

Case Report

Monophasic Synovial Sarcoma of Pharynx: A Diagnostic Challenge

Gurshinder Pal Singh, Sandeep Dachuri, Shaila T Bhat, Ankur Kaur Shergill

Abstract

Synovial Sarcomas of the head and neck region are very rare. The histologic monophasic variant of this tumor in the pharynx is even more uncommon. The benign appearance of an otherwise aggressive neoplasm offers a diagnostic challenge to both the surgeons and pathologists worldwide. Immunohistochemistry augments the conventional histopathology and offers a definitive diagnosis. Surgery forms the main mode of treatment followed by adjuvant radiotherapy. A rare case of monophasic synovial sarcoma of pharynx of a young boy is reported here. Any benign appearing chronic lesion with sudden exacerbation in head and neck region in young age group should be envisaged with high index of suspicion.

Keywords: Diagnosis; Immunohistochemistry; Pharynx; Sarcoma; Synovial; Radiotherapy.

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Introduction

Although common in extremities, Synovial Sarcoma (SS) is rarely seen in the head and neck region. It is an aggressive soft tissue tumor most commonly seen in the younger age group associated with a poor prognosis. This tumor arises from the mesenchymal tissue but is so named due to its histological resemblance to synovium. Its classic histological subtypes include the biphasic and the rare monophasic type. The latter subtype is extremely uncommon in the head and neck region.¹⁻⁵ The diagnosis of synovial sarcoma is difficult in cases which present with a benign chronic nature. Early diagnosis and precise treatment of synovial sarcomas are utmost challenges today. Complete surgical excision and adjuvant therapy are the most accepted treatment modalities.⁵

Case report

A 13 year old boy reported to the out-patient department of tertiary care center in Manipal with a chief complaint of painless swelling on the right side of neck since one year. It was insidious in onset and had gradually progressed to the present size. He also gave a history of change in voice and occasional pain in the swelling. There was no relevant medical or family history. A thorough clinical examination revealed a 3 x 3cm cystic swelling in the right submandibular region. The overlying skin was normal. The intra oral examination revealed no abnormalities. On further endoscopic examination of the upper aerodigestive tract, a smooth sub mucosal swelling measuring 7 x 5cm in greatest dimensions extending from the

inferior pole of right tonsil to the lateral wall of right pyriform fossa involving lateral pharyngeal wall was seen. On the basis of above clinical findings, a provisional diagnosis of a benign cystic swelling was made. The swelling was subjected to Fine needle aspiration cytology (FNAC) and Computed tomography (CT) scan. FNAC findings revealed the swelling to be a benign cyst. In addition to this, CT scan exhibited a well-defined peripherally enhancing cystic lesion which measured 3.2 X 5 X 4.6 cm. It possessed enhancing septae in the right Para pharyngeal region extending and effacing the right pyriform fossa. Anteriorly, and on the right side, it displaced the right submandibular gland and right cornu of the hyoid bone. It extended medially which resulted in narrowing of the oropharynx (Fig 1a). Superiorly, it was seen abutting the right tonsil and the base of tongue. The epiglottis was pushed to one side. Due to the benign picture of the lesion, it was subjected to surgical removal.

Intraoperatively, a well-defined cystic mass with solid component was found abutting the oropharynx on right side behind the right greater cornu of the hyoid bone (Fig 1b). The right greater cornu was resected and pharynx was opened for complete excision of the mass. The excised specimen was sent for histopathological examination. The microscopic sections revealed a hypercellular neoplasm consisting of spindle cells with plump nuclei and coarse chromatin. Focal areas demonstrated haemangiopericytomatous arrangement of

vessels with minimal mitotic activity (Fig 1c). The tumor showed diffuse positivity for Bcl-2 (B-cell lymphoma 2) concordant with its high mitotic activity (Fig 1d). Focal areas were positive for EMA (epithelial membrane antigen) confirming its epithelial origin (Fig 1e). The tumor cells were negative for S-100 and α -SMA (alpha smooth muscle actin). The tumor was negative for cytokeratins (CK) and showed a weak expression for

MIC2 (CD99). The above features pointed to a clear diagnosis of Monophasic SS. The patient refused the adjuvant radiotherapy treatment and returned after four months with local recurrence. He was treated with wide local excision of the recurrent tumor followed by chemoradiotherapy. The patient has remained disease free since two years from then.

