Section: Radiation Oncology

Case Report

ISSN (O): 2347-3398; ISSN (P): 2277-7253

Primary Nasal Neuroendocrine Tumor: Rare Case Report

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Abstract

Nasal cavity small cell neuroendocrine tumor is an exceptionally rare and violent tumor. The tumor carries a poor prognosis in spite of active treatment, with a high risk of local recurrence and distant metastases. Platinum-based chemotherapy combined with radiotherapy is used in the treatment of small cell cancer. We have a rare case of neuroendocrine primary nasal carcinoma. A 62-year-old female patient comes to OPD with a history of Epistaxis from the left nostril and watering from the left eye.

Keywords: Neuroendocrine tumor, nasal cavity, locally aggressive.

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Received: 22 November 2020 **Revised: 03 January 2021** **Accepted: 17 January 2021** **Published: 27 March 2021**

Introduction

The nasal cavity and paranasal sinuses are the foremost common site for numbers of neoplastic lesions because of their anatomic complexity and tissue variability amongst which squamous cell carcinoma is the foremost common, taken after by adenocarcinoma, lymphoma, undifferentiated carcinoma, melanoma, and olfactory neuroblastoma.[1] Paranasal sinuses and nasal cavity are an uncommon location for neuroendocrine tumors. Small cell carcinoma of the sino-nasal region is very locally destructive and usually presents with recurrence even after treatment. To date, very small numbers of cases have been reported in textbooks and counts for less than 1% of all malignancy and also for 3% of the upper aerodigestive tract.[2]

Neuroendocrine carcinomas are classified into three types depending on their grades of differentiation:

1. Well-differentiated (normal carcinoid)
2. Moderately differentiated (atypical carcinoids), and
3. Poorly differentiated (small and non-small cell type).

Well-differentiated type and moderately differentiated type of neuroendocrine tumor have somewhat superior prognosis as compared to poorly differentiated carcinoma.[3,4]

Poorly differentiated Sino-nasal neuroendocrine tumor is a very uncommon variant and is very aggressive in nature with a very high rate of recurrence and a propensity to metastasize.[5]

Case Report

A 62-year-old female patient has had symptoms of left nostril epistaxis (nasal bleeding) and left eye-watering and obstruction of the nasal cavity on and off for 3 months.

On examination, a proliferative mass was seen in the left nostril 3*3 cm black colored and with bleeding.

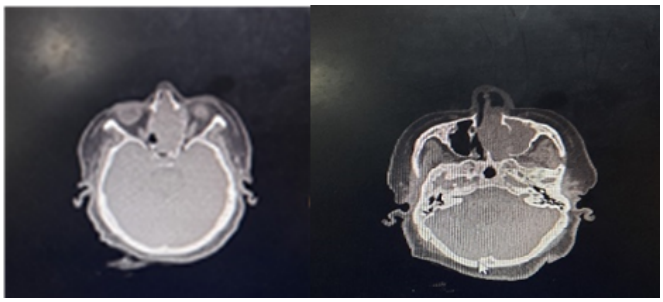
Investigations

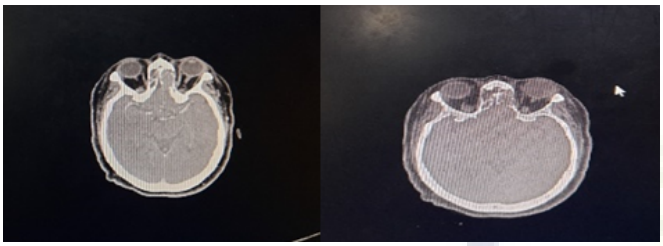
CECT face+neck shows: shows enhancing soft tissue density lesion noted epicentered in the left nasal cavity and ethmoidal air cells measuring 3.34 x 6.87 x 5.15 cm causing bulging and focal erosion of nasal septum with non-enhancing necrotic areas within.

Disease extension: superiorly causing erosion of cribriform plate of the ethmoid bone and ethmoid air cell with the intracranial extension of the soft tissue of size 1 x 0.7 cm into the left basi-frontal region.

