Case Report

Primary biphasic synovial sarcoma of left parapharyngeal region: an exceedingly rare site.

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Abstract

Primary synovial sarcoma is an unusual tumor of the head and neck region and again primary parapharyngeal synovial sarcoma is a rare tumor entity. In our case a male patient, aged 27 years, presented with feeling of swelling and discomfort in the left parapharyngeal region. CT findings suggestive of soft tissue mass in prestylar region on left side. A transcervical excision of the tumor was performed and histopathological examination of H & E stained sections show biphasic tumor composed of epithelial and spindle cells components. The epithelial areas show glandular structure lined by cuboidal cells while spindle cells areas show densely cellular fascicles surroundings the epithelial cells. Immunohistochemistry shows strong positivity of cytokeratin, vimentin and weak positivity of S-100, BCl2, CD99 and SMA. Synovial sarcoma may be mistaken for other tumors due to its rarity. Treatment is essentially surgical, requiring wide margins; radiotherapy is usually associated. The value of chemotherapy has yet to be assessed.

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Introduction

Synovial sarcoma is a distinct soft tissue sarcoma that occurs in young and middle-aged adults however, it may occur in children and the elderly in a variety of locations.[1] It has a predilection for the extremities but may occur rarely in head and neck region and parapharyngeal space.[2] It accounts for about 6.8% of head and neck tumors.[3] Its relationship to anatomic synovial structures often is tenuous or nonexistent. We describe a case of biphasic synovial sarcoma of parapharyngeal region confirmed by various immunohistochemistry stains. The clinical and morphologic spectrum of synovial sarcomas is quite wide. [1] This case demonstrates a rare occurrence of a synovial sarcoma in the parapharyngeal region.

Case Report

A 27-year-old male presented with feeling of swelling in oral cavity at left tonsillar region. Since six months he noticed bulge at left tonsillar region which was initially smaller in size and gradually increased in size reaching up to uvula and left retromolar triangle (RMT) associated with restricted mouth opening and upper aero-digestive tract obstruction. On local examination there was painless firm to hard mass lesion arising from the left-sided parapharyngeal space with no cervical lymphadenopathy.

Initial computerized tomography scan (CT scan) revealed the presence of well defined predominantly hypo dense lesion in the prestyloid space in the neck on left side and conclusive findings suggestive of soft tissue mass in prestyloid region on left side. Possibility of neurogenic tumor or minor salivary gland tumor was considered pre-operatively. A transcervical excision of the tumor was performed and sent for histopathological examination at our department.

Gross findings: The single gross lesion was a roughly ovoid, somewhat lobular mass growing in an expansile manner that rendered it circumscribed and measuring 6 × 5 × 4 cm in size. Outer surface was smooth, capsulated with grayish white and brownish in color (Fig. 1A). On cut section homogeneous mass was mainly solid and firm in consistency with grayish white in color (fig. 1B). Hemorrhage and necrosis were not seen.

Microscopic findings: H & E stained sections show biphasic tumor composed of segregated epithelial and spindle cells components. The epithelial areas appear in the form of gland-like spaces lined by epithelium filled with secretion (fig. 2) and presence of few solid nests of epithelial cells with foci of squamoid differentiation. Spindle cell areas show hypercellularity and fibroblast-like appearance with monotonous appearance and plump and vesicular nuclei (fig. 3). Spindle cells are mainly arranged in long fascicles with intersecting bundles and whorled pattern and also presence of hemangiopericytoma-like areas. Stroma shows hyalinization and myxoid areas.

