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SYNOVIAL SARCOMA OF LARYNX: A COMPREHENSIVE RARE CASE REPORT

Otorhinolaryngology

Rahul S. Gosavi Assistant Professor, Department Of ENT, Sanjeevan Medical Foundation, ENT PG Training Institute, Miraj, Sangli, IND

Anushka B. Patil* Junior Resident, Department Of ENT, Sanjeevan Medical Foundation, ENT PG Training Institute, Miraj, Sangli, IND *Corresponding Author

Vishal K. Magdum Associate Professor, Department Of ENT, Sanjeevan Medical Foundation, ENT PG Training Institute, Miraj, Sangli, IND

Vikas S. Gosavi Professor, Department Of Head And Neck Surgery, Sanjeevan Medical Foundation, ENT PG Training Institute, Miraj, Sangli, IND

ABSTRACT

Laryngeal synovial sarcoma is a rare form of malignancy in the head and neck region. Here we present a case of a 26-year-old young male with progressive difficulty in swallowing solids and mild breathing discomfort for a few months. Rigid video laryngoscopy revealed a globular mass in the post-arytenoid region. The patient underwent an elective tracheostomy, followed by direct laryngoscopy and biopsy, which confirmed the diagnosis on immunohistochemistry. Treatment included wide local excision of the tumour, reconstruction, and adjuvant radiation therapy. Follow-up indicates successful remission.

KEYWORDS

spindle cell neoplasm, larynx, neoplastic lesion, synovial sarcoma.

INTRODUCTION

Synovial sarcoma is an aggressive and rare soft tissue tumour originating from mesenchymal tissue. This malignancy accounts for approximately 5% to 10% of all soft tissue sarcomas and predominantly affects adolescents and young adults.[1] Primary synovial sarcomas occurring in the head and neck region are exceedingly uncommon, representing less than 10% of all synovial sarcoma cases. Within this region, the hypopharynx is the most frequently affected site, while laryngeal involvement is sporadic.[2]

The first documented case of laryngeal synovial sarcoma was reported by Gatti and Miller in 1975.[3,4]

A growing amount of research on cases of laryngeal synovial sarcoma has been published within the past four decades. Laryngeal synovial sarcomas usually manifest as a painless lump in the neck that is accompanied by airway symptoms, such as dysphonia, dyspnoea, and in more advanced cases, stridor. Tumour morphology, immunohistochemistry, and molecular investigations are used to make the final diagnosis, even when the imaging pathway for the initial investigation is still unknown. Histologically, the components of spindle and epithelial cells make up varying amounts of synovial sarcoma tumours. The proportionate presence of each component determines the subtype of tumour. Exclusively spindle cells or, less frequently, exclusively epithelial cells are found in monophasic tumours. Both spindle and epithelial cell components can be seen in biphasic tumours.[5]

At (X;18) (p11.2;q11.2) translocation involving the genes SS18 on chromosome 18 (which encodes SYT or SSXT protein) and SSX1, SSX2, and SSX4 on the X chromosome characterises synovial sarcoma as a translocation-associated sarcoma.[6]

The cornerstone of treating synovial sarcoma is wide local tumour resection; adjuvant radiation therapy and, less frequently, chemotherapy are also given to the affected area to lower the known high recurrence rate.[7] As a result, treating laryngeal synovial sarcoma poses a special set of difficulties for both patients and doctors, necessitating striking a balance between aggressive tumour treatments and attempts to maintain intrinsic laryngeal function. Prolonged neck radiation and laryngeal resections are frequently linked to long-term dysphagia, decreased breathing, and altered phonation, all of which can have a significant adverse impact on a patient's quality of life.[8]

In this report, we present the case of a 26-year-old male who experienced progressive dysphagia to solids and mild respiratory difficulty over a 2 to 3-month period. This case underscores the importance of considering rare malignancies in the differential diagnosis of laryngeal masses and highlights the role of timely intervention in achieving favourable clinical outcomes.

