Spindle Cell Sarcoma of Pharynx: A Rare Case with Successful Management

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Lalit Ray,[1] Aritro Bhattacharji,[2] Sabyasachi Chakravarty[3]

ABSTRACT

Introduction

Spindle cell sarcoma is a rare and aggressive malignancy characterized by spindle-shaped tumour cells. It can arise from various anatomical sites from soft tissue or bone. Involvement of the pharynx is relatively rare. This case report presents a unique case of spindle cell sarcoma in the pharynx, initially misdiagnosed as a vallecular space-occupying lesion on Fiber Optic Laryngoscopy but later confirmed as arising from the lateral wall of pharynx on HRCT. Accurate diagnosis and prompt management are essential for optimizing patient outcomes.

Case Report

A 57-year-old male patient presented to the ENT clinic with a two-month history of stridor and difficulty in swallowing. The patient had no significant medical or surgical history. Fibreoptic laryngoscopy revealed a large vallecular space-occupying lesion obstructing the airway and impeding deglutition. The patient was scheduled for surgery after a thorough clinical and radiological evaluation. Contrast enhanced CT scan reveals SOL arising from the left side of pharynx with contrast uptake.

Discussion

Spindle cell sarcoma of the pharynx is a rare entity with limited reported cases. It poses a diagnostic challenge due to its atypical presentation and overlapping histopathological features with other spindle cell tumours. Immuno histochemistry plays a crucial role in confirming the diagnosis by evaluating specific markers. IHC, P-16 is a specific marker differentiating peripheral nerve sheath tumour from spindle cell sarcoma. Surgical excision remains the primary treatment modality, often combined with adjuvant therapy depending on the stage and extent of the disease.

Keywords

Spindle Cell Sarcoma; Vallecular Mass; Malignant Peripheral Nerve Sheath Tumour; Immunohistochemistry

Spindle cell sarcoma is a rare and aggressive malignancy characterized by spindle-shaped tumour cells. It can arise from various anatomical sites in the soft tissue like rarely Tonsillar area[5] or bone usually affecting long bones in the arms[1]. This case report presents a unique case of spindle cell sarcoma in the pharynx initially misdiagnosed as avallecular space-occupying lesion on FOL but later confirmed as arising from lateral wall of pharynx on HRCT.

The cancer typically starts in the connective tissue layers, such as those between muscles or the surrounding organs.[1,2] Accurate diagnosis and prompt management are essential for optimizing patient outcomes. Understanding the cell type and characteristics allows healthcare providers to better understand the probability of the spread of cancer. It can also identify the best possible treatments.

Case Report

A 57-year-old male patient presented to the ENT clinic with a two-month history of stridor and difficulty in swallowing. The patient had no significant medical or surgical history. Fibreoptic laryngoscopy revealed a large vallecular space-occupying lesion obstructing the airway and impeding deglutition. The patient was scheduled for surgery after a thorough clinical and radiological evaluation. Contrast enhanced CT scan reveals SOL

1 - Department of ENT, Midwest Hospital, West Bengal
2 - Department of ENT, Asansol District and Super-speciality Hospital, West Bengal-713303
3 - Department of ENT, Gouri Devi Medical College, West Bengal
Corresponding author:
Dr. Lalit Ray
email : lalitray66@gmail.com

arising from the left side of pharynx with contrast uptake.

The patient underwent an intraoral excision of the mass and pre op tracheostomy under general anaesthesia as intubation difficulty was anticipated. The surgical team achieved adequate exposure and excised the lesion completely. A tracheostomy was performed to secure the airway during the pre-operative period. Endoscopic endo-laryngeal approach was taken and the mass was removed with radiofrequency ablation using suction diathermy. The procedure was well-tolerated and the patient was transferred to the recovery unit for further monitoring. Decannulation was done on the 2nd post operative day.

