Sinonasal neuro endocrine carcinoma extending into orbit – a case report

I. V. Renuka · B. Srinivasa Rao · R. Sasank

Abstract Small Cell Neuroendocrine Carcinomas of the Sinonasal tract are extremely uncommon and distinct neoplasms with aggressive clinical behavior. They have similar morphological and immunohistochemical features to those of small cell carcinomas of the lung. They should be distinguished from Olfactory Neuroblastomas. We report a case of this rare entity.

A 46-year-old woman presented with a history of injury to the right eye with facial pain and proptosis as the primary symptoms. Later she developed nasal stuffiness. C.T. Scan revealed a space occupying lesion in the right nostril extending into ethmoid sinus and orbit. Functional endoscopic sinus surgery was done with decompression of the orbit. The biopsy material was obtained from ethmoid sinus and orbit. Histopathological examination of the biopsy specimen along with Immunohistochemical stains confirmed a Small Cell Neuroendocrine Carcinoma. Patient was treated with Radiotherapy and chemotherapy and responded well.

Keywords Small cell Neuroendocrine carcinoma · Olfactory Neuroblastoma · Small Cell Carcinoma · Paranasal Sinuses

Introduction

Small Cell Neuroendocrine Carcinomas of the Sinonasal tract are extremely uncommon and distinct neoplasms with aggressive clinical behavior. They are also referred to as Sinonasal neuroendocrine carcinoma (SNEC). They should be distinguished from Olfactory Neuroblastomas (ONB). SNEC resides within the less differentiated end of the spectrum of neuroendocrine tumors relative to ONB [1]. They have similar morphological and immunohistochemical features to those of small cell carcinomas of the lung. Here we report a case of SNEC in a 46 year old woman with primary orbital symptoms. FESS (Rt) and Medial Orbitotomy was preformed followed by radiotherapy and chemotherapy. The present case is being reported because of its rarity.

Case report

A 46-year-old woman presented with swelling, pain, proptosis of the right eye of 1 month duration. She gave a history of trauma to the right eye and later developed nasal stuffiness. On examination, there was chemosis of eyeball, swelling of eyelids, mild discoloration of upper and lower eyelids and proptosis. The vision of the right eye was decreased but the eyeball movements were normal. CT scan showed iso-dense space occupying lesions in right nostril, ethmoid sinus and orbit with destruction of lamina papyracea. The right eyeball was pushed laterally and anteriorly. Brain study was normal. Intraoperatively tumor was extensive and occupied the right middle and posterior ethmoid, and right frontal sinus extending into medial part of orbit. FESS (Rt) and Medial Orbitotomy was preformed and a major part of the tumor was removed.

Pathological findings: The tissue received were multiple grey white, soft friable bits measuring 3 cm size in total. Microscopic examination under low power showed a highly cellular and widely infiltrative tumor. Tumor cells were seen in large sheets and nests surrounded by fibrous septae.

I. V. Renuka¹ · B. S. Rao² · R. Sasank¹

¹Professor

²E.N.T Hospital, Guntur
Siddhartha Medical College,
Vijayawada,
Andhra Pradesh,
India

I. V. Renuka (✉)

Ph: +91 / 9848134166, 0866 2450626

e-mail: repriya56@hotmail.com

Cell nests were irregularly shaped varied in size from small aggregates with well defined margins to larger confluent sheets with extensive necrosis and haemorrhage. The individual tumor cells were small to intermediate size, with a high nucleocytoplasmic ratio and nucleoli were not seen. Nuclear molding and crush artifact were seen focally. High mitotic activity was seen 8–10/10hpf. No fibrillary background, ganglion cells, rosettes, calcifications, squamous or glandular differentiation were seen. Immunohistochemistry showed positivity for cytokeratin and chromogranin. The dual positivity for cytokeratin and chromogranin favors a diagnosis of small cell Neuroendocrine Carcinoma.

Fig. 1 Clinical photograph of the patient showing proptosis of right eye

Fig. 2 CT scan film of the tumor in right Nostril, ethmoid sinus and orbit

Fig. 3 Microphotograph showing irregular sheets of tumor cells separated by fibrous stroma (H&E x 100)

Fig. 4 Microphotograph showing cells with round to oval hyperchromatic nuclei with dense chromatin and scanty cytoplasm. (H&E x 400)

Discussion

SNEC occur in the same sites as ONB [superior nasal cavity / superior turbinate/ ethmoid]. However they may also arise from antra or middle turbinates- a location that would be unusual for ONB. Patients typically present with facial pain, epistaxis, and nasal obstruction and they may have orbital symptoms. Smaller lesions can be polypoid and confined to one nasal fossa. Larger tumours can erode the cribriform plate and may extend into the orbit and the cranium¹. In our case the patient had orbital symptoms primarily and later she developed nasal block. She had no evidence of intracranial extension or lymph node metastasis. In a series of 6 cases that were reported by Perez-Ordóñez et al only 2 cases had invasion of the orbit but they also presented with facial pain and epistaxis.

SNEC should be differentiated from ONB and Sinonasal Undifferentiated Carcinomas (SNUC), because of the

prognostic implication. Immunohistochemically ONB does not express epithelial differentiation. SNUC do not show neuroendocrine differentiation. SNEC show a dual positivity for both neuroendocrine and epithelial markers. Our case showed diffuse positivity for the neuroendocrine marker (chromogranin) and the epithelial marker (cytokeratin).

The distinction of SNEC from ONB is important since the latter has a better prognosis. The reported 5 to 10 year survival rates for ONB ranged from 64% to 77% [2, 3]. Silva et al reported that SNEC had a higher tendency to multiple recurrences (54%) than does ONB (25%) [4]. SNUC are uniformly high grade and usually fatal. Despite aggressive chemotherapy and radiotherapy the prognosis is poor. In SNEC the grade and survival rates are variable. In present case post operatively external radiation of 6250 Rads was given followed by 5 cycles of chemotherapy. The drugs used were Dacarbazine and Adriamycin was given. The patient responded well.

SNEC should also be distinguished from other Sinonasal Neuroendocrine lesions, including carcinoid tumor, pituitary adenoma and paraganglioma. The histological features in the present case did not have the features of these tumors. Other small cell non neuroendocrine lesions of the sinonasal tract which include Malignant Melanoma, Non Hodgkins lymphoma, Ewings sarcoma and Rhabdomyosarcoma were excluded based on the morphological features and immunohistochemistry.

SNEC is one of the extrapulmonary small cell carcinomas. Extrapulmonary small cell carcinomas are usually

fatal with a 13% 5yr survival rate [5]. In a study of extrapulmonary small cell carcinomas from the Mayo clinic, the median survival of 14 patients with primary head and neck tumors was only 14.5 months. This included 7 cases involving the paranasal sinuses ⁵.

In conclusion SNEC of the sinonasal tract can present primarily with orbital symptoms and should be distinguished from ONB and SNUC. It can have an aggressive clinical behavior, recurrence is frequent and prognosis is poor.

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