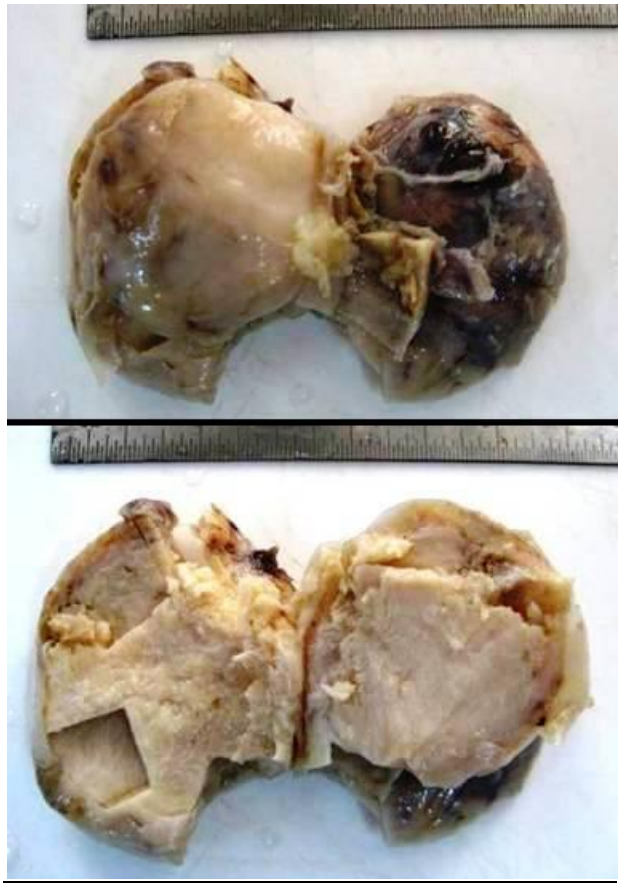

Figure 1(A&B): gross appearance and cut section of tumor.

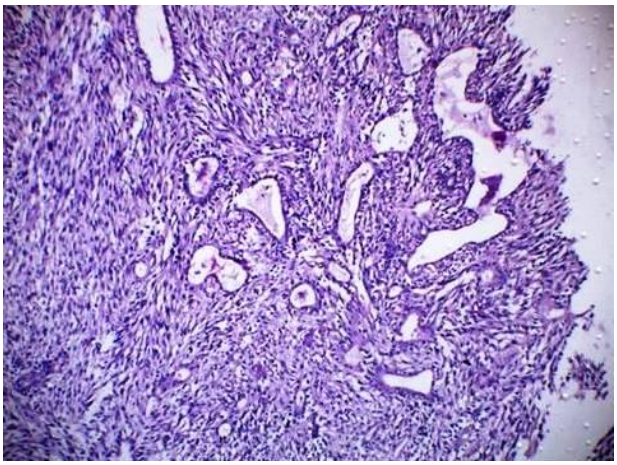

Figure 2: Section showing glandular epithelial component (H&E, 10x)

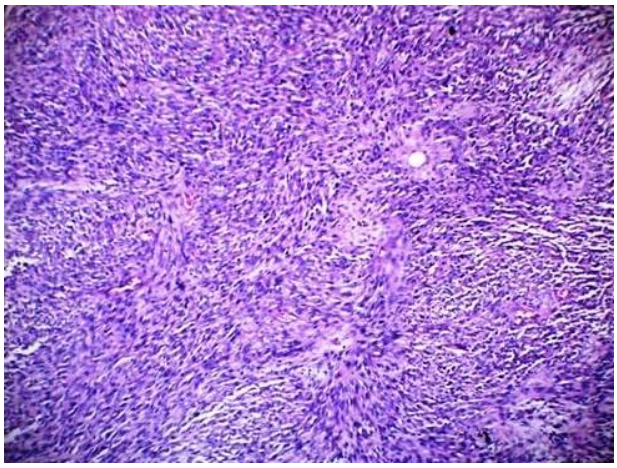

Figure 3: Section showing spindle cell area with fasciculation (H&E, 10X)

As in this case, the cells stained strong positive reactivity for cytokeratin supported the diagnosis since normal or reactive synovial cells do not express keratin.[4] Vimentin, Epithelial membrane antigen (EMA) also show positive reactivity and smooth muscle actin (SMA), S-100 & bcl-2, also show weak positive staining and CD99 stains focally[5] (fig. 4). Weak expression of desmin, CD34, and HMB – 45 are also found in our case.

vessels, abdominal wall, prostate, nerves, pleural cavity, kidney, and heart.[7] The diagnosis of synovial sarcoma can be very challenging to the pathologists especially at uncommon sites. Salivary gland and neurogenic tumors are the most common neoplasms involving the parapharyngeal space [3]. Synovial sarcoma accounts only 3–10% of all head and neck soft tissue sarcomas. Guadagnolo reported a large series of 150 cases of synovial sarcoma and only 9 involving the head and neck region. In the head and neck region, synovial sarcoma originates from primitive mesenchyma with no synovial association.[8] Patients usually present a painless slow growing mass during their third and fourth decade of life with associated compressive or infiltrative symptom of surrounding structures [8] .Some of the predisposing factors to development of sarcoma include genetic factors, viral infection in immunocompromised patients, irradiation, chronic lymphoedema, environmental carcinogens and trauma. [8] They often present as a slow growing well-circumscribed mass that mimics benign pathology. Histopathologically, synovial sarcoma is characterized by presence of epithelial and spindle cells with various different histopathologic subtypes.

