Synovial Sarcoma Oropharynx - A Case Report and Review of Literature

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Introduction

Synovial sarcoma is a rare malignant tumor that derives from a mesenchymal precursor stem cell that is unrelated to mature synovial tissue. It is primarily a disease of young adults typically arising in the deep soft tissues of the extremities, most commonly, the lower thigh-knee region. Reports in the literature have underscored the occurrence of this tumor in unusual sites, many seemingly distant from synovium-lined spaces, particularly, the abdominal wall [1] and head and neck region. The vast majority of this latter group, mostly case reports, were confined to the cervical and parapharyngeal region [2–8]. In this article, we report a case of synovial sarcoma that originated in oropharynx which is extremely rare. This is the first of its kind at our institute.

Case Report

A 40-year-old man presented with a history of foreign body sensation in the throat mainly on the left side & progressively worsening dysphagia and nasal regurgitation. He did not have other symptoms relating to the ear, nose and throat. He had no

history of a loss of weight or appetite. He had given up smoking 7 years earlier.

Examination revealed the presence of a pedunculated mucosal lesion projecting into oropharynx arising from just behind the posterior pillar of tonsil on left side. No cervical lymphadenopathy was noted. Findings on the remainder of the ENT examination were normal. A lateral neck x-ray demonstrated an abnormal protrusion of soft tissue from the lateral wall of oropharynx on left side.

The patient underwent a pharyngoscopy showing well demarcated globular mass of 4×3 cm arising from left lateral pharyngeal wall just behind posterior pillar of tonsil. Computed Tomography scan revealed a lobulated homogeneously enhancing soft tissue density mass lesion in left lateral pharyngeal wall extending from tip of epiglottis to hyoid bone. The mass was removed as an excisional biopsy by transoral endoscopic assisted approach under general anaesthesia (Figs. 1 and 2).

Histo-pathologic examination showed biphasic tumour with slit like spaces lined by *epithelial cells and Spindle cells*. Epithelial cells were cuboidal and pale with vesicular nuclei and *spindle cells* were in fascicles and bundles, closely approximated to epithelial and glandular spaces (Fig. 3) consistent with *synovial sarcoma/carcinosarcoma*. Excision of lesion was deemed complete. Immunohistochemistry highlighted spindle cells strongly expresses for *vimentin*, focal weak cytoplasmic positive for *bcl-2*, *pancytokeratin* positive in epithelial cells and *CD99* was negative consistent with *Synovial sarcoma/Carcino sarcoma*. It was subjected to polymerase chain reaction technique for t(X; 18) (p11.2; q11.2) SYT-SSX analysis. Results turned positive for *SSX1* and *SYT genes* and confirmed as biphasic synovial sarcoma [9].

Patient received adjuvant radiotherapy 66 Gy in 33 fractions. Case was followed up for 3 years at regular intervals for both local and metastatic work up and is free of any symptoms. Residual or recurrent mass ruled out by VLS.

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Figs. 1 and 2 CT scan showing lobulated homogeneously enhancing soft tissue density mass lesion in left lateral pharyngeal wall extending from level of epiglottis to hyoid bone in axial view (1) and coronal view (2)

Fig. 3 HPE: biphasic tumour with slit like spaces lined by epithelial cells and spindle cells in fascicles and bundles, closely approximated to epithelial and glandular spaces

Discussion

Synovial sarcoma is a malignant soft-tissue neoplasm that arises from primitive pluripotential mesenchymal cells near to or remote from articular surfaces. Other terms used to describe this entity are synovial cell sarcoma and malignant synovioma [9, 10]. Although the term synovioma implies a benign condition, further investigations have exposed the aggressive nature of this neoplasm and eventually led to its classification as a high-grade tumor by the American Joint Committee on Cancer [11].

Synovial sarcomas account for 8 to 10 % of all sarcomas [2]. Although these tumors occur predominantly in the extremities, they are also known to arise in the head and neck region [9]. Since the first description of a head and neck synovial sarcoma of hypopharynx by Jernstrom [3] in 1954, fewer than 90 cases have been reported in the literature, and they account for 3 to 10 % of all reported cases [12].

The most common site of synovial sarcoma of the head and neck is the hypopharynx [4, 10]. These tumors have been encountered in other areas of the head and neck as well, including prevertebral and parapharyngeal areas, pharyngeal, laryngeal, nasopharynx, soft palate, tongue, maxillofacial region, mandible corner, sternoclavicular region, scapular region and the cervical oesophagus [5]. Synovial sarcoma classically affects patients between the ages of 15 and 40 years [6], and the proportion of male-to-female patients is 3:2 [7]. Patients with synovial sarcoma of the head and neck tend to be younger than those who have such tumors in the extremities [5, 12].

