

Case Report

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Spindle cell squamous cell carcinoma of postcricoid region of hypopharynx: a case report of a rare tumour in uncommon location with atypical presenting complaint

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ABSTRACT

Spindle cell carcinoma (SpCC) is an aggressive subtype of squamous cell carcinoma characterized by a biphasic pattern with both sarcomatous spindle cell and epithelial squamous cell components. It accounts for less than 3% of all head and neck cancers, with a male predominance, presenting in the 5th to 6th decades of life. The larynx is the most commonly affected site, with dysphagia being the most frequently presenting complaint. Hypopharyngeal involvement is uncommon, particularly in the postcricoid region. We present a 55-year-old male with a complaint of bilateral neck swellings without dysphagia. An exophytic growth on the postcricoid region of the hypopharynx was evident on laryngoscopy, and histopathology confirmed SpCC. A computed tomography (CT) scan showed necrotic cervical nodes, and chemotherapy was initiated for treatment.

Keywords: Spindle cell squamous cell carcinoma, SpCC, Sarcomatoid carcinoma, Hypopharynx, Post cricoid region

INTRODUCTION

Spindle cell carcinoma (SpCC) is a rare and aggressive subtype of squamous cell carcinoma (SCC), accounting for fewer than 3% of all head and neck cancers of epithelial origin.¹ Histopathologically, it demonstrates a biphasic pattern with a sarcomatous spindle cell and epithelial squamous component.

It has a male predominance and most commonly presents in the 5th to 6th decades of life, with dysphagia being the most frequent presenting complaint.^{2,3} The larynx is the most commonly affected site (0.5%), while hypopharyngeal involvement, particularly the post-cricoid region, is rare.⁴

We describe an unusual presentation of post-cricoid SpCC with cervical lymphadenopathy without dysphagia.

CASE REPORT

A 55-year-old male presented to the Department of Ear Nose Throat Head and Neck Surgery with complain of Bilateral Neck Swellings for the past 4 months which were gradual in onset slowly progressive, never disappeared, not associated with fever however associated with weight loss. It was worth noting that the patient did not have any complaints about dysphagia at the time of initial presentation. On examination 3×2 cm large lymph node was palpable on the right level II, which is non tender, hard, fixed with excoriation of the overlying skin. Similarly, another lymph node was palpable at the left level II which was about 2×2 cm having similar finding but without excoriation of the skin. Fiber optic direct laryngoscopy was performed as a part of routine check-up which showed exophytic growth on post cricoid area involving the arytenoid and posterior aspect of bilateral ary-epiglottic fold (Figure 1).

Computed tomography (CT) scan head and neck were advised which showed multiple enlarged necrotic cervical lymph nodes, involving levels II-IV, were seen bilaterally on the CT neck with contrast. The largest nodes measured 1.4×1.4 cm on the right (level IIB) and 1.4×1.7 cm on the left (level IIA). In addition to an asymmetrical and bulky left aryepiglottic fold, a hypodense lesion was observed in the post cricoid area. As a result, direct laryngoscopy and biopsy were performed (Figure 2).

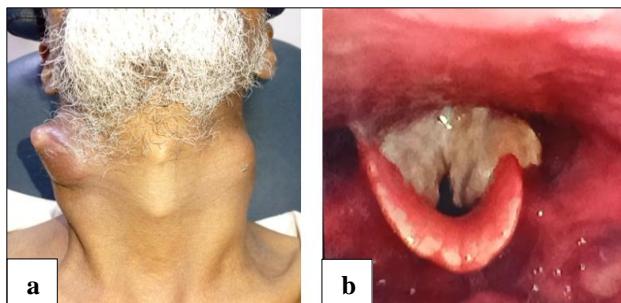


Figure 1 (a and b): A 3×2 cm hard, fixed, non-tender lymph node with excoriation of the overlying skin was palpable at the right level II. A similar 2×2 cm lymph node was noted at the left level II without skin excoriation. Fiber optic direct laryngoscopy revealed an exophytic growth on the post-cricoid area, extending to the arytenoid and posterior aspect of the bilateral ary-epiglottic folds.