Posterio-superiorly causing erosion of the anterior wall of the sphenoid sinus with intra-sphenoid extension.

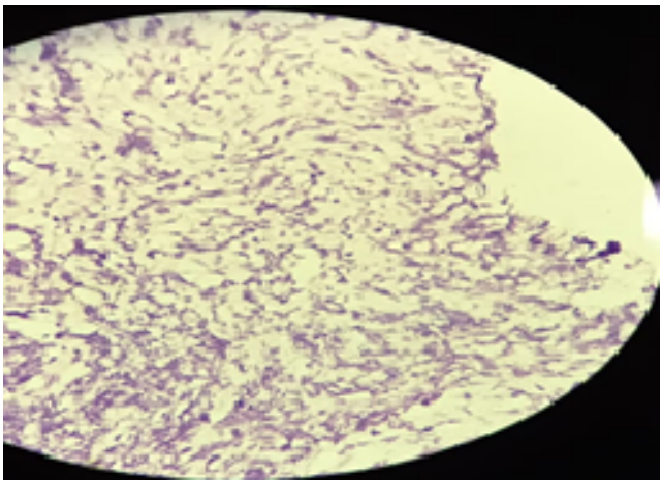
Left laterally eroding the medial wall of the left maxillary sinus and right laterally causing erosion of the nasal septum and bulging into the right nasal cavity.

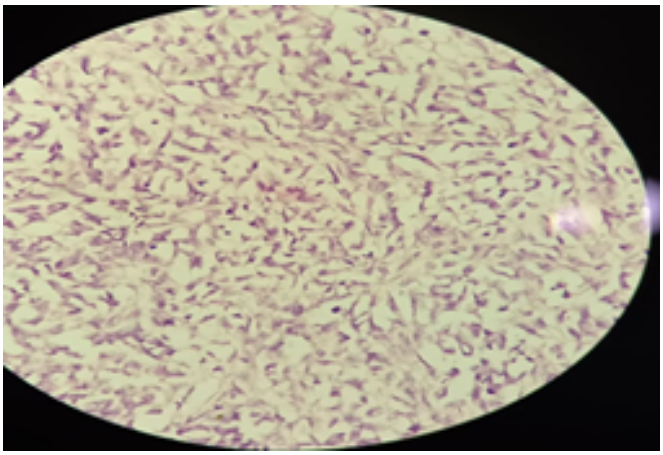




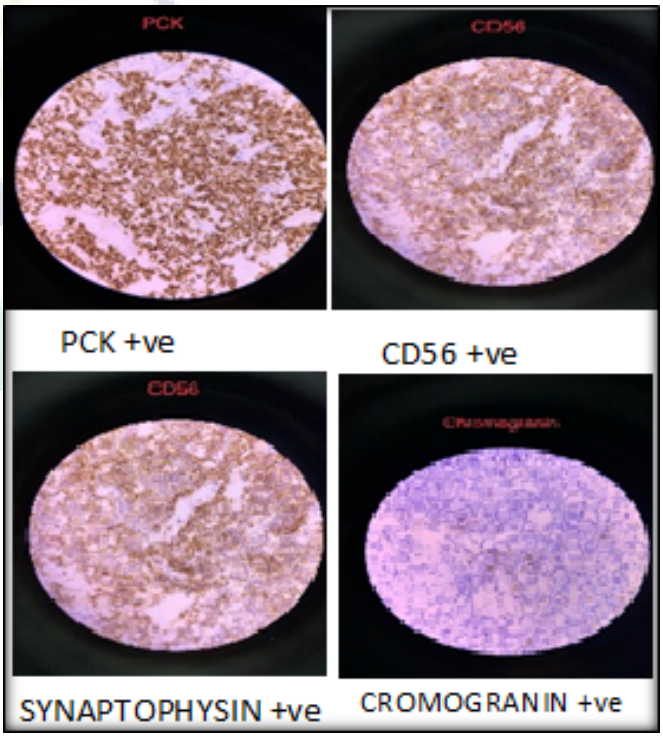
FNAC from the proliferative growth suggestive of poorly differentiated epithelial malignancy.

