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Original Research Paper

Otolaryngology

A CASE SERIES ON CLINICOPATHOLOGICAL PROFILE OF RARE TYPES OF SINO-NASAL MASS IN ADULTS

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ABSTRACT

Introduction: Nasal cavity & paranasal sinuses are close to important structures such as skull base, orbit, cranial nerves and vital vascular structures. A Sino-nasal mass presents with complaints of nasal obstruction, nasal discharge, epistaxis, loss of smell sensation, post-nasal discharge. Therefore the patient may ignore this early symptoms and sometimes do not treat an early-stage malignancy thinking it to be a benign pathology.

Aim: To study the clinical presentation, histopathological types and management of some uncommon lesions of sino-nasal cavities in an adult.

Material & Methods: The study evaluates the clinic-pathological & treatment profile of some uncommon entities of sino-nasal mass in patients attending ENT Outpatient department of our Tertiary Centre from January 2020 to January 2021.

Results: Two rare varieties of benign lesions were sino-nasal schwannoma and paranasal pilomatrixoma. Pleomorphic type of adult rhabdomyosarcoma, chondrosarcoma and esthesioneuroblastoma were the other types of rare malignant lesions.

Conclusion: Majority of patients with sino-nasal malignancies present at a late stage. Therefore a proper correlation of history, clinical examination, nasal endoscopy and imaging is important for early diagnosis and treatment.

KEYWORDS : Sino-nasal mass; Adult; Histopathology; Surgery; Radiotherapy

INTRODUCTION

Sino-nasal mass is a common finding seen in patients attending our ENT outpatient department. The most common presentation is nasal obstruction followed by rhinorrhea.[1] A sino-nasal mass can have various differential diagnoses. It can be a congenital, inflammatory, benign or malignant or traumatic lesion. [2, 3] We have reported clinic-pathological profile of few rare pathologies presenting as a sino-nasal mass. Benign lesions are common in 2nd to 4th decade and malignancies present in 5th to 6th decade.[4] Most of the patients are diagnosed after clinical examination, nasal endoscopy guided biopsy, immunohistochemistry and imaging following which the final treatment is planned. It is important to delineate the site of origin and extension before any treatment.

CASE SERIES

CASE-1

A 60-year-old man presented with one-year history of bilateral nasal obstruction, 2 month history of frontal headache, with several episodes of epistaxis. There was no history of diminished vision, prolonged fever, neck swelling, loss of appetite, weight loss or any associated ear or throat symptoms. Her general and systemic examinations were normal.

Anterior and posterior rhinoscopy revealed presence of mucopurulent discharge and a fleshy mass blocking middle part of both the nasal cavities. No features of raised intracranial pressure was found. Hematological and biochemical investigations were within normal limits. Nasal endoscopy showed a mass which occupied the middle part of both nostrils arising from the nasal septum. It appeared to have a smooth surface with prominent superficial vessels on it. Plain X-ray of the paranasal sinuses revealed radioopaque shadow in midline of septum.

Computed tomographic scan showed non-enhancing hypodense mass with punctuate calcifications involving posterior one-third of nasal septum and both nasal cavities with features of bone erosion. (Fig.1)

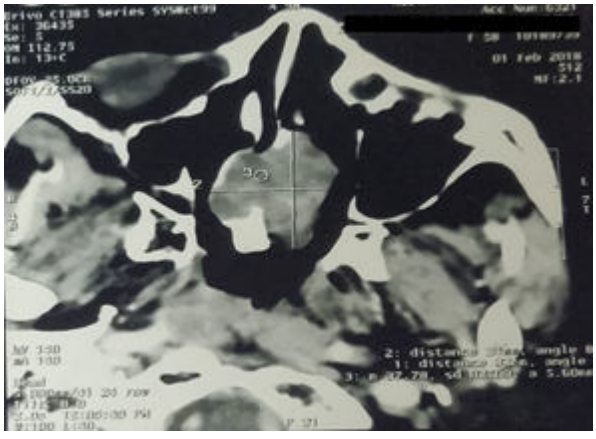

Fig.1 Axial View of nose & paranasal Sinus showing the non-enhancing mass in posterior part of nasal cavity.

Endoscopic excision was performed under general anesthesia. Mucoperichondrial and mucoperiosteal flap was raised after giving an incision at bony-cartilaginous junction of nasal septum. A firm reddish mass was attached to the nasal septum filling the nasopharynx in midline. (Fig. 2) Complete removal of the tumour was done and the tissue was sent for biopsy. (Fig. 3) Postoperative period was uneventful.

