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A Rare Localization Of Small-Cell Carcinoma In The Vallecula.

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ABSTRACT

Small-cell carcinoma (SCC) of the head and neck region is a rare and aggressive malignancy associated with poor prognosis due to its high metastatic potential and resistance to conventional therapies. Most cases originate in the larynx; involvement of the vallecula is exceedingly rare and sparsely documented in medical literature. We report a unique case of small-cell carcinoma located in the vallecula of a 51-year-old male. The diagnosis was confirmed through histopathological examination and immunohistochemistry (IHC) using CD56 marker positivity. This case underscores the importance of considering rare neuroendocrine tumors in the differential diagnosis of head and neck lesions and highlights the essential role of IHC in establishing a definitive diagnosis.

Keywords: Small-cell carcinoma, Vallecula, Neuroendocrine tumor, Head and neck cancer, CD56, Immunohistochemistry.

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INTRODUCTION

Head and neck cancers (HNCs) encompass a diverse group of malignancies that arise from various anatomical structures, including the oral cavity, pharynx, larynx, nasal cavity, paranasal sinuses, and salivary glands. Among these, squamous cell carcinoma (SCC) is the most prevalent histological subtype, accounting for nearly 90% of all HNCs, excluding non-melanoma skin cancers [1].

Small-cell carcinoma, also referred to as small-cell neuroendocrine carcinoma (SCNEC), is a rare and highly malignant tumor characterized by small, round-to-oval cells with scant cytoplasm, hyperchromatic nuclei, nuclear molding, and high mitotic activity. It represents approximately 0.3% of all head and neck cancers and is typically aggressive, with a high tendency for early metastasis and poor clinical outcomes [2]. While the lungs are the most common site of origin, extrapulmonary small-cell carcinomas (EPSCCs) can occur in various organs, including the gastrointestinal tract, genitourinary tract, and the head and neck region [3].

In the head and neck area, small-cell carcinomas most commonly arise in the larynx, particularly the supraglottic region. Less frequently, they may originate from the nasal cavity, paranasal sinuses, nasopharynx, salivary glands, and middle ear [4]. Involvement of the vallecula is extremely rare and has only been reported sporadically in the literature. Vallecular tumors often present late due to their deep anatomical location and lack of early symptoms, resulting in diagnostic challenges.

Given the rarity of this tumor in such an unusual site, it is important to document individual cases to improve awareness, aid in the identification of histological features, and guide treatment strategies. Here, we describe a rare case of small-cell carcinoma arising in the vallecula, confirmed by histopathology and CD56 immunopositivity, in a middle-aged male presenting with dysphagia and cervical swelling.

Review Of Literature

Small-cell carcinoma of the head and neck region is a well-established but rare entity, with most cases reported in the larynx. Vallecular involvement is even rarer. Ferlito et al [4] emphasized the aggressive nature and neuroendocrine features of such tumors. Chan [5] reviewed extrapulmonary small-cell carcinomas and noted their shared histopathological characteristics with pulmonary variants, along with their poor prognosis.

Patil et al [1] presented a case involving both the larynx and base of tongue with extensive local invasion, similar in behavior to our vallecular case. Wang et al [2] described a myoepithelial carcinoma at the vallecula, stressing diagnostic difficulty in that region. To date, only a handful of vallecular SCC cases have been described, making this case an important contribution to the literature.

Comparative review suggests that all reported cases presented with symptoms like dysphagia, neck swelling, or voice changes, with diagnosis reliant on biopsy and IHC. Treatment modalities varied, but most adopted chemotherapy and radiotherapy. Prognosis remained uniformly poor, supporting the need for early detection and aggressive management.

Case Presentation

A 51-year-old male presented to the otorhinolaryngology outpatient department with a 4-month history of progressive difficulty in swallowing, associated with a gradually enlarging swelling on the left side of the neck. There was no history of hoarseness of voice, cough, hemoptysis, or significant weight loss. The patient had no history of tobacco or alcohol use.

Physical examination revealed a firm, non-tender mass in the left cervical region, measuring approximately 3.5 x 2.5 cm. Indirect laryngoscopy showed an exophytic growth arising from the vallecular region. No lesions were seen in the vocal cords or adjacent structures.

A contrast-enhanced computed tomography (CECT) scan of the neck revealed a heterogeneously enhancing mass lesion at the base of the tongue involving the vallecula and extending into the pre-epiglottic space. Enlarged level II and III cervical lymph nodes were noted, raising suspicion of nodal metastasis.

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A biopsy was obtained from the vallecular mass was processed and stained with hematoxylin and eosin (H&E). Microscopy revealed a tumor composed predominantly of small round cells with high nuclear-to-cytoplasmic ratios. The cells were arranged diffusely in sheets, nests, as well as forming rosettes at places [Figure 1 and 2]. The nuclei were round to oval, with finely stippled chromatin and inconspicuous nucleoli. Nuclear molding was a prominent feature, suggestive of the aggressive nature of the tumor. Numerous mitotic figures and apoptotic bodies were seen, indicating high proliferative activity. Areas of necrosis and crush artifact were also noted, typical of small-cell morphology. The differential diagnosis included small-cell carcinoma, lymphoma, and other small round blue cell tumors.

