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CASE REPORT

PRESENTATION OF AN UNCOMMON LESION IN THE NASAL CAVITY- NEUROENDOCRINE CARCINOMA

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ABSTRACT:

Introduction: Neuroendocrine carcinomas are very rare be it in the nasal cavity, paranasal sinuses or nasopharynx. A lesion more common in the lungs with the extra-pulmonary forms accounting for only 4% of cases.

Case report: A male patient aged 62 years with complaints of long standing nasal obstruction with intermittent episodes of epistaxis for almost 4 months

Discussion: A nasal endoscopic evaluation suggested a mass arising from the posterior nasal cavity and extending to the nasopharynx, which did not appear to bleed on probing. Surgical debulking of tumour along with histopathological examination was done, suggestive of poorly differentiated carcinoma. Immunohistochemistry confirmed the diagnosis of poorly differentiated neuroendocrine carcinoma. Patient was soon after taken up for chemoradiotherapy.

Conclusion: Extrapulmonary Neuroendocrine carcinomas due to their rare incidence have posed a dilemma in their diagnosis and management. Their presentation 12 and GD markt in advanced stages worsens the prognosis. This lesion should be differentiated from other lesions such as Olfactory Neuroblastoma which show similar features.A surgeon must be thorough with nasal endoscopic examination to evaluate a long standing nasal obstruction associated with epistaxis

Keywords: Nasal cavity mass, Neuroendocrine carcinoma, Immunohistochemistry.

INTRODUCTION:

Nasopharynx and the sinonasal tract is the location for a wide variety of benign and malignant tumours. The use of nasal endoscopes and radiological imaging enhancements has helped in the detection and diagnosis of these lesions. One such lesion is the Neuroendocrine carcinoma. They are defined as epithelial neoplasms with predominant neuroendocrine differentiation[1]. Primary small cell neuroendocrine carcinoma (SNEC) of the nasal cavity and sinuses is an extremely‘ rare tumour and difficult to diagnose by conventional methods of histological examination[2]. First described by Ray Chowdhuri as a differentiated histological entity in the paranasal sinuses in 1965[3], it accounts for just 4% of all cases of neuroendocrine carcinomas.[4,5]

At first thought to arise from the lung due to similar features to anaplastic small cell carcinomas of lung[6], they are now considered to be completely different due to their behavioural differences in relation to metastasis and local spread. They should be differentiated from olfactory neuroblastomas which show similar clinical and morphological features.[7,8] Sinonasal malignancies such as SNEC present with an undifferentiated or poorly differentiated morphology and are composed of small, medium, and large round or polygonal atypical cells. [8] These lesions pose significant diagnostic difficulties for the surgical pathologist, especially with limited biopsy material. The role of immunohistochemistry is of vital nature.Depending on their histopathological and biological characteristics they are classified into well differentiated and poorly differentiated carcinomas.[6]

No specific treatment exists at present for Neuroendocrine tumours of the head and neck and despite improved histological classification they are mostly treated as conventional squamous cell carcinomas or less often as

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small cell carcinomas of the lung[6]. These tumours are aggressive with a poor prognosis and frequent local recurrence and distant metastasis despite multimodal therapy.[1]

CASE REPORT:

A 62 years of age male patient presented with nasal obstruction with intermittent episodes of nasal bleeding for almost 4 months. Patient had been examined and evaluated previously several times in other hospitals but no clear diagnosis had been reached. On nasal endoscopic examination

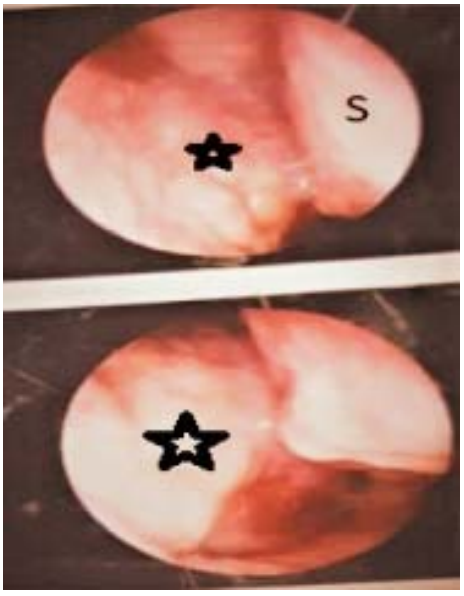

Fig.-1 nasal endoscopic picture. S – Nasal Septum. Star sign depicts the mass in the posterior Nasal cavity and Nasopharynx.