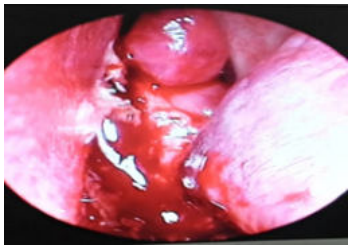

Fig. 2 Nasal endoscopic picture showing a reddish mass in posterior 1/3rd of nasal cavity completely filling the nasopharynx

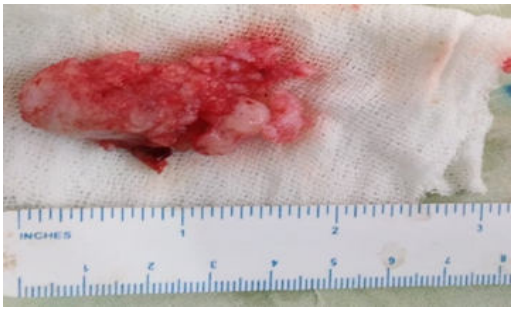

Fig.3 Tumour Tissue

Histopathology revealed lobules of chondromatous tissue at places lined by nasal mucosa. Few lobules were relatively acellular and resembled mature cartilage, while others showed marked increase in cellularity with large binucleated and multinucleated chondrocytes. Stroma consisted of collagen strands having congested blood vessels. (Fig.4) Thus, a diagnosis of low- grade well differentiated chondrosarcoma was made. Following the surgery, he received radiotherapy with a total of 60 Gray in 40 fractions. Clinical and radiological examinations (CT scan) after 1 year follow-up did not show any evidence of recurrence.

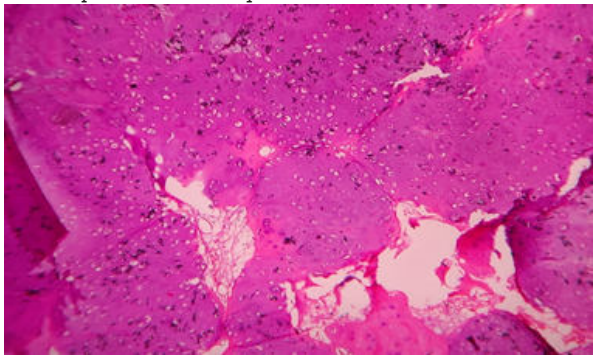

Fig. 4 (H & E, 10 x): showing lobules of chondromatous tissue at places consisting of large binucleated and multinucleated chondrocytes

CASE-2

A 32-year old male presented to our ENT outpatient department with the chief complaints of progressive nasal obstruction for 2 months, bleeding from right nose, blurring of vision and pain in right eye for 20days. Anterior and posterior rhinoscopy shows a smooth pinkish vascular mass present in right nasal cavity occluding the nasal valve anteriorly, and posteriorly extending to nasopharynx occluding the right choana. On examining eyes, it showed non axial proptosis with inferior and lateral displacement of the orbit with movements slightly restricted on medial gaze. Patient visual acuity was decreased in right eye. To further investigate, Contrast enhanced MRI of nose and paranasal sinus was done.

It revealed an extensive polypoidal mass occupying the right frontal, ethmoid, sphenoid sinus and right nasal cavity with extension to right maxillary sinus. It was extending into nasopharynx and deviating the nasal septum to left. There was bulging of medial wall of right orbit along with compression of optic nerve at orbital apex. (Fig.5)This finding explained the ongoing ophthalmoplegia in right eye. Nasal endoscopy guided biopsy of the mass was done under general anaesthesia with proper hemostasis.

Histopathology revealed it to be a rare pleomorphic variety rhabdomyosarcoma. (Fig.6) Immunohistochemistry was positive for desmin and vimentin and negative for cytokeratins, S-100 which confirmed our diagnosis.

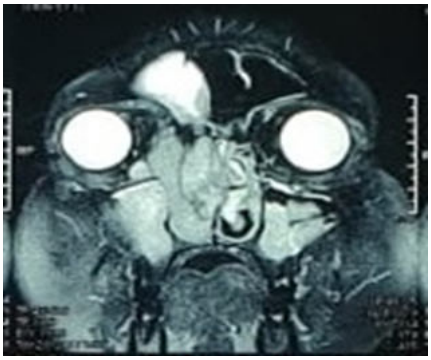

Fig. 5 Contrast enhanced MRI of nose and paranasal sinus revealed an extensive polypoidal mass in all the right paranasal sinuses with an orbital extension.