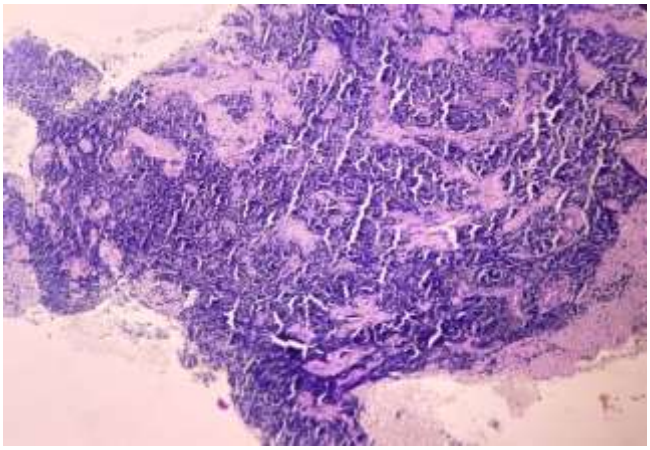

Figure 1: A tumor cells arranged diffusely in sheets and nests [H & E; 40X].

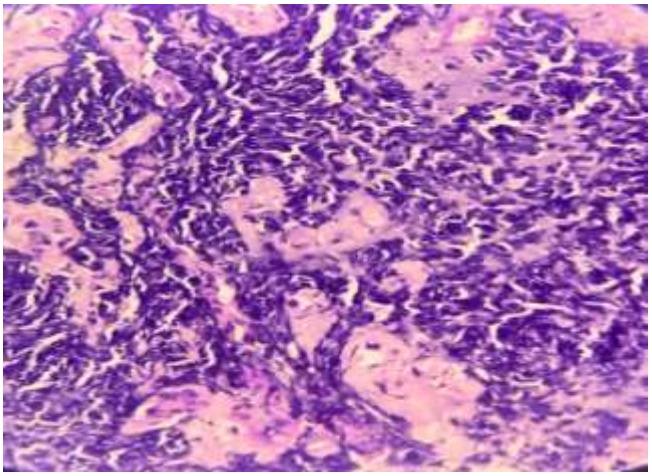

Figure 2: A tumor cells composed predominantly of small round cells with high nuclear-to-cytoplasmic ratios [H & E; 400X].

Given the overlapping histological features with other small round blue cell tumors, such as lymphoma, Merkel cell carcinoma, and poorly differentiated squamous cell carcinoma, immunohistochemistry was crucial for definitive diagnosis. The tumor cells exhibited strong membranous positivity for CD56, a neural cell adhesion molecule indicative of neuroendocrine differentiation [Figure 3].

Additional markers such as synaptophysin and chromogranin were considered but not performed due to resource constraints.

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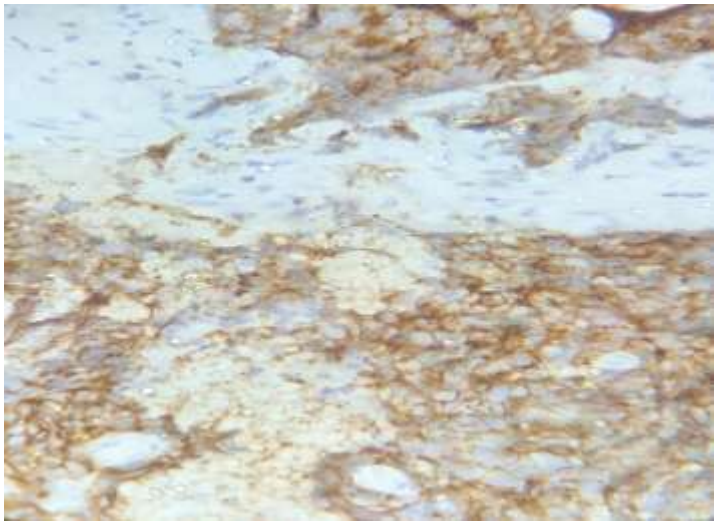

Figure 3: CD56 membranous positivity [IHC; 100X].

Based on the histomorphology and immunohistochemical findings, a final diagnosis of small-cell carcinoma of the vallecula was rendered.

DISCUSSION

Small-cell carcinoma of the vallecula is exceptionally rare, with limited cases reported in literature. It falls under the broader category of extrapulmonary small-cell carcinomas, which behave aggressively and have a predilection for rapid growth and early metastasis. SCCs of the head and neck share morphologic and immunohistochemical characteristics with pulmonary SCCs, including expression of neuroendocrine markers [4].

The pathogenesis of these tumors is not completely understood but is believed to involve transformation of multipotent basal epithelial cells or existing neuroendocrine cells. Various risk factors, including tobacco and alcohol use, human papillomavirus (HPV) infection, and prior radiation exposure, have been implicated in head and neck cancers but their role in SCC of the vallecula remains unclear due to the rarity of the condition.

Management of SCC in the head and neck is not standardized due to the low incidence. Therapeutic strategies are generally extrapolated from pulmonary SCC and involve multimodal approaches. Chemotherapy with etoposide and cisplatin is commonly used, often in combination with radiation therapy. Surgical excision is typically reserved for localized disease with no evidence of metastasis [5].

Prognosis remains poor, with most patients developing recurrence or distant metastases within a year of diagnosis. Long-term survival is rare, and the five-year survival rate is dismal. Close follow-up and comprehensive imaging are required to monitor treatment response and detect recurrence.

CONCLUSION

Small-cell carcinoma of the vallecula is a highly aggressive and rare malignancy that poses diagnostic and therapeutic challenges. Due to its rarity, awareness among clinicians and pathologists is essential for early recognition. Histopathological examination supported by immunohistochemistry is vital in establishing an accurate diagnosis. Treatment strategies must be tailored individually, often involving a combination of chemotherapy and radiotherapy. Documentation of such rare cases is essential to expand our understanding and improve management strategies.

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