

Synovial Cell Sarcoma of the Hypopharynx

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ABSTRACT

Synovial sarcomas of head and neck are very rare and account for only 3% of all sarcomas. Approximately 5% of synovial sarcomas arise in the head and neck region and hypopharynx and larynx are the most and least often affected anatomic sites respectively. We describe a rare case of primary hypopharyngeal synovial sarcoma in a young adult.

Keywords: Synovial cell, Head and neck, Sarcoma.

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INTRODUCTION

Sarcomas are malignant soft tissue tumors accounting for only 1% of all malignant tumors and represent 1% of all head and neck malignancies.¹ Synovial cell sarcoma (SS) is a mesenchymal spindle cell tumor which displays variable epithelial differentiation, including glandular transformation and has a specific chromosomal translocation t(X;18) (p11;q11).² SS usually involves the lower limbs of young adults with a peak incidence in those aged 20 to 40 years and males are affected twice as often as females.³ Synovial sarcomas of head and neck are very rare and account for only 3% of all sarcomas.⁴ Approximately 5% of synovial sarcomas arise in the head and neck region. Hypopharynx and larynx are the most and least often affected anatomic sites respectively.⁶ We describe a rare case of monophasic synovial sarcoma treated with palliative chemotherapy and radiation.

CASE REPORT

A 22-year-old male was seen in our hospital in January 2007 with a 3 months history of lump left neck, progressive dysphagia for 2 months and worsening shortness of breath in the past 4 weeks. A biopsy of the neck mass done elsewhere was reported as inflammatory myofibroblastic tumor.

Magnetic resonance imaging (MRI) neck (Fig. 1) demonstrated a large soft tissue mass arising from the posterocricoid location extending cranially up to the cricoid level and involving the left side of the larynx with extension beyond the cartilage. Caudally the mass extended to the level of C6 vertebra, infiltrating the left lobe of thyroid, thyroid cartilage and the upper esophagus. The mass abuts the left carotid sheath without definite evidence of vascular invasion. The oropharynx proximal to the mass was dilated. Posteriorly,

the tumor was in contact with the vertebral bodies with obliteration of the prevertebral fat. No enlarged cervical lymph node enlargement was seen.

Examination under anesthesia revealed a large tumor occluding the entire hypopharynx and an incisional biopsy was performed. Histology revealed a spindle cell lesion with extensive necrosis. There were sheets and fascicles of uniform small spindle cells (Fig. 2). Individual tumor cells had oval nuclei, inconspicuous nucleoli, finely dispersed chromatin and scanty cytoplasm (Fig. 3). Hemangiopericytoma like vascular pattern was seen in between sheets and fascicles of tumor cells. Twelve mitoses per 10 high power fields were noted. Monophasic SS, spindle cell carcinoma and malignant peripheral nerve sheath tumor (MPNST) were considered in the differential diagnoses. Cytokeratin immunohistochemical stain was performed which was focally positive in spindle cells (Fig. 4). On the basis of histological and immunohistochemical findings the lesion was diagnosed as monophasic SS.

The lesion was deemed inoperable and three courses of palliative chemotherapy with adriamycin and cyclophosphamide were given. Postchemotherapy scans showed stable disease and palliative radiation 30 Gy in 10 fractions was added to the treatment. The disease progressed 4 months later and the patient died in March 2009.

DISCUSSION

SS are histologically classified as either biphasic, monophasic or poorly differentiated. Biphasic synovial sarcomas are composed of epithelial and spindled cells. Usually, the spindle cell component predominates. Mast cells, mitoses, areas of calcification, and scant collagen production are typical of biphasic synovial sarcoma. Secretions within the epithelial cells and pseudoglandular spaces are PAS positive and diastase resistant, alcian blue and mucicarmine positive. A rare poorly differentiated subtype has been described. These tumors may consist predominantly of epithelial cells, spindle cells, or a small cell variant that forms rosettes. The stromal mucin secreted by the spindle cells is alcian blue positive but PAS negative. Reticulin stain helps in demonstrating the biphasic pattern of the tumor. Nests of plump rounded cells are highlighted by the reticulin stain. Monophasic synovial sarcoma is composed of a single cell type and may be derived from epithelial or spindle cells. Both epithelial and spindle cells stain positively for cytokeratin and epithelial membrane antigen (EMA).² CD99 stains 62%

of synovial sarcomas with spindle cells staining in membranous pattern and epithelial cells showing cytoplasmic positivity.⁵ In addition BcL2 stains almost all SS.⁶ SS also



Fig. 1: MRI neck showing a huge soft tissue mass of neck arising from the hypopharynx

