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Ethmoidal Sinus Schwannoma: Case Report and Review

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Abstract: ***Aim: To highlight the need for benign nasalshwannoma to be included in any differential diagnosis of any soft tissue mass of the sinonasal spaces. Methodology/ Case Description: A 45 year old female presented with Right sided nasal obstruction for 1 year along with mouth breathing, with no history of anosmia or hyposmia, epistaxis, difficulty in breathing. On examination, Anteriorrhinoscopy right side nasal polypoidal mass, Examination of Eye and Oral cavity found to be normal. On nasal endoscopy right side nasal polyp extending to posterior choana occupying the right nasal cavity with deviated septum towards left side. CT PNS shows findings of polypoidal lesion noted at right sided ethmoidal air cells and right nasal cavity extending posteriorly up to right side nasopharynx. Intraoperatively polyp identified and removed as a piece meal and sent for HPE. Histopathology suggestive of schwannoma with spindle cell proliferation. Follow up Nasal endoscopy shows no evident of recurrent polyps. Result: A case of Ethmoidal sinus shwannoma, a bening nasal mass presented as a right Antrochoanalpolyp, CT Scan reported as a antrochoanal polyp which was removed completely surgically by Functional Endoscopic sinus surgery procedure without any post operative complication of nerve involvement and patient has been not yet reported with recurrent polyp or smell disturbances till date. Conclusion: Aparanasal sinus shwannomas arising from ethmoid sinus can be removed completely removed by Functional endoscopic sinus surgery without any Nerve injury /cranial nerve palsy.***

Keywords: ethmoidsinus, shwannoma, malingnant

1. Introduction

The paranasal sinuses are a common site for polypoid lesions, particularly those of inflammatory nature; however nerve sheath tumours are a rare presentation. Most of these lesions arise from the branches of the trigeminal nerve. The pathologists should be aware of such a presentation in this rare location. Careful assessment of these lesions is important to exclude malignancy.

2. Case Description

Discussion

A 45 year old female, attended ENT OPD of PDU civil hospital, Rajkot with Right sided nasal obstruction for 1 year along with mouth breathing, with no history of anosmia or hyposmia, epistaxis, difficulty in breathing. On examination, Anteriorrhinoscopy right side nasal polypoidal mass, Examination of Eye and Oral cavity found to be normal. On nasal endoscopy right side nasal polyp extending to posterior choana occupying the right nasal cavity with deviated septum towards left side. CT PNS shows findings Symptoms was not relieved by taking steroids medications. Patient was not a known case of diabetes mellitus, hypertension and any chronic illnesses. There was no history of smell disturbance. On examination anterior rhinoscopy right side nasal polypoidal mass, on nasal endoscopy right side polyp extending to posterior choana occupying the right nasal cavity with deviated septum towards left side. CT PNS (Fig: 1) shows findings of polypoidal lesion noted at right sided ethmoidal air cells and right nasal cavity extending posteriorly up to right side nasopharynx. Intraoperatively (Fig.2) polyp identified and removed as a piece meal and sent for HPE. Histopathogoly shows it was a schwannoma with spindle cell proliferation. Follow up Nasal endoscopy shows no evident of recurrent polyps.