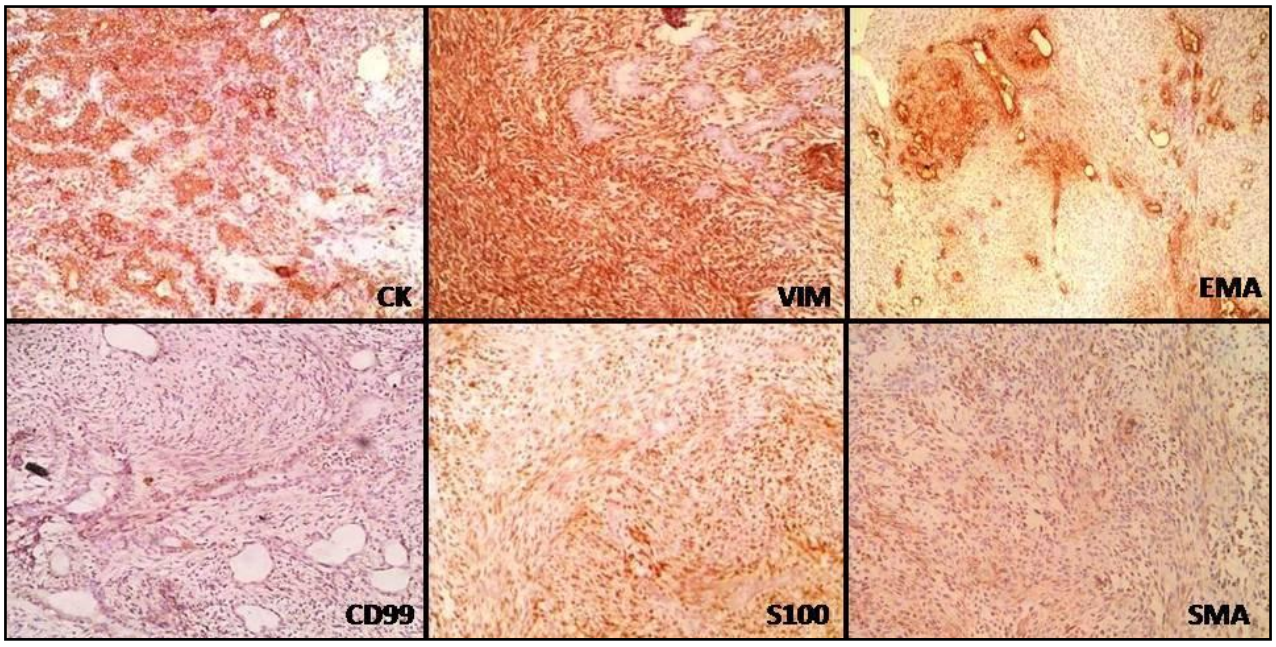

Figure 4: Section showing strong positivity for CK, Vimentin, EMA and weak/focal positivity for CD99, S-100 and SMA.

Discussion

Synovial sarcoma typically arises about the knee and ankle joints of children and young adults.[6] It rarely occurs in other locations of body such as head & neck, tonsils, mediastinum, skin, blood

The most recognized subtypes are monophasic and biphasic variants. The classic monophasic subtype has a single cellular component composed solely of spindle cells and may be misdiagnosed as fibrosarcoma or malignant peripheral nerve sheath tumors.

Whereas the biphasic subtype is composed of a mixture of elongated basophilic spindle cells and glandular structures made of columnar epithelial cells[9] similar to our case. The differential diagnosis between synovial sarcoma with other benign or malignant sarcomas such as rhabdomyosarcoma, fibrosarcoma, malignant hemangiopericytoma or lymphoma is difficult to make clinically or radiologically. Tissue biopsy is still necessary for confirmative diagnosis of synovial sarcoma. The diagnosis of synovial sarcoma requires an integrated multidisciplinary approach. Immunochemistry helps to identify these tumors as they show positive reactions for keratin and epithelial antigen [4,5,10] as seen in our case. Vimentin, EMA, and occasionally S-100 protein are also expressed by this tumor[4,5] and was also expressed in our case. It has also been found that a significant number of cases of synovial sarcoma are immunoreactive for CD99, the marker characteristically associated with Ewing sarcoma/PNET and BCL2.[5] The treatment of choice is surgery with or without radiotherapy and chemotherapy. The survival rate at five years varies from 25% to 62% and at ten years range from 10% to 30%.

Conclusion

The possibility of occurrence of this rare tumor at this unusual site should be kept in mind and Immunohistochemistry should always be advised to rule out and to know the nature of the tumor at this location.

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Competing Interests

None declared.

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