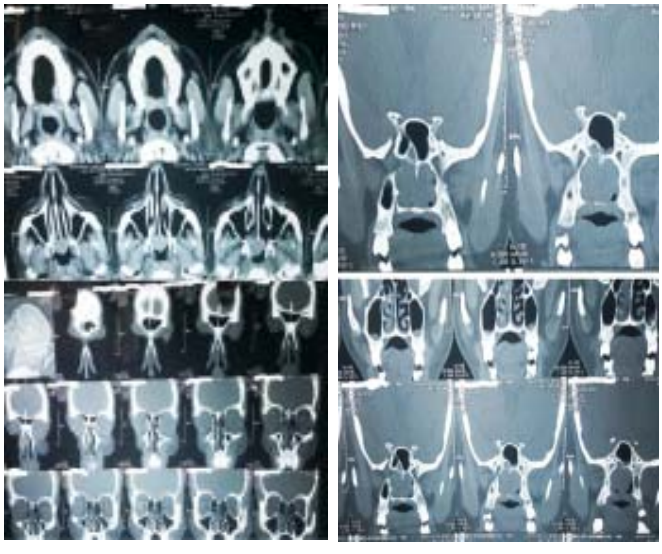

Fig.-2 CT scan showing homogenous soft tissue Mass involving posterior nasal cavity and nasopharynx.

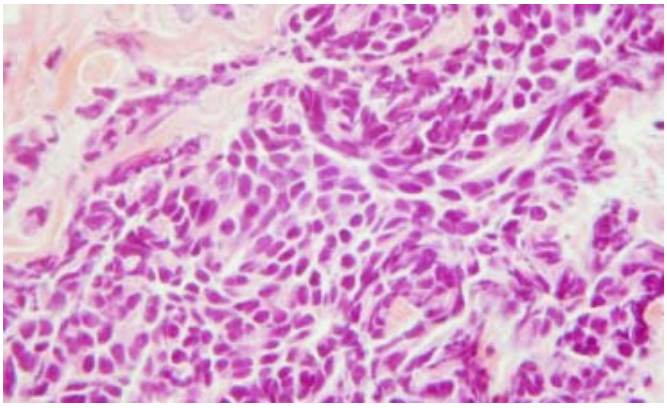

Fig.-3 histopathological examination suggestive of poorly differentiated Sinonasal Carcinoma.

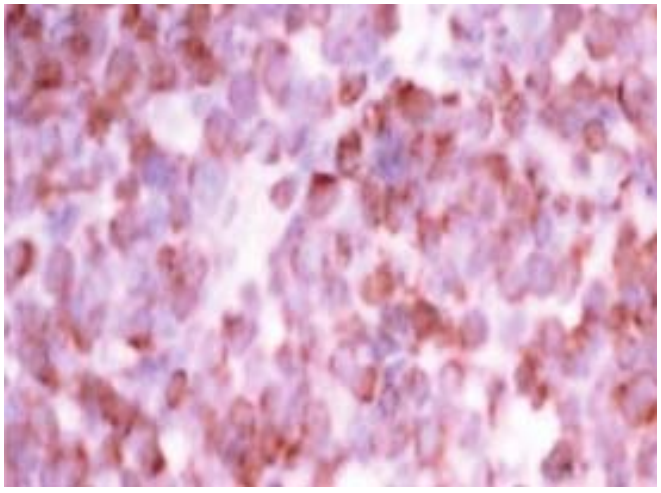

Fig.-4 Cytokeratin staining.