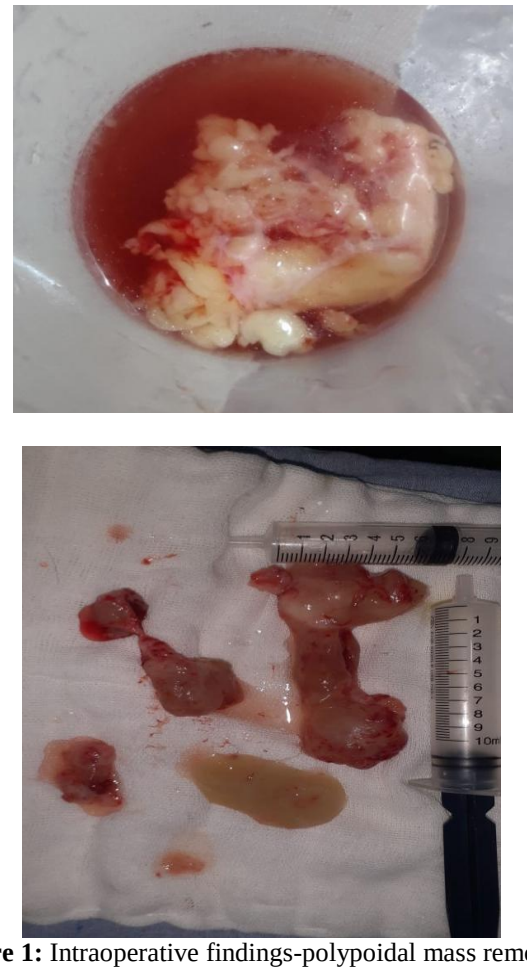


Figure 1: Intraoperative findings-polypoidal mass removed after FESS

Schwannomas are benign nerve sheath tumors. Involvement of the sinonasal region is a rare presentation. Malignant counterparts have been rarely described. Schwannomas occurring in the head and neck accounted for 25–45% of cases, of which only 4% involved the sinonasaltract. The lesion originates from branches of the trigeminal nerve. Symptoms of sinonasalschwannomas resemble other common inflammatory sinonasal conditions such as polyps, mucocele or sinusitis.] The most frequent presenting symptom is nasal obstruction. Less frequent symptoms include exophthalmos, facial swelling, cranial nerve palsy and visual disturbances. Schwnnomas originating from the nasal septum tend to be more symptomatic when compared to shwannomas of the paranasal sinuses due to the confined area of the nasal cavity. Berlucchi et al., reviewed the cases published on schwannomas of the nasal septum. [3] Herein, we review the clinical and the radiological presentation of the previous cases published on nasal cavity schwannoma since 2000 [Table 1]. Only 17 cases were reported during the period 2000–2013 in PubMed. The mean age of presentation of nasal cavity schwannoma was 39 years; ranging from 11 to 82 years. Female predilection was observed with F: M ratio of 1.8: 1.4 cases (23%) originated from the nasal septum. The principal clinical presentation was nasal obstruction followed by epistaxis with rare cases of dysphonia, insomnia and rhinorrhea. Most of the cases were related to the nasal septum, which often showed deviation. The radiological features were mainly of a well-circumscribed homogeneous soft tissue mass that may extend to the paranasal sinuses and the nasopharynx with no destructive growth. Hu *et al*. studied the magnetic resonance imaging of nasal schwannoma in a series of 5 cases. They found that these tumors have an intermediate intensity on T1-weighted and T2-weighted;

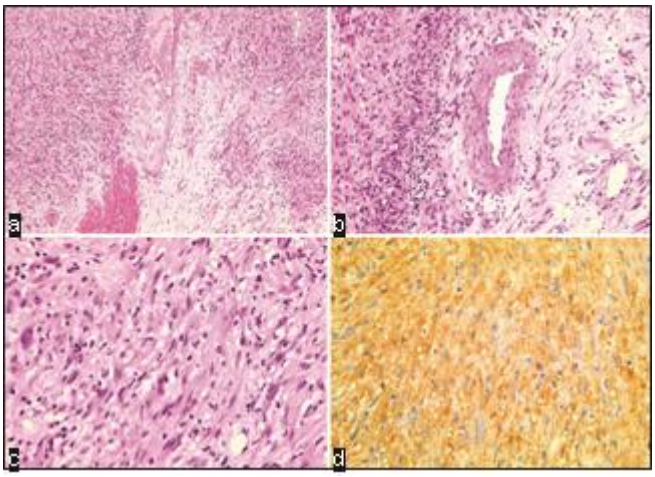
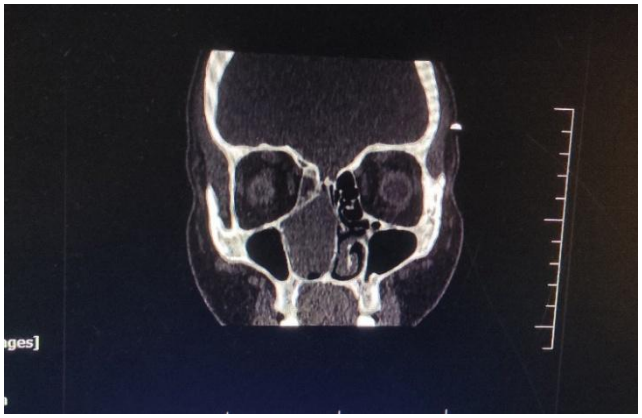


Figure 3: (a) Spindle cell proliferation in hypocellular and hypercellular zonation (b) Thick walled vascular spaces and vague palisading of tumour cells (c) The cells show random mild cytological atypia. No increased mitotic activity or necrosis (d) S100 shows diffuse positivity.

with sometimes higher intensity on T2-weighted. Homogenous enhancement was seen after contrast. Follow-up of the cases revealed that the lesion was unlikely to recur, which reflects the indolent nature of the neoplasm.



