

Synovial Cell Sarcoma of the Larynx

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KEYWORDS

• Synovial cell sarcoma • Ifosfamide • Larynx • Head neck

KEY POINTS

- Synovial sarcoma of the larynx represents a rare entity, with challenging diagnosis and management issues.
- A multidisciplinary therapeutic approach is essential for management of this malignancy.
- Long-term follow-up is essential to monitor for recurrence and improve disease-free survival.

INTRODUCTION

Synovial cell sarcomas are rare, aggressive, mesenchymal malignancies constituting a high-grade histologic variety of sarcomas. These tumors represent the fourth most common type of sarcoma after malignant fibrous histiocytomas, liposarcomas, and rhabdomyosarcomas.¹ Synovial cell sarcomas predominantly affect soft tissues of the extremities such as large joints, bursal structures, and tendon sheaths,² particularly in adolescents, among whom they account for 10% of all soft-tissue sarcomas.³ This malignancy has preponderance in males with a male/female ratio of 1:2.⁴ Approximately 3% of synovial cell sarcomas can also occur in the head and neck region, which is poor in synovioblastic tissue,⁴ and fewer than 100 cases in this region have been described in the literature to date. It has been reported to occur in various sites within the head and neck, most commonly the hypopharynx.⁵ Other sites include the oropharynx and soft tissues of the neck. However, synovial cell sarcomas of the larynx are extremely rare, with only a handful of cases reported to date.^{6–14} As a result it is not often diagnosed until later in the course of the disease, which therefore affects management and prognosis. Being such a rare entity, the treatment of head and neck synovial cell sarcomas is not well researched and classified.

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EPIDEMIOLOGY

Synovial cell sarcoma is a rare malignancy with an incidence of 2.75 per 100,000.¹⁵ The incidence in the head and neck is significantly lower, with approximately 80 cases of head and neck synovial sarcomas reported to date.¹⁶ Laryngeal synovial cell sarcomas are extremely rare, with about 20 cases reported in the literature.^{6-14,17-26}

Synovial cell sarcoma is primarily a malignancy affecting adults in the third to fifth decade of life. In the head and neck region, synovial cell sarcoma is noted to occur during this age range as well, although it can also occur outside of this range. Of the specific cases of laryngeal synovial cell sarcoma, the youngest known patient was 14¹² and the oldest 79 years old.²⁶ Furthermore, males are predisposed to this malignancy more than females, with a male/female ratio of 2:1.

ETIOLOGY

Synovial sarcomas are thought to arise from pluripotent mesenchymal cells that have the ability to differentiate into epithelial and mesenchymal lineages.²⁷ Despite the name, synovial cell sarcomas can arise from cells outside of the synovium as exemplified by synovial cell sarcomas arising in the parapharyngeal, hypopharyngeal, or laryngeal regions.²⁸

The underlying defect in synovial cell sarcoma is primarily a specific chromosomal translocation t(X;18)(p11;q11). This translocation creates a so-called fusion gene *SYT-SSX1*, *SYT-SSX2*, or *SYT-SSX4*.¹ The SYT component arises from chromosome 18 and the SSX gene arises from chromosome X.

PATHOGENESIS

The pathogenesis of this tumor involves the creation of aberrant transcriptional regulators resulting in the inhibition of tumor suppressor genes or the activation of proto-oncogenes. The SYT component (chromosome 18) fuses with homologous genes at Xp11 with 1 of 3 sites: *SSX1*, *SSX2*, or *SSX4*.¹ Monophasic tumors may have any of the transcripts, but biphasic tumors predominantly express the SYT-SSX1 transcript.

PATHOLOGIC FINDINGS

Macroscopic Pathology

On gross appearance, no distinguishing characteristics have been described for synovial cell sarcomas. The tumors may have a gray-white almost ivory appearance with an oleaginous texture on palpation. In the head and neck, synovial cell sarcomas most commonly occur in the hypopharynx and very rarely in the larynx. Laryngeal synovial cell sarcomas have been known to occur in the supraglottis, glottis, and subglottis.^{10,11,14} Macroscopically, synovial cell sarcomas may have a cystic, solid, or necrotic appearance.²⁹ In the upper airways, these cancers may appear as exophytic lesions with superficial ulceration.²⁵ **Fig. 1** depicts the gross appearance of a laryngeal synovial cell sarcoma encountered by the authors.

Histopathology

Up to 3 subtypes of synovial cell sarcomas have been described in the literature: monophasic, biphasic, and poorly differentiated.

The monophasic and biphasic subtypes are the 2 main histologic subtypes of synovial cell sarcomas. Histologically both subtypes have proliferating spindle cells and branching, dilated, thin-walled blood vessels (hemangiopericytomatous pattern)

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