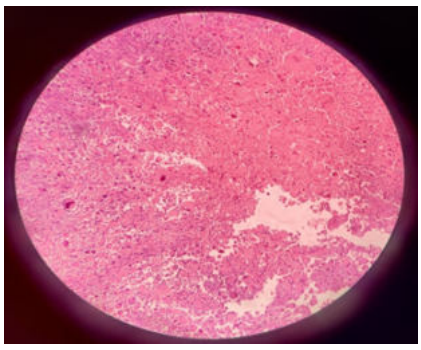

Fig.6 (H & E, 10): showing Pleomorphic type cells with nuclear atypia and thin walled blood vessels suggestive of Pleomorphic variety Rhabdomyosarcoma.

Final diagnosis of paranasal adult type rhabdomyosarcoma with orbital invasion was proposed. A multidisciplinary team of ENT and oncologists decided to start initially Adriamycin based chemotherapy regimen and radiotherapy. There was rapid deterioration of his general health with elevated liver and renal function test parameters. Later on, he developed cerebral edema and died 7 weeks after initiation of chemotherapy treatment.

CASE-3

A 30 year old male presented with complaints of left sided nasal obstruction, nasal discharge and decreased smell sensation for the past 1 year. There was no history of epistaxis, diminished vision and change in voice. General and systemic examination was normal. Anterior rhinoscopy revealed a firm, non-tender pale-white polypoidal lesion with a smooth surface on anterior one third of left nasal cavity. On probing it was found to be extending through the lateral wall of nose to left maxillary sinus. On nasal endoscopy, the lesion was arising from the left middle meatus and extending to left nasal cavity and choana. Computed tomography Scan (CT) showed a minimal enhancing soft tissue dense lesion filling the left maxillary sinus extending to left nasal cavity with widening of maxillary sinus ostium and posteriorly up to nasopharynx. (Fig. 7)

Biopsy was taken from the left nasal mass and provisional diagnosis of nasal polyp was done. Endoscopic excision was planned under general anaesthesia. Middle meatal antrostomy and ethmoidectomy was done. Histopathology of excised nasal mass revealed a focus of stratified squamous epithelium on one side and respiratory epithelium on the other side. The presence of irregularly shaped epithelial cells called basophilic or shadow cells, areas of calcification, ghost cells with clusters of lymphocytes and plasma cells is suggestive of sino-nasal pilomatrixoma. (Fig.8) Follow up examination of the patient after 1st and 6th month did not show any signs of recurrence.

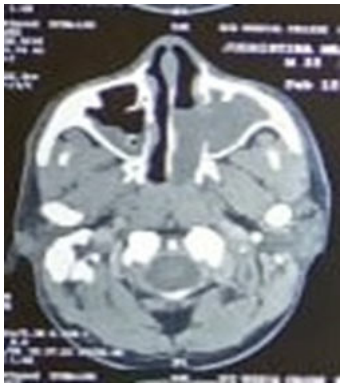

Fig. 7 CT Scan nose & paranasal sinus shows minimally enhancing soft tissue density filling the left nasal cavity extending to left maxillary sinus and nasopharynx

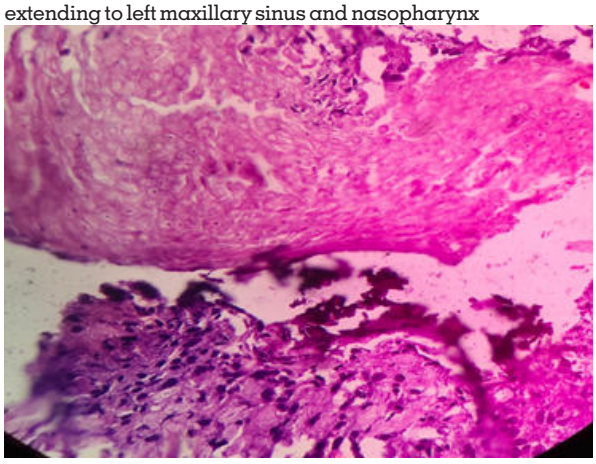

Fig.8 (H & E): showing stratified squamous epithelium with shadow cells, calcifications, basaloid cells

CASE-4

A 45 year old female presented to our outpatient department with complaints of nasal obstruction, blood stained nasal discharge and decreased smell sensation on right side since 1 year. On examination, a pinkish mass seen completely filling the right nasal cavity. The mass was insensitive to touch with smooth surface and it bled on probing. The mass was free from the septum and extended superiorly to roof of the nose. Computed tomography of paranasal sinuses and orbit revealed a homogenously well-defined enhancing mass involving right side nasal cavity and its roof, right maxillary and frontal sinus with mild erosion of frontal bone. (Fig.9)