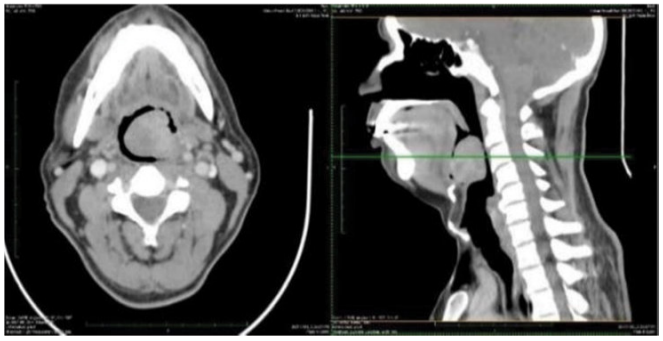

Fig. 1. Pre-Operative Contrast Enhanced Ct Scan Of Neck Showing Mass Is Arising From Lateral Wall Of Pharynx

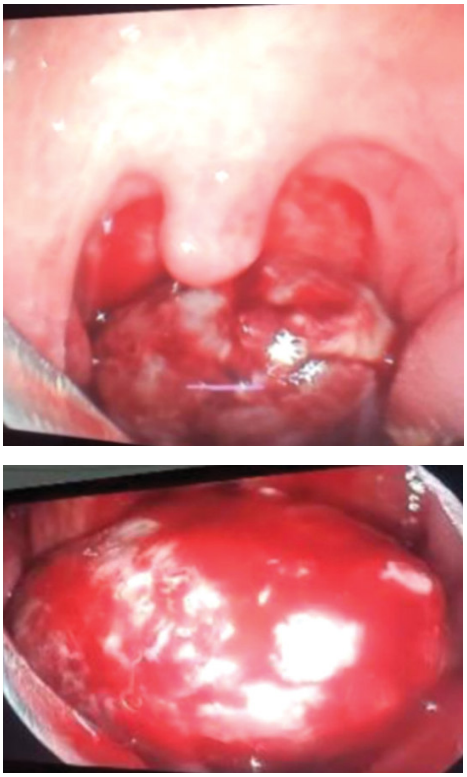

Fig. 2. Conventional D/L Scope With In Built 30 Degree Endoscope and Monitor showing a huge pre operative mass in the vallecular region