The tumor usually appears as an asymptomatic mass until it attains sufficient volume to cause pressure effects on neighbouring structures. This process is usually more rapid in patients with head and neck synovial sarcoma than in those whose tumor is in an extremity. Lockey reported that patients with head and neck tumors were diagnosed an average of 20 months earlier than those with extremity tumors [12]. Head and neck synovial sarcomas have better prognosis because of their slow growth rate and lower rate of lymph node metastasis. Pai et al. reported that survival in cases of neck disease was higher than that in cases of limb tumors [8]. On the other hand, Kartha and Bumpous found that the 5-year survival rate for patients with head and neck synovial sarcoma was 40 %, which is substantially lower than the 60 % 5-year survival rate reported in the literature for synovial sarcoma at all other sites [13].

Histologically, three variants of synovial sarcoma have been described: biphasic, monophasic fibrous, and monophasic epithelial. Although monophasic forms of synovial sarcoma have been reported, its primary histopathologic feature is biphasic cellularity (Fig. 3) [14]. Microscopically, the tumor is made up of epithelial and sarcomatous components. Epithelial areas are present in the form of glands lined by cuboidal or columnar cells

or as solid nests as was seen in this case. Sarcomatous component is made up of hypercellular spindle cells with fibroblast-like cells or hemangiopericytoma-like areas. Extensive sampling is required to display epithelial features. Synovial sarcoma with only spindle cells or purely glandular component is called monophasic synovial sarcoma. Calcifying Synovial sarcoma is another subtype, which has extensive calcification and carries a much better prognosis. Histologically, no tumour in the body resembles biphasic Synovial sarcoma. However, monophasic Synovial sarcoma simulates fibro sarcoma, malignant peripheral nerve sheath tumour and hemangiopericytoma. Reticulin stain highlights the imperceptible clusters of epithelial cells. Immunohistochemically S-100, CD-99, bcl-2 and vimentin positivity is seen in the spindle cells of Synovial sarcoma. Cytokeratin expression is seen in 90 % of all Synovial sarcoma both in the epithelial and a few cells of the sarcomatous component. Specific expression of Cytokeratin subtypes 7 and 19 along with EMA is observed, and expression of epithelial markers in the epithelial as well as sarcomatous element excludes the possibility of Ewing's sarcoma and Malignant peripheral nerve sheath tumour, which may also show positivity for CD-99 and S-100 on immunohistochemistry in the spindle cell areas. Ultrastructurally, true glandular epithelial features are seen in epithelial areas. Spindle cell areas also show subtle epithelial features. The t (x;18)(p11;q11) is the cytogenetic hallmark of synovial sarcoma. FISH and RT-PCR have been employed for the rapid diagnosis of Synovial sarcoma [14].

Preoperatively, computed tomography and magnetic resonance imaging are the investigations of choice to determine the location and extent of the tumor.

Small number of patients with synovial sarcoma of the head and neck precludes us from drawing firm conclusions with respect to a precise treatment protocol. The primary mode of treatment is wide surgical excision [10] that includes a normal-appearing tissue cuff in order to obtain tumor-negative margins. However, this cannot always be accomplished in head and neck area because of the proximity of the tumor to vital structures. The results of multimodality treatment appear to be better than those seen with any single-modality treatment or with any form of salvage therapy [12, 15]. The role of radiotherapy is confined to reducing the risk of local recurrence following resection; radiation does not improve long-term survival [16]. The role of chemotherapy is controversial [17], although Lenoir et al. [18] reported a dramatic regression in tumor size in a 15-month-old patient with a retropharyngeal synovial sarcoma after treatment with a combination of ifosfamide, vincristine, and actinomycin D. The future role of both radiotherapy and chemotherapy in the management of synovial sarcoma needs to be determined by a prospective study.

Favourable prognostic indicators include an early diagnosis, small tumor size (<5 cm), younger age (<20 years), distal location of tumor, mitosis <15/10 HPF, calcification and a wide surgical excision [16, 17, 19]. However, necrosis, poorly

differentiating areas and aneuploidy are worse prognostic indicators [14].

We presented this case because of its rarity in its presentation as an oropharyngeal synovial sarcoma, not more than ten cases reported in the world literature as far as our knowledge [19–23].

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