Case Report

A 26-year-old male presented to the ENT department of our hospital with a complaint of dysphagia to solids and slight difficulty in breathing for the past 2-3 months. The individual gave no history of weight loss, otalgia, haemoptysis or any other underlying disease. No history of tobacco chewing, gutakha chewing or smoking. Upon 70° rigid video laryngoscopy, a post-arytenoid solitary globular smooth swelling was seen obstructing the laryngeal inlet (Fig. 1). Vocal cords were not visible.

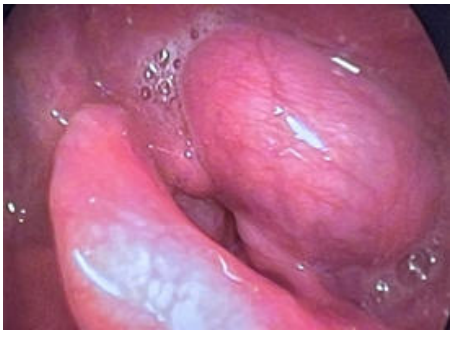

Figure 1: 70° rigid video laryngoscopy

Due to the obstruction of the glottic opening and signs indicating an impending stridor, it was decided to proceed with an elective tracheostomy. This was followed by direct laryngoscopy and biopsy, after a thorough discussion with the patient, to confirm the diagnosis.

A computed tomography (CT) scan (Fig. 2) proved valuable in determining the site of origin and extent of the heterogeneously enhancing lesion involving supraglottic, post-cricoid, and cervical oesophagus, suggestive of a neoplastic lesion with a tracheostomy tube in situ.

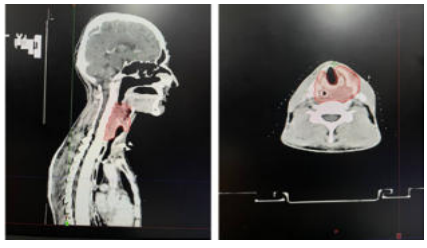

Figure 2: CT neck (left- sagittal view, right- axial view)

Histopathological examination (HPE) revealed a spindle-cell neoplasm (Fig.3a) with differential diagnosis of synovial sarcoma and spindle cell tumour of neurogenic origin, post which immunohistochemistry (IHC) was advised. Upon IHC, tumour cells expressed TLE1 (Fig. 3b), BCL2 (Fig.3c), Calponin and Pancytokeratin (focal) and were immunonegative for Cd34, S100, Desmin and SMA. Hence, a diagnosis of synovial sarcoma was rendered.

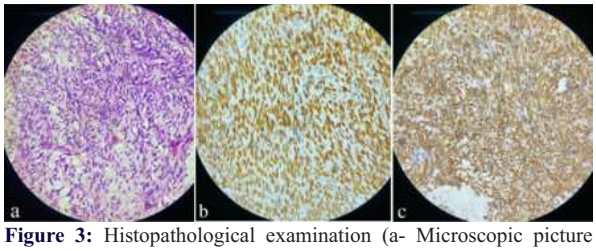

Figure 3: Histopathological examination (a- Microscopic picture showing a spindle-cell neoplasm; b - On IHC, tumour cells expressed TLE1; c - On IHC, tumour cells expressed BCL2.)

Metastatic workup showed no distant metastasis.

Surgery included wide local excision and reconstruction (the raw area was covered with local mucosal flap) (Fig. 4a,b,c,d). Phonation, swallowing and breathing were preserved successfully. The HPE of the excised specimen was the same as that of the previous biopsy specimen, i.e. synovial sarcoma. Histopathology revealed the tumour size to be 5.3*3.4*3.3 cm. Tumour reached the inked deep surgical margin and medial surgical margin.

Figure 4: Intraoperative images (a,b - Surgical wide local excision; c - reconstruction; d - excised specimen.)