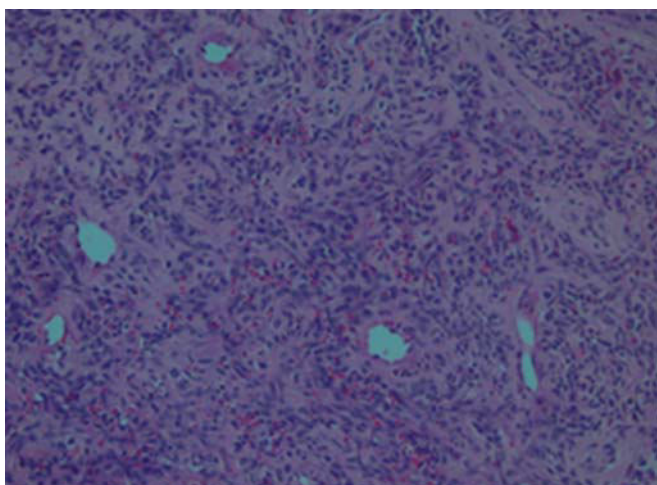


Fig. 2: 4x: Spindle cells arranged in sheets and focal hemangiopericytomatous areas

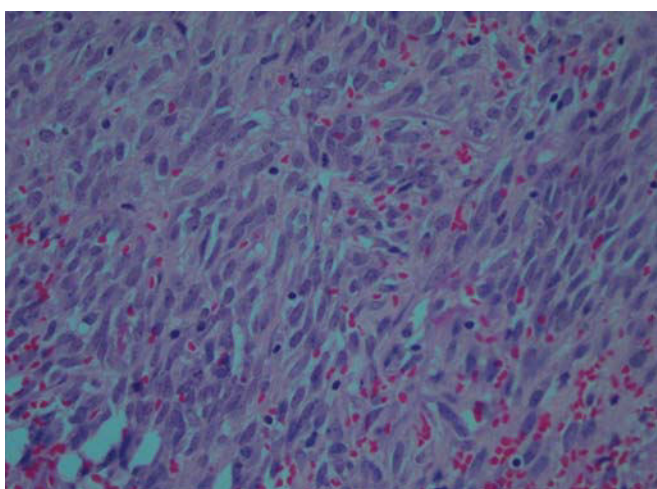


Fig. 3: 40x: Spindle cells having oval nuclei with dispersed chromatin and inconspicuous nucleoli

stains positively with cytokeratin 7 and 19, other soft tissue sarcomas including malignant peripheral nerve sheath tumor stain positively with cytokeratin 8 and 18. Cytokeratin positivity is noted in only 50% cases of poorly differentiated SS and also in monophasic SS immunoreactivity of cytokeratin is reduced by almost 60%.²

In the differential diagnosis of 'round cell tumors expressing epithelial markers', poorly differentiated synovial sarcoma should be included. Application of a panel of immunohistochemical markers is suggested to avoid diagnostic pitfalls.

A translocation $t(X;18)(p11.2;q11.2)$ is the hallmark of synovial sarcomas whatsoever the histologic type may be; $t(X;18)(p11.2;q11.2)$ seems to be specific. It is not found in other spindle cell sarcomas, and very rarely detected in tumors, such as malignant fibrous histiocytoma or fibrosarcomas. Cytogenetic studies are helpful in establishing the diagnosis in poorly differentiated synovial sarcomas.⁷

The AJCC stage is determined by the size of the tumor, the histologic grade, and whether there is spread to lymph nodes or distant sites. Synovial sarcomas can recur locally and metastasize distantly. The reported incidence of nodal metastasizes in SS is 10 to 15%; a risk for nodal metastasis that is higher than other soft tissue sarcomas of adults.⁸ Factors associated with a poor prognosis include increasing age >60 years, grade, tumor size greater than 5 cm, and mitotic activity.⁹

Synovial sarcomas are usually treated aggressively with combination of surgery and postoperative radiotherapy. Limited excision is associated with a high incidence of local recurrence (60-90%) within 2 years of the original surgery.¹⁰ The surgical excision is followed by postoperative radiotherapy and chemotherapy to help control metastasis.¹¹

SS is a rare tumor in the head and neck region. The diagnosis of SS should be considered in the differential of

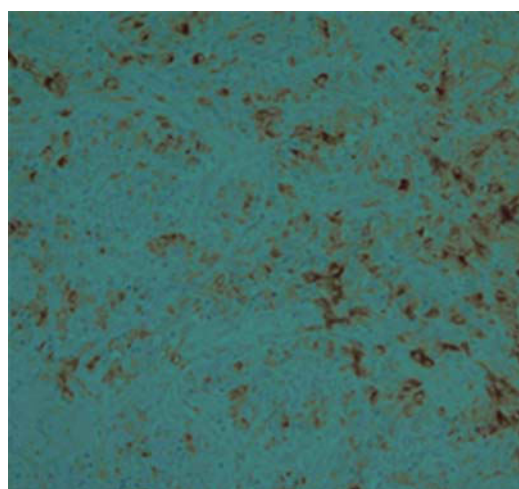


Fig. 4: Focal cytokeratin positivity in spindled tumor cells of monophasic SS

spindle cell and round cell tumors expressing epithelial markers. The use of a panel of immunohistochemical markers is suggested to avoid diagnostic pitfalls.

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