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CASE REPORT

Small-Cell Neuroendocrine Carcinoma of Nasopharynx

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ABSTRACT

Small-cell neuroendocrine carcinoma occurring in nasopharynx are extremely rare. Only three cases of small-cell neuroendocrine carcinoma occurring in nasopharynx have been reported uptodate. To our knowledge this is the fourth case of small-cell neuroendocrine carcinoma of the nasopharynx reported in world literature. We present this case for its rarity.

Keywords: Small-cell neuroendocrine carcinoma, Extrapulmonary, Nasopharynx.

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INTRODUCTION

Small-cell neuroendocrine carcinoma is frequently encountered in bronchogenic tissue. However, its occurrence in extrapulmonary sites is extremely rare accounting for about only 5% of all cases; these are commonly found in the esophagus, pancreas, skin, uterus and breast. A head and neck origin for small-cell carcinoma is even rarer. Both pulmonary and extrapulmonary small-cell carcinomas usually have a good response to chemotherapy initially, but a poor prognosis finally. We report here a case of small-cell carcinoma of the nasopharynx. The patient underwent chemotherapy and irradiation for the disease and is under follow-up.

CASE REPORT

We report a case of a 52-year-old male presenting with nasal blockage for 6 months associated with epistaxis on and off. The blockage was initially unilateral (right side), insidious in onset and gradually progressive. He also gave history of associated right-sided aural fullness. Nasal endoscopy revealed a growth in the nasopharynx. Otoscopic examination showed features of a right-sided serous otitis media. A biopsy was taken from the growth which was reported as small-cell carcinoma (Figs 1 and 2). A thorough investigation was done to rule out primary in the lungs or any other site. Epstein-Barr virus was shown to be absent by Epstein-Barr virus encoded RNA *in situ* hybridization, which further distinguished it from conventional nasopharyngeal carcinoma.

DISCUSSION

Extrapulmonary small-cell carcinomas are uncommon. In a series of 81 patients from the Mayo Clinic, 19 cases (23%) originated in the head and neck, of which six originated in the salivary glands, seven in the paranasal sinuses, four in the larynx, one in the trachea and one in the thyroid gland.[1] In addition, there were eight small-cell carcinoma cases originating in the esophagus. In other series, laryngeal small-cell cancer was more prevalent.[2]

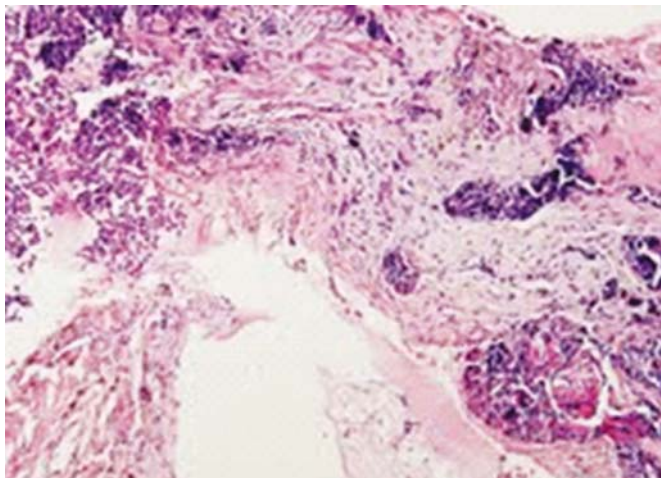

Fig. 1: Infiltrating small cells arranged in sheets and acini with perineural invasion (hematoxylin and eosin, ×100)

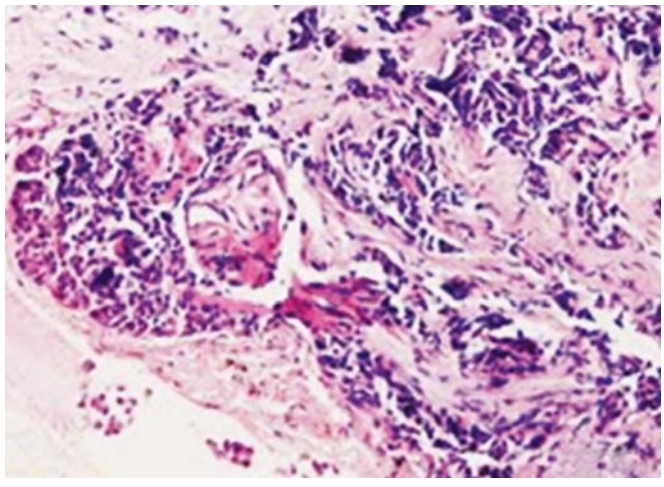

Fig. 2: Infiltrating small cells arranged in sheets and acini with perineural invasion (hematoxylin and eosin, ×400)

To the best of our knowledge only three cases of nasopharyngeal small-cell carcinoma have been reported.[3-5] Clinical features of this case and three previously reported cases when compared do not differ from conventional nasopharyngeal carcinoma, but the prognosis seems to be worse.

Small-cell carcinoma accounts for 10 to 20% of all primary malignancies of the tracheobronchial tree. It is

defined as high-grade epithelial neoplasm with neuroendocrine differentiation at both immunohistochemical and ultrastructural levels.[6] This highly lethal aggressive carcinoma is also referred as small-cell undifferentiated carcinoma, oat cell carcinoma, anaplastic carcinoma.[7,8] The majority of neuroendocrine neoplasm in the head and neck arise from larynx.[9,10] The laryngeal neuroendocrine tumors have an overall male predilection and the same seems to be true of nonlaryngeal neuroendocrine carcinomas of the head and neck. Neuroendocrine carcinoma of the head and neck may be stratified into a number of subsets on the basis of appearance under light microscopy, immunohistochemical staining, ultrastructural findings and clinical course.[6] These tumors have an epithelial or neural origin. The neural subgroup includes paraganglioma and epithelial subgroup includes typical carcinoids, atypical carcinoids and small-cell neuroendocrine carcinoma. The use of immunostains, electromicroscopy and molecular genetics has increased our understanding of this lesion, but the mainstay of diagnosis of this tumor remains light microscopy.[6,8] Differentiation from the more common nasopharyngeal squamous cell carcinoma is important for management and prognostic purposes. These tumors are often disseminated at diagnosis, thus it is important to perform thorough metastatic workup to detect regional and or distant metastases before initiating the treatment. The most common sites of spread are cervical lymph nodes, liver, lung and bone. Approximately 50% of patients have cervical metastasis on initial presentation.[7,8,10] However, in our patient there was no cervical or distant metastasis. Small-cell carcinoma of the head and neck is also notable for hematogenous dissemination.

Therapeutic options include various combinations of surgery, chemotherapy and radiotherapy.[6-8] Surgical results for this tumor have been disappointing and are reserved for cases of local relapse with no evidence of metastases.[9-11] Despite combined modality treatment prognosis is poor; the high likelihood of dissemination resulting low survival rates indicates that further advances in systemic therapy are desperately needed. Due to the rarity of these tumors, no specific treatment guidelines exist at present.

CONCLUSION

It is very important to distinguish highly lethal small-cell neuroendocrine carcinoma of nasopharynx from conventional nasopharyngeal squamous cell carcinoma which carries relatively good prognosis and good response to radiotherapy. A large number of case series with improved clinical, morphological and treatment response correlation studies are necessary to provide best treatment options.

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