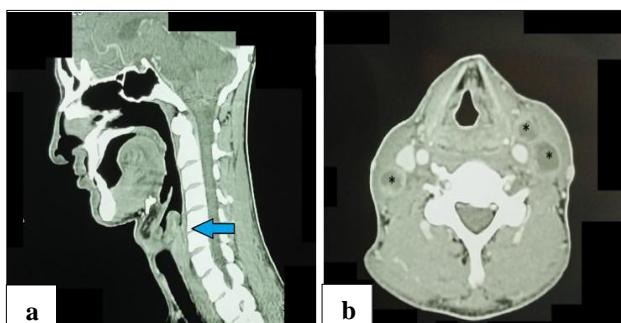


Figure 2 (a and b): CT scan of the neck showing multiple enlarged necrotic cervical lymph nodes, the largest measuring 1.4×1.4 cm (right level IIB) and 1.4×1.7 cm (left level IIA), with an asymmetrical bulky left aryepiglottic fold and a hypodense lesion in the post-cricoid region.

Histopathological report revealed stratified squamous epithelium showing infiltrating poorly differentiated neoplastic lesion arranged in the form of sheets and aggregates of spindly cells having abundant cytoplasm and hyperchromatic markedly pleomorphic nuclei showing frequent mitotic activity. Special stain mucin does not show mucin within tumor cells. Immunohistochemical stains performed show reactivity pattern in neoplastic cells for CK AE1/AE3: positive and p40: positive. All these findings suggest spindle cell carcinoma (sarcomatoid carcinoma). Hence, keeping in mind age and location the

patient was advised for chemoradiotherapy and currently under treatment (Figure 3).

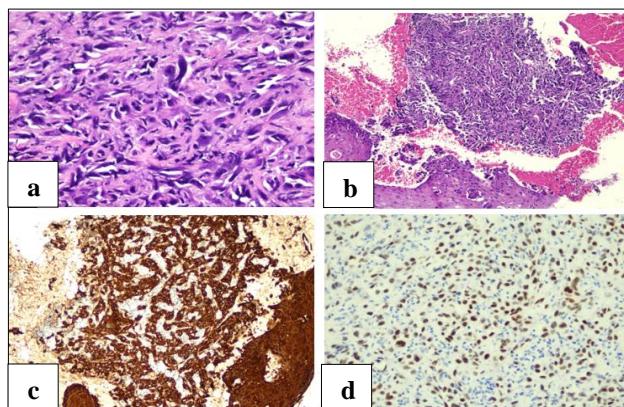


Figure 3 (a-d): Histopathology showing spindle-shaped neoplastic cells with pleomorphic nuclei and frequent mitoses; immunohistochemistry positive for CK AE1/AE3 and P40, confirming spindle cell (sarcomatoid) carcinoma.

DISCUSSION

SCC of the head and neck is the seventh most common cancer globally, with 450,000 deaths annually per GLOBOCAN estimates.⁵ It arises from squamous cells lining the oral cavity, pharynx, larynx, and other head and neck parts. Hypopharyngeal SCC is rare, with a global incidence of 0.8 per 100,000 population, and the highest burden is in South-Central Asia, accounting for an estimated 35,000 annual cancer-related deaths.⁶

Verrucous carcinoma, basaloid SCC, papillary SCC, spindle cell squamous carcinoma, adenosquamous carcinoma, and lymphoepithelial carcinoma are various subtypes of SCC, out of which many tumors now have established genetic profiles that are useful when considering them for differential diagnosis of complex cases.

SpCC, also called sarcomatoid carcinoma, is a rare and aggressive biphasic malignant tumor in which SCC has undergone epithelial-mesenchymal transition as per the World Health Organization (WHO) classification system.⁷ It is characterized by the presence of spindle-shaped epithelial (carcinomatous) and mesenchymal (sarcomatoid) cells. Studies have reported it to occur frequently in white people, with a peak incidence during the seventh decade of life. Predisposing risk factors include use of tobacco, alcohol consumption, smoking, radiation exposure, and poor oral health.