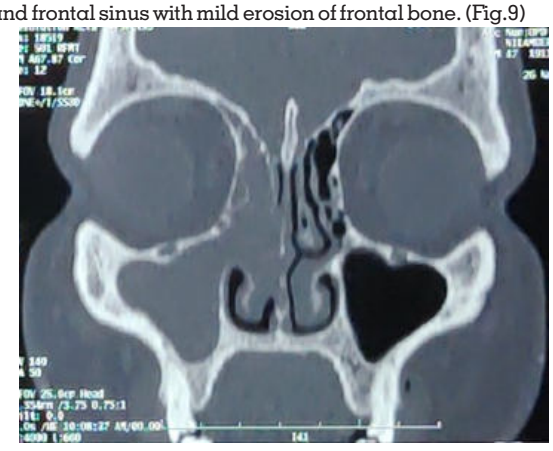

Fig. 9 Coronal CT Scan view showing a homogenous enhanced mass attached to roof of nasal cavity

Biopsy was done under endoscopic guidance under local anaesthesia with nasal packing. Hematoxylin and eosin stained sections revealed nasocilliary epithelium with neoplastic small blue round cells arranged in clusters separated by fibrovascular stroma. These tumour cells are arranged in pseudorosettes and glandular patterns. (Fig. 10) Immunohistochemistry was positive for chromogranin and neuron specific enolase which confirmed our diagnosis as olfactory neuroblastoma. Trans-nasal endoscopic excision of the entire tumour mass was done. The patient was then referred to the oncology department to receive radiotherapy. 6 months follow up did not reveal any recurrence or residual tumour.

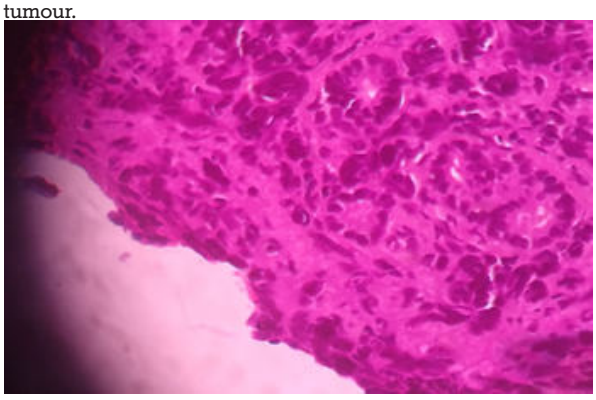

Fig. 10 (H & E, 40 x) : showing Homer-Wright rosettes

DISCUSSION

There is a wide spectrum of common to rare pathologies for any sinonasal mass in an adult. Some of the rare differential diagnosis includes benign lesions like paragangliomas, neurofibroma, basal cell adenoma, fibroma, chordoma, rhabdomyoma and chondroma.

Some of the rare malignant lesions are sinonasal undifferentiated carcinoma, hemangiopericytoma, olfactory neuroblastoma, sarcomas, plasmacytomas, metastatic carcinomas, melanomas. Most of the tumors originate from the maxillary sinuses. Less common sites are nasal cavity, ethmoid sinuses and rarely frontal or sphenoid sinuses. Patients present with vague symptoms like nasal obstruction, nasal discharge, bleeding, snoring, headache or facial pain. [5, 6] In advanced stages patients have visual disturbances, neurological deficits and facial deformities. Rigid or flexible nasal endoscopy guided biopsies under local anaesthesia are essential for confirming the final diagnosis. Imaging studies & histopathological analysis is conclusive to determine the aetiopathogenesis of the sino-nasal mass as inflammatory or neoplastic. CT scans are best to evaluate the bony structures and skull base where as MRI is preferred for defining soft tissue details like orbital, vascular and intracranial extension. Most of the benign pathologies are curable by surgical excision which can be an open or endoscopic approach. Radiotherapy can be advocated alone or following surgical resection. Combination of surgery, radiotherapy and chemotherapy are given in certain specific pathologies.[7,8]

Rhabdomyosarcoma is a highly aggressive tumor with poor prognosis, making it necessary to diagnose it in early stages. They are common in pediatric age group, and are extremely rare in adults. Around 35% is localised to the head and neck region. We have reported a rare pleomorphic type of rhabdomyosarcoma in a 32 year old male. Anatomically, it is classified as parameningeal, non-parameningeal, orbital and non-orbital. Histologically, 4 subtypes of RMS are recognized: alveolar (20%), embryonal (50%), pleomorphic (20%), and spindle cell/ sclerosing (10%). [9]