Since the patient showed no signs of breathing difficulty post-surgery, slow weaning of the tracheostomy tube was started, and tracheostomy closure was done after 3 weeks. The patient was advised to take adjuvant radiotherapy. Radiation treatment was given through External Beam Radiotherapy on a Linear Accelerator machine during which 35 cycles of 70 Gy dose were administered.

He is alive in remission at 1 year 3 months.

DISCUSSION

Usually affecting the lower limbs, synovial sarcoma is an aggressive malignant soft tissue tumour believed to develop from pluripotent mesenchymal cells.[9] Sarcomas occur uncommonly in the head and neck region in adults, and synovial sarcoma is extremely rare in this site. Squamous cell carcinoma accounts for over 90% of all laryngeal cancers,[9] with fewer than 40 cases reported to date.[5]

Gatti and Miller described the first laryngeal case in 1975.[3,4]

Head and neck sarcoma typically manifests as a painless submucosal or subcutaneous lump, with symptoms differing depending on the site. Sarcomas in this area, depending on their size, present with symptoms due to mechanical interference with function, much like primary squamous cell carcinomas of the larynx. Usually, the first sign is hoarseness, followed by stridor and dyspnoea. Usually, dysphagia only happens when the tumour grows sufficiently to intrude into the hypopharynx.[10] In our case, dysphagia and mild respiratory difficulty prompted laryngoscopy, which revealed a post-arytenoid mass. Given the unusual location, histopathological evaluation was essential.

Biphasic and monophasic are the two main histologic subtypes of synovial sarcomas. The monophasic spindle cell type, like the one we have, can mimic other sarcomas; hence, immunohistochemistry is required for a conclusive diagnosis.[11] Tumour markers such as TLE1, BCL2, cytokeratin, and vimentin confirm the diagnosis.[5] Our case was positive for TLE1 and Bcl-2. A t(X;18) that represents the fusion of SYT (at 18q11) with either SSX1 or SSX2 (both at Xp11) is found in nearly all of them. Genetic testing is particularly useful in poorly differentiated tumours.[11]

Multimodal treatment is the best approach to treat synovial sarcoma. The size, location, and grade of laryngeal sarcoma determine the course of treatment. It is widely acknowledged that the cornerstone of treatment is radical surgical resection. To reduce the risk of local recurrence, radiotherapy (RT) is a crucial adjuvant in the management of soft tissue sarcoma.[12] High-grade lesions, positive surgical margins, larger tumours (>5 cm), and recurrent lesions are the primary indications for postoperative radiotherapy.[13] Our patient underwent wide local excision and adjuvant radiation.

There have been cases of high-grade synovial sarcoma treated with adjuvant chemotherapy. In the treatment of soft tissue sarcomas, ifosfamide and doxorubicin have been demonstrated to increase disease-specific survival.[1,14]

Thus, patients and clinicians face particular problems in managing synovial sarcoma, which necessitates a delicate balancing act between aggressive tumour treatment and preserving intrinsic laryngeal function.

At 1 year 3 months post-treatment, our patient remains in remission, highlighting the importance of early detection and multidisciplinary care.

CONCLUSION

Synovial sarcoma of the larynx is an exceedingly rare and aggressive neoplasm, often posing diagnostic and therapeutic challenges. Through this case report of a 26-year-old male presenting with dysphagia and breathing difficulties, we highlight the importance of a comprehensive approach involving clinical assessment, histopathology, and immunohistochemistry for accurate diagnosis. Early intervention with wide local excision and adjuvant radiotherapy, as demonstrated in this case, can effectively preserve vital functions like phonation and swallowing while minimising the risk of recurrence. Though the prognosis of synovial sarcoma remains guarded due to its aggressive nature, a multidisciplinary approach, as outlined in this report, offers promising outcomes. Continued research and accumulation of similar case reports are essential for improving our understanding and management of this rare malignancy.

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