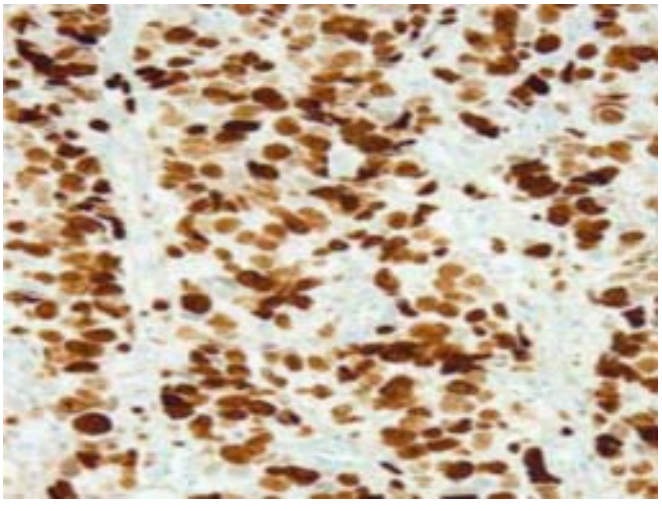

Fig.-5 Synaptophysin staining.

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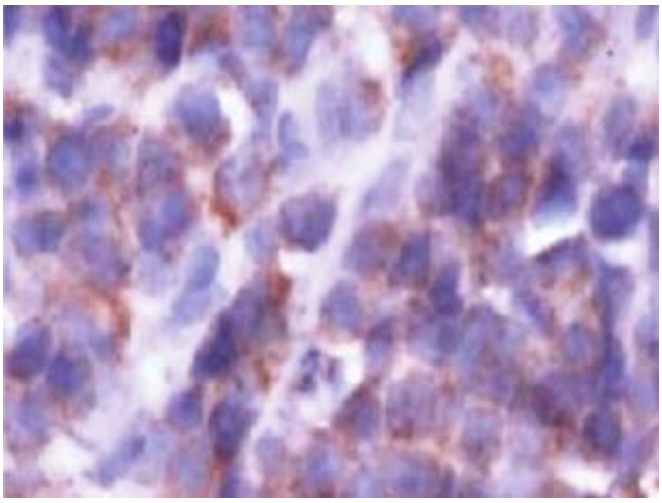

Fig.-6 Ki 67 index greater than 95%.

patient was found to have a proliferative tumour mass in the posterior aspect of the nasal cavity and extending to the nasopharynx [Fig.-1]. Computed Tomography scan revealed a soft tissue mass extending from the posterior aspect of the nasal cavity to the nasopharynx predominantly on the right side. [Fig.-2]

Patient was moderately built and nourished, with normal gait and satisfactory vital signs. There was no history of smoking, drinking alcohol or exposure to radiation or environmental irritants.

Endoscopic debulking of the mass to relieve nasal obstruction and also to obtain biopsy was done. The histopathological examination was suggestive of a poorly differentiated carcinoma [Fig.-3]. Immunohistochemistry of the lesional cells were positive for Cytokeratin (CK) [Fig-4], Synaptophysin [Fig-5], Chromogranin and CD 56 and were negative for p63 and CD45. Ki67 labelling index was found to be about 95% [Fig-6]. These features were suggestive of a poorly differentiated neuroendocrine carcinoma or a small cell neuroendocrine carcinoma and potentially ruled out nasal lymphoma.

DISCUSSION:

Small cell neuroendocrine carcinoma (SNEC) is one of the subtypes of Neuroendocrine carcinoma with the others being Carcinoid, atypical carcinoid and Large cell neuroendocrine carcinoma[8]. These tumours are mainly located in the lungs, and account for 20% of all lung carcinomas[9]. Extrapulmonary SNECs represent 4% of all SNECs[5]. It is a rare tumour with no sex, racial or geographic predilection and no known association with smoking or radiation. The age range is from 26 to 77 years with a mean of 49 years[7]. Most commonly it arises in the superior or posterior nasal cavity, and often extends into the maxillary or ethmoid sinuses. Secondary involvement of nasopharynx is present in a minority of patients as in this case. Advanced tumours can involve the skull base, orbit or brain. Rarely elevated serum levels of adrenocorticotropic hormone and calcitonin may be seen.[10]

