateral rhinotomy, partial maxillectomy, midfacial degloving, external approach, combined approach	Average follow up period- 29.1 months (1-240 months), no recurrence (91.1%), locoregional recurrence (7.9%), distant recurrence (1%)

Table 2: Two largest case series on intranasal Pas.

Conclusion

We conclude that PAs are rare and unique tumors affecting the nasal cavity. Owing to their increased myoepithelial and lesser stromal constitution as compared to those arising from major salivary glands, intranasal PAs have a likelihood of getting misdiagnosed at early stages which might result in institution of a more radical and aggressive management. Hence, it is pertinent to know these differences. Also, despite its rarity, PA should be considered as a differential diagnosis for a unilateral nasal mass especially if the patient presents with unilateral nasal obstruction and/or epistaxis. Clinical follow up after surgical excision with routine DNEs is essential. We advise an adequate follow up period as deemed suitable by the surgeon keeping in mind the recurrence potential, risk of malignant transformation and ability to metastasize especially when clear margins are not reported on HPE.

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