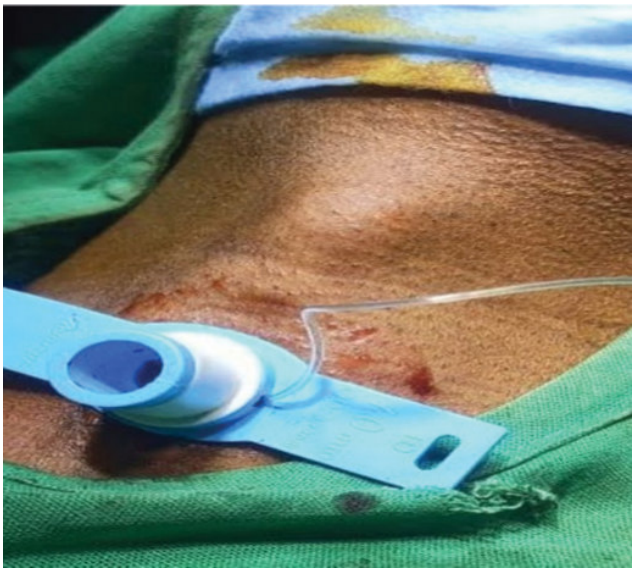

Fig. 3. Intra Operative Image

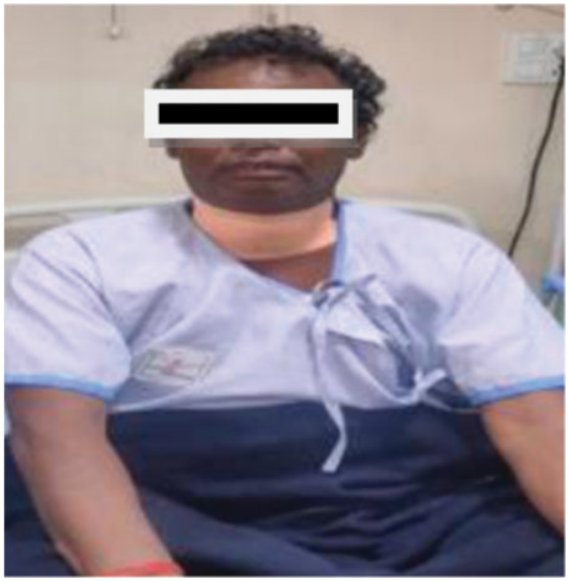

Fig. 4. Post operative image

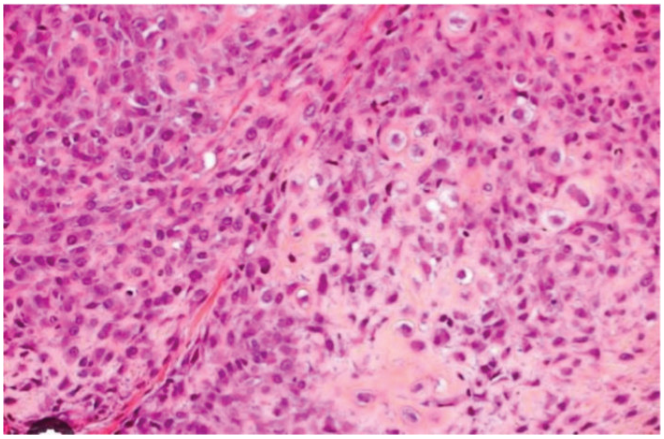

Fig. 5. Histopathology with 40x magnification using H & E stain revealed a high-grade spindle cell sarcoma consisting of elongated cells with ovoid nuclei and variable amount of cytoplasm

Immuno histochemical analysis demonstrated positive staining for vimentin, S100, EGFR, and p16[4,5]. These findings supported the diagnosis of spindle cell sarcoma. Two IHC reports have been done to emphasise and clear the diagnostic dilemma. One report is suggestive of malignant peripheral nerve sheath tumour whereas the other report suggests spindle cell sarcoma with myofibroblast differentiation revealed malignant peripheral nerve sheath tumour. However HPE and IHC done in another institute reveals spindle cell sarcoma and thus we confirmed it as Spindle cell Sarcoma.Following surgery, the patient received comprehensive postoperative care including pain management, tracheostomy care and enteral nutrition support. The patient's progress was closely monitored for any signs of complications or recurrence. The patient showed gradual improvement in symptoms postoperatively. Regular follow-up visits were scheduled to monitor for disease recurrence and to provide necessary supportive care.

Discussion

Spindle cell sarcoma of the pharynx is an extremely rare entity with limited reported cases[2]. It poses a diagnostic challenge due to its atypical presentation and overlapping histopathological features with other spindle cell tumours.[3]

Incidence of Spindle cell carcinoma in the head and neck region is 3% of all carcinomas and amongst them Spindle cell sarcoma in the pharynx is so rare that relevant incidence data is not known in the literature.[4]

Immuno histochemistry plays a crucial role in confirming the diagnosis by evaluating specific markers.[2] In this case, positive staining for vimentin, S100, EGFR and p16[4,5] supported the diagnosis of spindle cell sarcoma. IHC, P-16 is a specific marker differentiating peripheral nerve sheath tumour from spindle cell sarcoma. Surgical excision remains the primary treatment modality often combined with adjuvant therapy depending on the stage and extent of the disease.

However precautions related to the Anatomical site must be kept in mind. In our case, since the mass inloved pharynx, so preoperative Tracheostomy was done in order to protect the airway. Later on IHC and histopatholgy proved the case and the patient was treated adequately.

Conclusion

Spindle cell Sarcoma involving the pharynx is an extremely rare condition as per incidence. Importance of accurate diagnosis through histopathological examination and immune histochemical analysis is very ceratin[1,3]. Surgical excision followed by appropriate adjuvant therapy remains the cornerstone of treatment[5]. Further research and studies are needed to better understand the pathogenesis, prognosis, and treatment and it remains as a diagnostic dilemma.

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