

*Case Report***Sinonasal Neuroendocrine Carcinoma Presenting As Epistaxis-
A Rare Case Report with Review of Literature**Avinash Mane¹, Sujata Kanetkar², Rajesh Karambelkar², Shakuntala Armani¹, Sharda Sarda³¹Assistant professor, ²Professor, ³Tutor;
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*Received: 03/04/2015**Revised: 22/04/2015**Accepted: 27/04/2015***ABSTRACT**

Sinonasal neuroendocrine carcinoma is an uncommon aggressive malignant tumor. Most common sites of these tumors are esophagus, GI tract, salivary glands, etc; however sinonasal tract is a rare site. These tumors show bimodal peak- young adults and elderly. Tumor shows male predominance. Common presenting complaints are nasal mass, epistaxis and nasal discharge. Tumor has capacity to metastasize locally and distantly resulting in poor outcome. Due to complex anatomy of sinonasal tract patient presents in advanced stage and thus requires more aggressive treatment by multimodality approach. We here by present a case of 28 year old female presented with history of recurrent epistaxis.

Keywords: Sinonasal tract, sinonasal tumors, neuroendocrine carcinoma, epistaxis.

INTRODUCTION

Cancers of the upper aerodigestive tract constitute approximately 4% of all malignancies. Amongst which nasopharyngeal carcinoma are relatively rare. Generally they present with epistaxis (46%) followed by nasal mass (42%). [1] Neuroendocrine carcinoma of nasopharynx is extremely rare tumor presenting as a rapidly growing mass particularly in a young age group. Constant association with Epstein-Barr virus, high levels of volatile nitrosamines in the diet are proved to be major risk factors. We from KIMS present a case of SNEC occurring in 28 year old female who presented with profuse epistaxis.

CASE REPORT

28 year old female presented to an ENT OPD with history of repeated episodes of epistaxis for a period of last one month. Nose examination revealed large fungating polypoidal mass obstructing the nasal cavity. It was fleshy and bleeding on touch. CT scan was done and biopsy was sent for histopathological examination. Indirect laryngoscopy was normal. Ear examination revealed no significant pathology. There was no history of cough, fever, rhinorrhoea, hearing loss and headache. CBC examination revealed no significant findings.

GROSS-

Multiple irregular friable gray white to gray brown tissue fragments altogether amounting to 2 g was received.

MICROSCOPY-

Polypoidal lesion lined by pseudostratified ciliated epithelium and stratified epithelium shows a tumor composed of sheets of neoplastic cells which are round to oval having large pleomorphic vesicular nuclei salt and pepper chromatin (fig 1) and scant amount of eosinophilic cytoplasm. Stroma shows increased vascularity (fig2) H & E staining of vessel wall was noted referred to as Azzopardi effect (fig.3). Few areas of necrosis and hemorrhage are also seen. Based on these findings diagnosis of SNEC was offered.

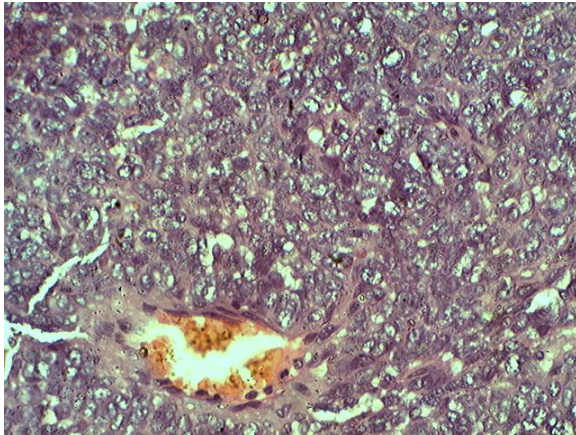


Fig 1- Neoplastic cells showing salt and pepper chromatin.

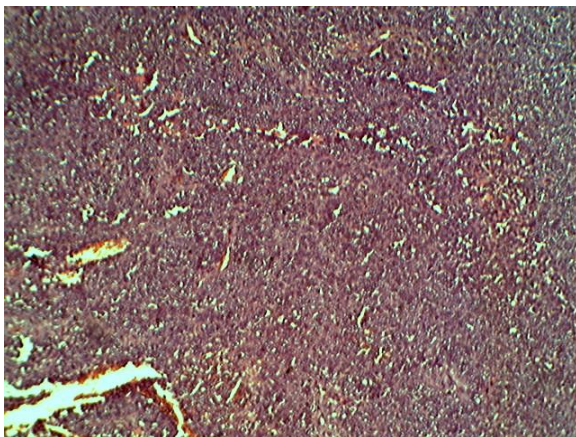


Fig 2: Sheets of neoplastic cells with increased vascularity in stroma.



Fig 3: Blood vessel lined by bluish coloured material (Azzopardi effect).

DISCUSSION

Primary SNEC was first described by Ray Chaudhari in 1965. [2] NEC have been classified into 4 types- carcinoid, atypical, large cell neuroendocrine and small cell NEC. [3] SNEC is rare malignancy, clinical behaviour of which is not known. [4,5] Primary sites for NEC include esophagus, salivary glands, GI tract, etc. Primary sinonasal NEC is extremely rare. [6] In nasal and paranasal malignancies squamous cell carcinoma is most common followed by adenocarcinoma, malignant lymphoma, plasma cell tumor, malignant melanoma and olfactory neuroblastoma. [7] The age incidence is bimodal with peak occurring between 15-25 years and another between 60-69 years. In our case, young adult 28 year old. [8] Men are more commonly affected 2-3 folds more than females. In present case, patient is female. [1] The initial presentation of nasal obstruction, nasal discharge and recurrent episodes of epistaxis is practically indistinguishable from those of benign diseases and hence is likely to result in delay presentation. [7] The aggressiveness coupled with complex anatomy of this region; ensures that most patient present in advanced stage of disease. [7] SNEC is aggressive tumors with high potential for local invasion as well as distant metastasis. As most patient present in

advanced stage, the prognosis is extremely poor. A recent review of literature by Han et al stated that local recurrence rate was 33% and rate of distant metastasis was 31%.^[9] The Kapla-Meier estimate of overall survival at 5 years was 64.2% for patients with SNEC. The local control rate at 5 years was 72.6% for NEC. The regional failure rate at 5 years was 12.9% for patients with NEC.^[10]

CONCLUSION

SNEC is an aggressive tumor usually presenting in middle age as a nasal mass. SNEC has capacity to metastasize locally and distantly resulting in poor outcome however, with advent of newer chemotherapy and better personalized treatment protocols this is changing. So the outcome is improved with better 5 year survival.

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