Esthesioneuroblastoma is commonly known as olfactory neuroblastoma that lines the cribiform plate, superior aspect

of nasal septum, superior and middle turbinate. It originates from olfactory neuro-epithellium in the roof of nasal cavity. Multimodality approach is recommended which includes minimally invasive approaches to cranio-facial resection, radiotherapy or chemotherapy with palliative care. [10]

Pilomatrixoma is also called as "calcifying epithelioma of Malherbe" which originates from the matrix cells of a hair follicle. [11] The common locations are head and neck followed by upper extremities, trunk and lower extremities. The commonest subsites in head and neck are neck, frontal, temporal, periorbital, preauricular region. [14, 15]. We present a unique case of pilomatrixoma at a rare location which is the nasal cavity and paranasal sinus. Recommended treatment is complete surgical excision with clear margins.

Chondrosarcoma is an uncommon group of cartilaginous tumours arising from primitive mesenchymal stem cell or any tissue having chondroblasts. [12] It constitutes only 4% of non-epithelial tumors of the nasal cavity, paranasal sinuses and nasopharynx. Chondrosarcoma of the nasal septum is extremely rare and it rarely extends into cranial or intracranial areas. In recent trends, endoscopic surgery is more successful and adjuvant therapy like radiation and chemotherapy is reserved for residual or recurrent disease.

CONCLUSION

Majority of patients with sino-nasal masses present at an advanced stage. We have discussed here the clinic-pathological profile of some of the rare differential diagnosis of sino-nasal mass in adults.

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REFERENCES

1. Humayun AHM, Zahurul Huq AHM, Ahmed SMT, et al. Clinicopathological study of sinonasal masses. Bangladesh J Otorhinolaryngol 2010; 16:15-22.
2. Lathi A, Syed MM, Kalakoti P, Qutub D, Kishve SP. Clinico-pathological profile of sinonasal masses: a study from a tertiary care hospital of India. Acta Otorhinolaryngol Ital. 2011; 31(6):372-377.
3. Weymuller EA, Gal TJ. Neoplasms of the nasal cavity. In: Cummings CW, Flint PW, Harker LA, et al. editors. Otolaryngology - Head and Neck surgery. 4th ed. Mosby; 2005.
4. Bakari A, Afolabi OA, Adoga AA, et al. Clinico-pathological profile of sinonasal masses: an experience in national ear care center Kaduna, Nigeria. BMC Research Notes 2010; 3:186.
5. Dasgupta A, Ghosh RN, Mukherjee C. Nasal polyps - Histopathologic spectrum. Indian J Otolaryngol Head Neck Surg 1997; 49:32-6.
6. Pradhananga RB, Adhikari P, Thapa NM, et al. Overview of nasal masses. J Inst Med 2008; 30:13-16.
7. Jansen EP, Keus RB, Hilgers FJ, et al. Does the combination of radiotherapy and debulking surgery favor survival in paranasal sinus carcinoma? Int J Radiat Oncol Biol Phys 2000; 48:27-35.
8. Hoppe BS, Stegman LD, Zelefsky MJ, et al. Treatment of nasal cavity and paranasal sinus cancer with modern radiotherapy techniques in the postoperative setting-the MSKCC experience. Int J Radiat Oncol Biol Phys 2007; 67:691-702.
9. Fatusi O, Ajike S, Olateju S, Adebayo A, Gbolahan O, Ogunmuyiwa S: Clinico-epidemiological analysis of orofacial rhabdomyosarcoma in a Nigerian population. Int J Oral Maxillofac Surg 2009; 38:256–260.
10. Chadha S, Pannu KK. Esthesioneuroblastoma a case report. Indian J Otolaryngol Head Neck Surg. 2011; 63 (Suppl 1):44-46.
11. Duflo S, Nicollas R, Roman S, Magalon G, Triglia JM. Pilomatrixoma of head and neck in children: A study of 38 cases and a review of the literature. Arch Otolaryngol Head Neck. 1998; 124:1239.
12. Rassekh CH, Nuss DW, Kapadia SB, Curtin HD, Weissman JL, Janecka IP. Chondrosarcoma of the nasal septum: skull base imaging and clinicopathologic correlation. Otolaryngol Head Neck Surg. 1996 Jul; 115(1):29-37.