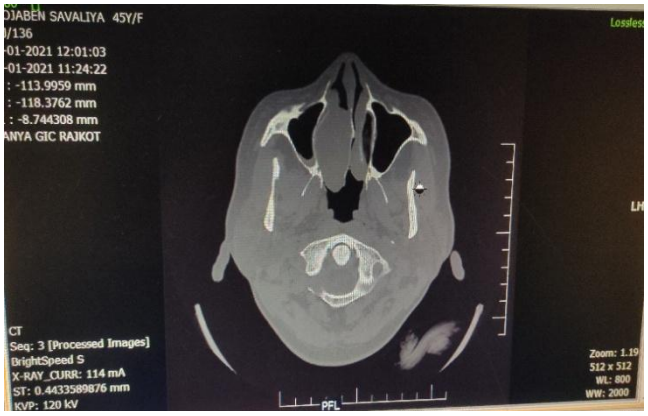


Figure 2: CT PNS showing right side nasal polyp occupying the right nasal cavity, extending till right nasopharynx.

The histological differential diagnosis of sinonasal schwannomas includes lesions characterized by spindle cells that include the following:

Neurofibroma; a benign peripheral nerve tumor composed of Schwann cells, perineural cells and intraneural fibroblasts. The tumor is also characterized by myxoid matrix and scattered mast cells. Immunohistochemically neurofibromas are reactive to S100 protein, EMA and NF. Diffuse neurofibroma is a distinctive variant of neurofibroma characterized by spindled cells embedded in a fibro-myxoidstroma, the tumor infiltrates adjacent fat creating a honeycomb pattern. Wagner-Meissner bodies are also seen in diffuse neurofibroma. This form of neurofibroma is more common in subcutaneous locations.

Ectopic or secondary meningiomas of the sinonasal cavity are mainly meningothelial or transitional type and rarely the fibroblastic type. The tumor cells have a syncytial arrangement with a whorled pattern. The cells have a pseudo-nuclear inclusions and psammoma bodies. Immunoreactivity for EMA can be helpful to exclude the entity.

Angiofibroma has a characteristic presentation in young males involving the postero-lateral wall of the roof of the nasal cavity. Histologically the lesion is characterized by hyalinized vascular stroma containing stellate and spindled myofibroblasts with numerous mast cells. The vascular spaces had variable wall thickness and characterized by The

cells are reactive to vimentin and β-catinin with staghorn appearance and lack elastic fibres in their walls focal SMA.

