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Case Report

Neuroendocrine Carcinoma of the Paranasal Sinus: A Case Report

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Abstract

Neuroendocrine carcinoma is an uncommon tumor. Primary NEC of the paranasal sinuses is extremely rare; only about 76 cases have been reported in literature. Unfortunately due to the rarity of this neoplasm, there are no specific recommendations regarding management of the disease. While Surgery was used in the past, chemoradiation now seems to be evolving as the treatment of choice. The clinical presentation, imaging studies, histopathological diagnosis with immunohistochemistry correlation, management protocols, and a brief review of literature of this rare tumor is discussed.

Keywords: *Management, paranasal sinus, prognosis, small cell neuroendocrine carcinoma*

Introduction:

Neuroendocrine carcinoma is an uncommon tumor. This tumor usually occurs in the lungs and the extra-pulmonary form accounts for only 4% of all cases[1]. Primary NEC of the paranasal sinuses is extremely rare and only about 76 cases have been reported in literature. Treatment approaches to these rare tumors has been controversial, with the trend changing from surgery in the past to chemoradiation. We report a case of sinonasal NEC primarily managed in NIENT.

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Case Report

A 55-year-old female from kalikabadi, Pabna admitted on 26/1/17 in NIENT with complaints of progressive nasal obstruction and occasional nasal discharge from left side of nose for 6 months and sudden loss of vision for last one and half months. Movement of eye ball was restricted for same duration. She was diabetic for last 7 years and hypertensive for last 2 years. She was habituated to tobacco powder for several years. On examination: A soft reddish mass in the left nasal cavity, bleeds on touch, pain sensitive, firm in consistency, seems to be attached on left nasal wall. Sense of olfaction was impaired; loss of vision as well as mobility of eyeball was restricted. There was no trismus. Oral cavity (teeth, gum, hard palate, gingivolabial sulcus) was normal. Orodental hygiene was average, Posterior rhinoscopy reveals post nasal mucopurulent discharge .cervical lymph nodes were not palpable.

Fig.-1: *Preoperative picture*

Fig.-2: *Postoperative picture*

Computerized tomography (CT) scan demonstrated a large contrast enhancing soft tissue density mass noted in left maxillary sinus with extensions into the left. Mass destroyed the medial wall of left orbit and extended to orbit. It had pushed the globe mildly forwards and laterally. Posteriorly posterior nare was obliterated. Sphenoidal sinus was also involved in the mass. Both eye balls are of normal size and having normal appearing vitreous & lens and proptosis of left eyeball. Optic nerves on both side showed normal course and caliber. Eye muscles were normal width and course. Retrobulbar fat was unremarkable.

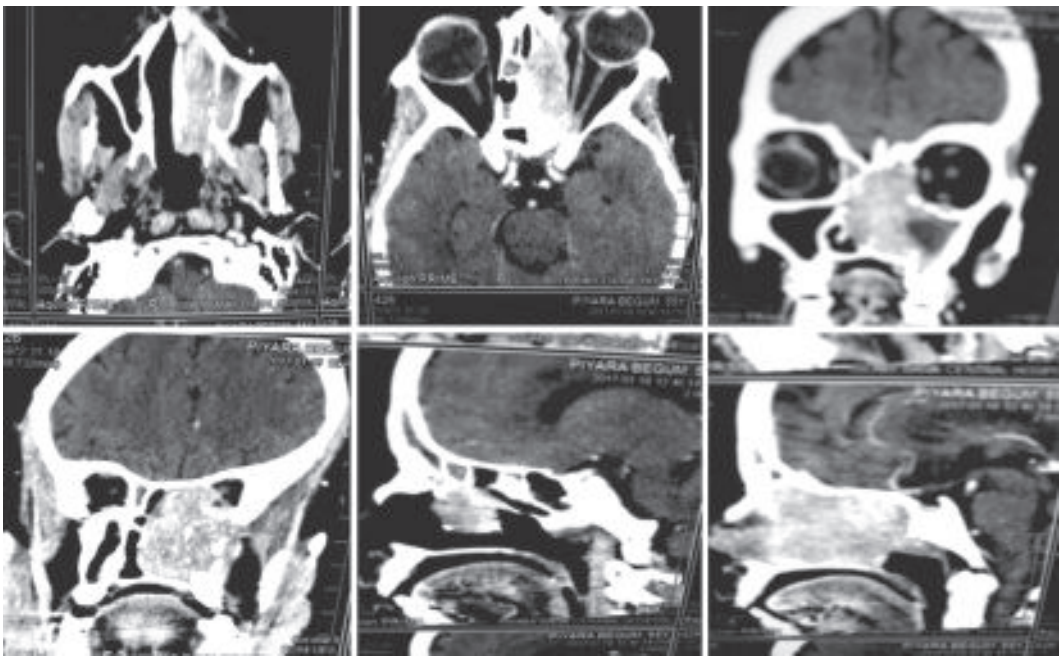

Fig.-3: *CT Scan Axial,Coronal and Sagittal section]*

Surgery was conducted and medial maxillectomy was done on 6/2/17.

Biopsy of the lesion was done which on microscopy revealed malignant neoplasm, composed of round cells, arranged in sheets and nests, separated by fibrous tissue. There is rosette like structures. Mitosis was noted and diagnosis was high grade malignant neoplasm, consistent with high grade neuroendocrine carcinoma.Differential diagnosis was high grade olfactory neuroblastoma and immunohistochemistry was recommended 9/2/17. Immuno-histochemical profile showed tumor cell positivity for Epithelial Membrane Antigen (EMA), pancytokeratin, and focally positive for synaptophysin, The tumor cell was negative for S-100 Protein.