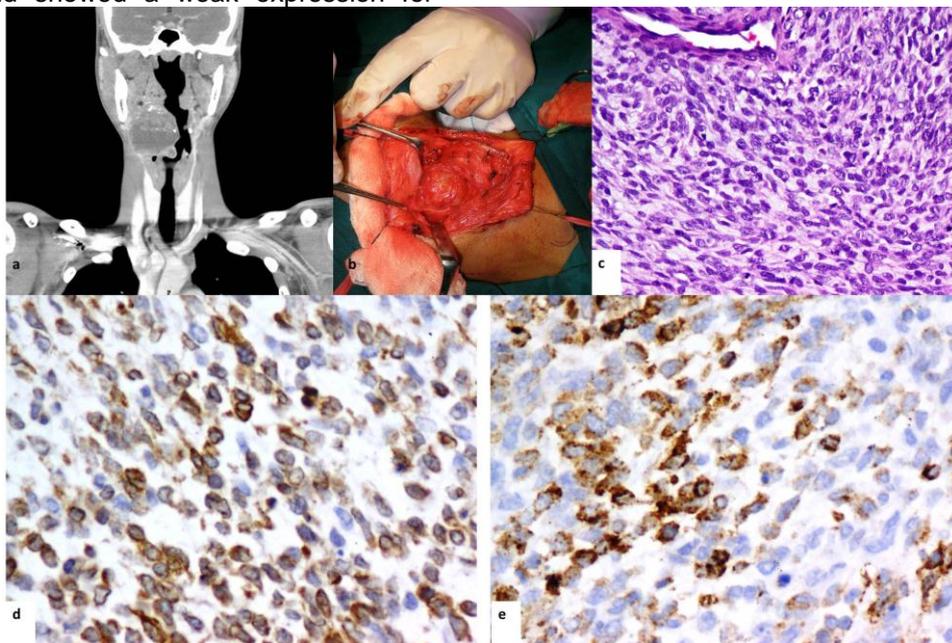


Figure 1: The computed tomography showing well circumscribed cystic mass in the right side of the neck (a). Intraoperative tumor mass seen in the submandibular region (b). The photomicrograph depicting fascicles of tumor cells with hemangiopericytomatous appearance of blood vessel (Hematoxyllin and Eosin x 10)(c), Tumor cells showing diffuse positivity for Bcl-2 (d) and focal EMA positivity (e).

Discussion

Synovial Sarcoma is a rare form of sarcoma in the head and neck region. In the past, monophasic synovial sarcomas have been reported in sites like lungs, nerves, gastrointestinal tract, liver, vulva and conjunctiva. Very few cases of Monophasic SS of pharynx have been reported in the world literature.²⁻⁴ It exists in children and young adults with preponderance in males.² This neoplasm usually follows a chronic protracted course with sudden exacerbation of symptoms occasionally.⁴ The definitive diagnosis is attained only after a complete histopathological examination together with immunohistochemical evaluation of the excised tumor.⁶ Synovial sarcoma usually presents as a slow growing well circumscribed solid mass. Although literature indicates the presence of some radiological findings including hemorrhage, calcification, heterogeneous intensity on T2 weighted images and heterogeneous enhancement

pattern in this tumor, SS is frequently misdiagnosed as a benign entity owing to its smooth margins, cystic component and lack of aggressive infiltration.⁶⁻⁸ In our case, a young boy presented with a cystic mass which was clinically and radiologically considered to be a benign cyst because of its chronicity, cystic nature and lack of aggressive infiltration.

Microscopically, SS consists of two predominant cell patterns which include the epithelial type and sarcoma like spindle cells. While biphasic type of the tumor consists of the distinct epithelial cells and spindle cells in different proportions, the monophasic type presents with a uniform pattern of a single type of cells. The poorly differentiated SS consists of predominantly epitheloid or round cell morphology and an increased mitotic activity. Our case presented with increased mitotic activity. Immunohistochemical evaluation of tumor

forms an indispensable tool for the definitive diagnosis of SS. Majority of SS exhibit immune positivity for EMA and cytokeratins⁹ The tumor cells in our case were EMA positive but CK negative which is a rare finding. Low expression of cytokeratins is a common finding in poorly differentiated tumors.¹⁰ This points towards a more aggressive nature of the tumor. Bcl-2 is diffusely expressed in almost all cases of SS especially of spindle cell type. Our case was no exception. MIC2 gene is expressed in 60-70% of SS.⁹ A weak expression of MIC2 was observed in our case.

The most accepted mode of treatment is surgical excision of the tumor mass with wide margins followed by postoperative radiation therapy especially in high risk cases. Chemotherapy has emerged as a promising treatment in the current times. Current literature points towards a multidisciplinary approach for treatment of the disease.⁵ In our case the patient denied the adjuvant treatment and came back with a local recurrence after 4 months.

Monophasic SS of the pharynx are very rare. Because of rarity of this disease and a benign presentation, diagnosis of this entity presents as a challenge to surgeons, radiologists and pathologists. A high index of suspicion is required for this type of presentation especially in children and adolescents. An increased awareness of this disease with more representation of cases can assist in a definitive diagnosis and prompt treatment.

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