Table 1: Summary of the schwannoma cases involving para nasal sinuses

Reference	Age (years) / Site	Clinical features	Radiological features gender		Follow-up
Wada et al., 2001	62/female	Bilateral nasal cavities, ethmoid sinuses and maxillary sinuses Nasal vestibule	Bilateral nasal obstruction	Enhancing mass in the nasal cavity and bilateral nonenhancing areas in both maxillary sinuses	Free of disease 15 months after surgery
Leu and Chang 2002	UK/female		N/A	N/A	N/A
Leu and Chang 2002	UK/female		N/A	N/A	N/A
Hazarika *et al.*, 2003	35/female		Bilateral nasal obstruction	Soft tissue mass occupying the right nasal cavity and extending to the maxillary sinus. The nasal septum is deviated	Free of disease 6 months after surgery
Khnifies *et al.*, 2006	42/female		Slowly progressive nasal obstruction	Soft tissue mass extending from left inferior turbinate to left nasopharynx	Free of disease 6 months after surgery
Rajagopal *et al.*, 2006	54/female	Inferior turbinate	Left sided nasal obstruction	Homogeneous mass within the mid portion of the nasal cavity suspected to be arising from the nasal septum	No recurrence at 6 months
Gupta et al., 2008	22/female	Right nasal cavity	Swelling, blockage, and watering from right eye	Well defined homogenously enhancing lesion involving the right nasal cavity. The mass extends through the cribriform plate to anterior cranial fossa	Free of disease 4 years after surgery
Pagella *et al.*, 2009	82/male	Left inferior turbinate	Recurrent dysphonia	Soft tissue mass occupying the posterior nasal cavity	Free of disease, 5 years after treatment
Jacopo et al., 2009	20/male	Left sided nasal cavity	Progressive right nasal obstruction and epistaxis	A mass involving the posterior part of the right nasal fossa and the ethmoid complex	Free of disease 8 months after surgery
Ramavat *et al.*, 2010	11/female	Right nasal cavity	Nasal obstruction and bloody nasal discharge	Sift tissue mass eroding the cribriform plate. The mass was of an intermediate intensity on T1- and T2-weighted images	No evidence of disease 4 months after surgery
Hu *et al.*2012	59/male	Left nasal fossa	N/A	Well defined right nasal vestibule mass with uneven density and mild enhancement	Free of disease 12.8 years
Hu *et al.*2012	51/male	Right nasal fossa and the right ethmoid	Nasal bleeding, headache, anosmia and rhinorrhea	Expansile well defined mass involving the left nasal cavity and extending to the left maxillary sinus. The nasal septum is deviated	Free of disease 12.3 years after surgery
Hu *et al.*2012	56/male	Left nasal cavity, frontal and ethmoid areas	Nasal bleeding	Well defined homogenous mass (1.0 cm × 1.0 cm) involving the left nasal vestibule. Homogenous enhancing on contrast	Free of disease 7.8 years
Hu *et al.*2012	48/female	Right nasal vestibule	Nasal bleeding	2 cm soft tissue mass extending into the nasopharynx with resorption of the nasal septum. The mass is enhancing in - homogenously	Free of disease in 4.8 years.
Hu *et al.*2012	27/female	Left nasal septum	N/A	Well defined soft tissue mass on the right nasal vestibule. Patchy enhancement on contrast	Died of heart attack
Hu *et al.*2012	78/male	Left nasal septum	Nasal obstruction	Nodular low-density homogeneous mass eroding the medial wall of the maxillary sinus	Free of disease 12.7 years
Pauna *et al.*, 2013	31/female	Right nasal septum	Nasal obstruction, anosmia and headache	Homogenous mass on the left nasal cavity and extending to the ethmoid and sphenoid sinuses	N/A
Ohashi *et al.*, 2013		Right nasal vestibule			No signs of disease postoperatively
		Right nasal cavity and septum			
		Polypid lesion on the left nasal cavity			

UK: Unknown, N/A: Not available

Glomangiopericytoma is a distinctive spindle cell lesion of the sinonasal cavity, originating from modified perivascular myoid cells. The tumor cells are uniform spindled to oval cells, densely packed with little intervening collagen. Staghorn capillary vessels with occasional hyalinized walls characterize the vascularity of the lesion. The immunohistochemistry of the cells shows reactivity to SMA, FXIIIa, vimentin and negative immunoreactivity to CD34, Bcl2 and CD99. Focal expression of CD34 and calretinin has been reported in nasal schwannomas.

Leiomyomas grow in intersecting fascicles of spindled cells. The cells have cigar-shaped nuclei and they appear to have perinuclear halos when the fascicles are visualized in cross-section. Immunohistochemically the cells react to desmin, SMA, calponin and H-caldesmon.

Ancient change in schwannoma could be misinterpreted as a malignant peripheral nerve sheath tumour. Malignant spindle cell lesions including, spindle cell carcinoma, melanoma and leiomyosarcoma. The absence of fascicular growth, increased mitotic activity and hypercellularity, excludes malignant peripheral nerve sheath tumor. Spindle cell carcinoma contains areas of squamous cell carcinoma or carcinoma-*in-situ* of the overlying epithelium and reacts to cytokeratins and occasionally to vimentin. Desmoplastic malignant melanoma shows reactivity to melanocytic markers. Leiomyosarcoma has a similar morphology and immunohistochemistry to leiomyoma but with more pleomorphism, increased mitotic activity or necrosis.

The treatment of nasal schwannomas involves surgical excision, and the extent of the lesions influences the surgical approach The importance of the sinonasal endoscopy in the diagnosis and the management has been emphasized. [2] Preservation of the nerve trunk could prevent postoperative neurological complications

3. Conclusion

Schwannomas are benign peripheral nerve sheath tumors that rarely involve the para nasal sinuses and often present clinically as nasal polyps. The radiology of the lesion usually reflects the indolent nature of this tumor with no bone destruction or soft tissue invasion. The treatment involves complete surgical excision with no reported cases of recurrence after surgery.

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