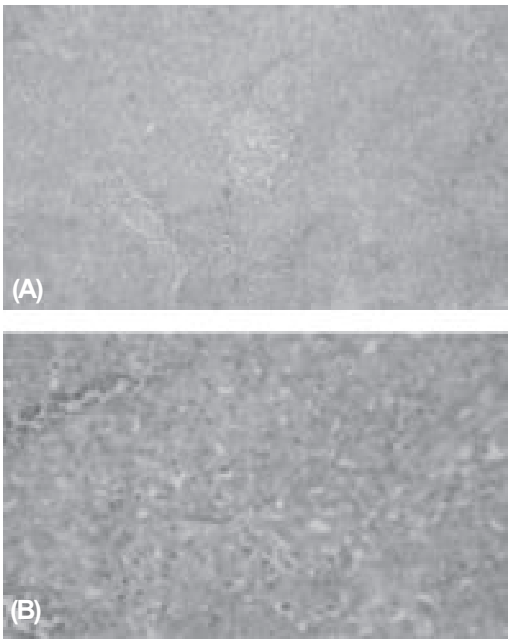

Fig.-4: (A) Neuroendocrine carcinoma demonstrating large sheets of tumoral cells with confluent necrosis (centre); **(B)** Detail showing large, round cells with moderate amounts of eosinophilic cytoplasm and large, round nuclei with finely dispersed chromatin.

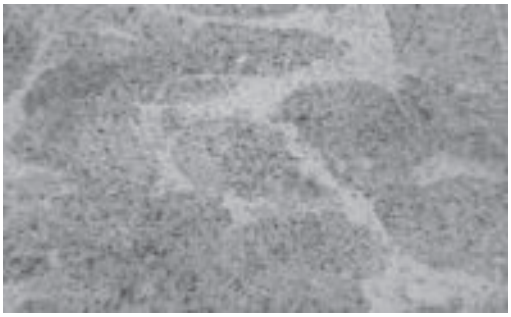

Fig.-5: *Immunohistochemical staining for chromogranin*

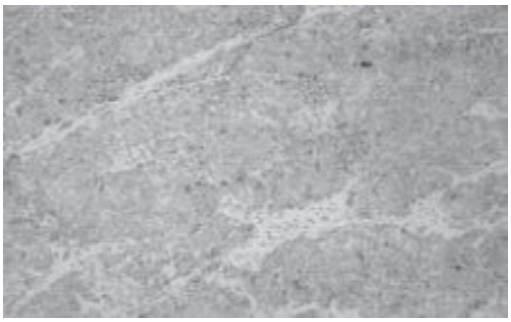

Fig.-6: *Perinuclear dot-like cytokeratin staining*

Discussion

Neuroendocrine carcinomas have been classified into from low grade to high grade into four types–carcinoid, atypical carcinoid, large cell neuroendocrine carcinomas, and SNECs[2].

Extra-pulmonary primary neuroendocrine carcinomas are uncommon malignant neoplasms, whereas primary sites documented may include esophagus, salivary glands, gastrointestinal tract (including small intestine and large intestine), pancreas, larynx, cervix uteri, uterus, urinary bladder, prostate, breast, and lacrimal gland. Primary sinonasal SNECs are extremely rare.

SNECs are thought to arise from a multi-potential stem cell. SNEC is a histological sub-type among a broad group of sinonasal malignancies together known as sinonasal

neuroendocrine tumors. Other sub-types include sinonasal undifferentiated carcinoma and esthesioneuroblastoma.

The initial presentation of nasal obstruction, nasal discharge, and recurrent epistaxis is practically indistinguishable from that of more benign diseases and hence is likely to result in delay in presentation. Occasionally, the presenting complaint may be swelling of the maxilla and expohthalmos. The aggressiveness coupled with the complex anatomy of this region, ensures that most patients present in an advanced stage of disease. The association between the paraneoplastic endocrine syndrome and SNEC is well documented. However, review of literature showed only in five cases of SNECs of nasal and paranasal sinuses with endocrine syndromes[2]. Our patient did not have any endocrine syndrome except diabetes which was detected 7 years back.

Diagnosis by histological examination is challenging. It is indistinguishable from its pulmonary counterpart. Both consist of small-sized cells arranged in sheets, nests, or cords, with moderate to scanty cytoplasm and hyperchromatic nuclei. It is important to differentiate this tumor from olfactory neuroblastoma, which is a low grade tumor. Definitive diagnosis is made by correlation with immunohistochemistry and/or electron microscopy.

Unfortunately due to the rarity of this neoplasm, there is no specific recommendation on management guidelines, treatment options are generally extrapolated from similar tumors of pulmonary origin. Treatment approaches to this aggressive tumor has varied over the years. Previously, surgery followed by radiotherapy or chemotherapy was preferred[4, 5]. In general, these patients usually present in advanced stages, surgery can be extremely disfiguring and may not be curative. Chemotherapy using cisplatin and etoposide followed by high dose proton-photon radiotherapy has been proven by some authors to be an effective line of treatment[6, 7]. Surgery is now reserved for non-responders. Despite presenting in an advanced stage, our patient responded well to a standard combination of chemotherapy and radiotherapy and continues to be in remission.

Conclusion

NECs are aggressive tumors with high potential for local invasion as well as distant metastasis. Metastatic deposits occur in the brain, bones, lungs, and skin.As most patients present in advanced stages, the prognosis is extremely poor.

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