While the respiratory system is most commonly affected, particularly in the larynx and oral cavity, hypopharyngeal involvement is rare. In a 7-year review of 103 head and neck SpCC cases, 10.7% had hypopharynx involvement.⁸ Additionally, Olsen et al reported 9 hypopharyngeal cases out of 34 SpCC patients.⁹ In the literature, the pyriform

sinus is the most frequently reported site of hypopharynx, followed by the posterior pharyngeal wall, whereas postcricoid involvement is exceedingly rare, making this case clinically significant.

Patients with SpCC of the hypopharynx typically present symptoms like sore throat, hoarseness, foreign body sensation, dysphagia, or odynophagia, which progress to breathing difficulties, hemoptysis, and weight loss as the tumor grows. In the post-cricoid region, it can infiltrate the posterior cricoarytenoid muscle, resulting in vocal cord paralysis and airway obstruction. While bilateral neck swellings were the primary complaint of our patient, the absence of dysphagia and other typical symptoms was noted despite an extensive lesion. Suspicion for malignancy was raised due to the associated weight loss, but the unusual presentation made this case noteworthy.

SpCCs are considered more aggressive than conventional SCCs, contributing to their poor prognosis with a mortality rate of 35% in 2.5 years.¹⁰ Prognosis is worse for cervical metastasis and hypopharyngeal involvement. High rates of recurrence and metastasis have been shown, with approximately 25% regional lymph node and 5–15% distant metastases. In a review, lymph nodal metastasis at presentation was seen in 42% of patients.⁸

Histogenesis of SpCC has been widely discussed and is now regarded as a monoclonal epithelial neoplasm characterized by the presence of both spindle or pleomorphic cells and an SCC component. The spindle cell is derived from divergent mesenchymal differentiation of squamous epithelium and forms the largest portion of the tumor with a wispy and fasciculated pattern, whereas the squamous cell component comprises a minor portion.¹¹ Histopathological examination remains the cornerstone of diagnosis, indicating SpCC if both spindle and SCC components are present. If not, it must be differentiated from sarcomas, which are exceedingly rare in the hypopharynx, and inflammatory myofibroblastic tumors that have ALK, ROS1, and NTRK3 genetic alterations, which are absent in SpCC. According to the WHO classification, a malignant spindle cell tumor at the hypopharynx is considered spindle cell squamous carcinoma until proven otherwise. Poorly differentiated spindly neoplastic cells arranged in sheets and fascicles with marked pleomorphism, and frequent mitoses were typical microscopic features present in our case, and the absence of mucin staining excluded other sarcomatoid or glandular tumors.⁷

Immunohistochemical studies have demonstrated the epithelial origin of spindle cell elements by positive keratin immunostaining and the presence of desmosomes and tonofilaments in the cells, with CK being the most sensitive epithelial marker. It is used to differentiate SpCC from true sarcomas or other spindle cell neoplasms by confirming the epithelial origin in cases where only the spindle cell component is identified on histology. In our case, tumor cells expressed cytokeratin (CK AE1/AE3)

and p40, supporting the diagnosis and underscoring the importance of IHC in differential diagnosis, especially in atypical sites such as the postcricoid region.⁸

Management of SpCC is individualized according to the stage, size, and metastatic potential of the tumor.¹⁰ Surgical removal is preferred, which can be done promptly in low-stage tumors that are detected early. However, in advanced-stage tumors, adjunctive therapies, such as targeted radiation and systemic chemotherapy, have to be supplemented with surgery. Given the patient's age, tumor location, and bilateral nodal disease, chemoradiotherapy was advised.

CONCLUSION

Spindle cell squamous cell carcinoma of the hypopharynx is a rare and aggressive malignancy that is often presented at an advanced stage with extensive nodal disease. It poses a significant diagnostic challenge due to overlapping histopathological features with other spindle cell tumors. Histopathology supported by immunohistochemistry is crucial for accurate diagnosis and early multimodal intervention for reducing metastatic risk and improving outcomes. Unusual presentation with bilateral cervical lymph node swelling without dysphagia in this case adds to the scarce literature underscoring the importance of maintaining a high index of suspicion for hypopharyngeal malignancies in similar cases.

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