Biopsy from the proliferative growth shows histopathological features of poorly differentiated malignancy.





IHC was positive for PCK, CD56, SYNAPTOPHYSIN, CHROMOGRANIN-A, and based on the above immunohistochemical finding, the diagnosis of the neuroendocrine tumor was made.



The patient was initially given 1 cycle of paclitaxel and carboplatin-based chemotherapy before the confirmation of diagnosis by IHC as the patient was having symptoms.

After the confirmation of diagnosis by IHC, the chemotherapy regimen was then changed to cisplatin plus etoposide.

Discussion

Nasal cavity and paranasal sinus carcinoma are extremely rare, with an occurrence of less than 1 per 100,000 individuals per year.[6] Neuroendocrine tumor of the extrapulmonary region (EPSNEC) are uncommon, accounts for 0.1-0.4% of all cancers.[7] Head and neck neuroendocrine tumors account for around 11% of all EPNEC. Among EPSNEC, essential Neuroendocrine Carcinoma of the nasal cavity and sinuses is very uncommon. Raychowdhuri in 1965 was the first to report this as separate differentiated histology in the paranasal region.[8] Only 76 cases of small cell neuroendocrine carcinoma of the nasal and paranasal region in the last 45 years have been published in the literature.[9]

World Health Organization (WHO), defines small cell tumors as malignant epithelial tumors comprising of small cells with very little cytoplasm, with ill-defined borders, granular chromatin, missing nucleoli with necrosis, and very high mitotic count.[10] The tumor frequently stains positive for neuroendocrine markers such as synaptophysin, CD 56, and chromogranin A.[11] All Small cell carcinomas whether having pulmonary origin or extra-pulmonary origin, all have comparatively similar morphologic, immunohistochemical, and ultrastructural features.[12] Now since neuroendocrine tumor of the sino-nasal region is very rare, the initial workup is to rule out lung as primary.[13]

The neuroendocrine carcinoma of the sino-nasal tract incorporates a slight male prevalence. It can be seen in any age but the mean age of presentation is 51–58 years.[14] In spite of the natural likenesses between pulmonary and extrapulmonary neuroendocrine carcinoma, the hazard components are not the same. In spite of the fact that a history of tobacco intake relates unequivocally with a pulmonary neuroendocrine tumor, but not for EPSCC.[15]

Based on the extent and degree of tumor spread, EPSCC can be classified as

- Limited
- Extensive

When the tumor is limited to or confined to the primary site and local lymph nodes it is said to be a limited (local) disease, whereas the disease is said to the extensive stage when it extends beyond the loco-regional boundaries.[16] Neuroendocrine tumor carries a very poor prognosis, in spite of holistic management the disease has a recurrence rate of 80% or distant metastasis within 2 years.[17] The median overall survival is 1.4 years for the limited disease and 0.7 years for extensive diease.

The clinical trials have not been done for determining the optimal treatment plan for EPSCC due to its rarity. The treatment plan for EPSCC is combination chemotherapy with 4 cycles of cisplatin and etoposide, together with radiotherapy of atleast 50 Gy in 2 Gy per fraction. Surgical management is usually reserved for any residual lesion.[18] Because of the very extensive disease and poor prognosis of the disease in our patient, we have decided to give platinum-based chemotherapy followed by localised radiotherapy.

Conclusion

Patient in this case is a 62 year old female who presented to us with nasal bleeding and nasal obstruction due to mass in the sino-nasal region with some intra-cranial extension. The diagnosis was confirmed by biopsy followed by IHC. Due to very extensive and advanced stage disease and also because of poor prognosis of the disease, our management plan is to give cisplatin based chemotherapy followed by palliative radiotherapy.

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How to cite this article: Bandhate K, Diwan AK, Mahobia V. Primary Nasal Neuroendocrine Tumor: Rare Case Report. Asian J. Med. Res. 2021;10(1):1-4.

DOI: dx.doi.org/10.47009/ajmr.2021.10.1.RO1

Source of Support: Nil, **Conflict of Interest:** None declared.