Of great importance is to distinguish SNEC from Olfactory Neuroblastoma (ONB). Their relationship remains confusing and controversial. In most cases, SNEC should be readily differentiated from low grade ONB. Sinonasal SNEC lacks lobular architecture, fibrovascular septa, neurofibrillary stroma, and does not contain neural or olfactory rosettes. The anaplastic cells of SNEC are small to intermediate in size with negligible cytoplasm, high nucleo/cytoplasmic ratio, round or oval dense hyperchromatic nuclei, numerous mitotic figures and apoptotic cells accompanied by extensive areas of necrosis[6,8]. Immunohistochemistry appears to play a significant role to distinguish the two entities. SNEC lacks the S-100 positive cells and is negative for Neurofilaments.Strong staining has been reported with synaptophysin and CD 56 nerve cell adhesion molecule[11] as was noticed in our case. A high Ki67 labelling index also favours the diagnosis of SNEC.[8]

Clinical features and behaviour of these lesions are like any other tumour of the sinonasal tract. It usually presents with epistaxis, nasal obstruction, followed by ophthalmic signs (exophthalmos, visual acuity trouble and limitation of eye movement) due to orbital involvement. Local pain, anosmia and cervical node metastasis have also been described[4,2,12]. Our patient too presented with complaints of nasal obstruction and intermittent episodes of epistaxis which goes on to show the importance of thoroughly evaluating these patients with a nasal endoscope.

Radiographically, the tumour always involves the nasal cavity and multiple paranasal sinuses. In our case a predominantly right sided nasal cavity mass extending to the nasopharynx was observed. CT scan can help to diagnose the malignant nature of the tumour, as it can reveal the presence of an osteolytic lesion. The signal of these tumours are homogenous isodense or mild hyperdense on CT. MRI with T1, T2 using I.V gadolinium improves differentiation between

inflammatory reaction, tumour and liquid retention and also involvement of the Meninges. Other lesions can be distinguished on CT scan such as, Inverted Papilloma which has a lobulated or cerebriform configuration, squamous cell carcinoma of sinonasal cavity with bony erosion, adenoid cystic carcinoma shows bony erosion and sclerosis and Olfactory neuroblastoma which is located high in the nasal cavity with peripheral areas of cystic degeneration and calcific foci.[13]

Kadish classification: the initial location of the tumour is rarely precise, usually because of its late discovery. This is why the Kadish classification is often used.[14] The extensive involvement of the nasopharynx and nasal cavity proper pushes the staging to Stage C in our case. Factors such as cerebral invasion, lymph node involvement, visceral metastasis and associated endocrine syndrome renders the usage of the Kadish classification as unreliable.[2]

The limited number of cases published, difficulties of diagnosis and heterogeneity of treatment approaches hamper evaluating the ideal treatment strategy. Though previously surgery followed by radiotherapy and chemotherapy was preferred[15], recent studies recommend neoadjuvant chemotherapy followed by Radiotherapy with surgery only reserved for non-responders[16,17]. However Mitchell et al in their study concluded that a favourable response to Chemotherapy could be used to stratify patients either for definitive surgical resection of disease or Radiotherapy[18]. In our case as the patient complained of significant nasal obstruction, debulking of the tumour was done first using a microdebrider under endoscopic guidance and also tissue for biopsy was obtained. Patient is currently undergoing Chemotherapy using cisplatin and Etoposide and radiotherapy for the disease.

Prognosis seems more favourable in the case of nasal and paranasal locations with 67- 100% of patients alive at 5 years[7,16,19] dropping to 77% at 10 years.[19]

CONCLUSION:

Extrapulmonary Neuroendocrine carcinomas due to their rare incidence have posed a dilemma in their diagnosis and management. Their presentation mainly in advanced stages worsens the prognosis. SNEC should be differentiated from other lesions such as Olfactory Neuroblastoma which show similar features. Proper histopathologic diagnosis is of utmost importance to dictate appropriate therapy. A systemic therapy may be warranted in most cases due to a late presentation. A surgeon must be thorough with nasal endoscopic examination to evaluate a long standing case of nasal obstruction especially when associated with epistaxis.

DISCLOSURES:

a) Competing interests/Interests of Conflict- None
b) Sponsorships – None
c) Funding - None
d) Written consent of patient- taken
e) Animal rights- Not applicable

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