Parapharyngeal Space Synovial Sarcoma

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SUMMARY

Synovial sarcoma is a rare soft tissue sarcoma that commonly involves the lower extremities and occurs predominantly in young adults. It very rarely occurs in the head and neck region and carries a poor prognosis. The tumor demonstrates both a loco-regional and a systemic pattern of spread that makes management challenging. Combined modality therapy of this aggressive tumor yields better results. Herein, we described a 58 years old lady presented with left parapharyngeal synovial sarcoma. This is to demonstrate the aggressiveness of the disease despite complete surgical excision followed by post operative radiotherapy. Local regional control was difficult and recurrence of disease was seen in this particular patient.

KEY WORDS:
Synovial sarcoma, Head and neck, Parapharyngeal space

INTRODUCTION

Synovial sarcoma is a mesenchymal malignancy which primarily occurs in the periarticular region and predominantly located in the extremities [1]. It is a rare for synovial sarcoma to occur in the head and neck region which is poorly supplied with synovial tissue [1-2], with only less than 100 cases have been reported in the literature [3]. To explain the occurrence in this unusual site it been suggested that synovial sarcoma arise from undifferentiated or pluripotential mesenchymal cells [2]. Synovial sarcomas can occur high in the superior aspect of the neck, in the prevertebral areas from the base of the skull to the hypopharynx, the retropharyngeal and parapharyngeal areas [2].

CASE REPORT

A 58 years old, Chinese lady presented to our ENT department with a complaint of difficulty in swallowing for 2 months duration, associated with change in voice for the past one month. There were no symptoms of fever, odynophagia or difficulty in breathing. Examination of the oral cavity and oropharynx revealed a huge smooth surface tumour occupying the left oropharyngeal space not crossing the midline, but extending anteriorly into the oral cavity, superiorly to the level of soft palate and inferiorly up to the supraglottic region. Contrasted computer tomography(CT) scan of the Head and neck showed a 5x4x2cm smooth surface mass arising from the left parapharyngeal space with solid and cystic components. There were no features suggesting malignancy or involvement of the prevertebral space. As the tumour was benign in characteristic and accessible transorally, a wide intraoral surgical excision was performed; revealing a fleshy tumour with cysts filled with serous fluid. There was minimal spillage of the cystic fluid in the oropharynx intraoperatively. Histopathological report showed a biphasic type of synovial sarcoma. A repeat CT scan post excision showed no residual tumour and the she was closely followed up. Seven months post excision; she developed recurrence at the same site with no distant metastasis. We decided to proceed with transoral wide excision again as the tumour was still accessible via this approach followed by post operative radiotherapy. She was disease free for one year, but unfortunately she developed another recurrence over the same site with no distant metastasis and the tumour was fast growing. A lateral pharyngotomy which provided a better access and exposure for the excision of the tumour was performed as the preoperative CT scan showed a tumour abutting the internal carotid artery. Intraoperatively, the tumour was noted to be encasing the internal carotid artery and adherent to the prevetebral space as well as invading the intervertebral space, thus the surgical margin was compromised. Options of chemotherapy or repeat surgery were given but she declined any further treatment and succumbed to the aggressive growing tumour 6 months post surgery.

DISCUSSION

Synovial cell sarcomas usually presents between third and fifth decade of life but it may also occur at any age [1]. Although the synovial sarcoma of the extremities and head and neck are histologically similar, the survival rate of head and neck synovial sarcoma is worse [1]. Synovial sarcomas of the head and neck usually present as a progressively painless enlarging mass in the neck that causes dysphagia and dyspnoea. Tumors can be spherical, well-circumscribed, lobulated or multinodular, covered with a pseudocapsule with cystic and haemorrhagic foci [2-3]. In the initial radiological imaging for this case it was difficult to distinguish between a benign and malignant lesion as synovial sarcoma mimics a benign mass because of its homogenous appearance and smooth margin with lack of aggressive infiltration [3]. Schwannomas, neurofibromas, and tumors of ectopic minor salivary glands are some of the benign tumours which can be hard to distinguish from synovial sarcoma.

Histopathologically synovial sarcoma are described in two subgroups; namely the monophasic and the biphasic cellular patterns [2]. The biphasic pattern is composed of two cell populations, epithelial cells and spindle cells [4]. The monophasic pattern often poses a diagnostic challenge

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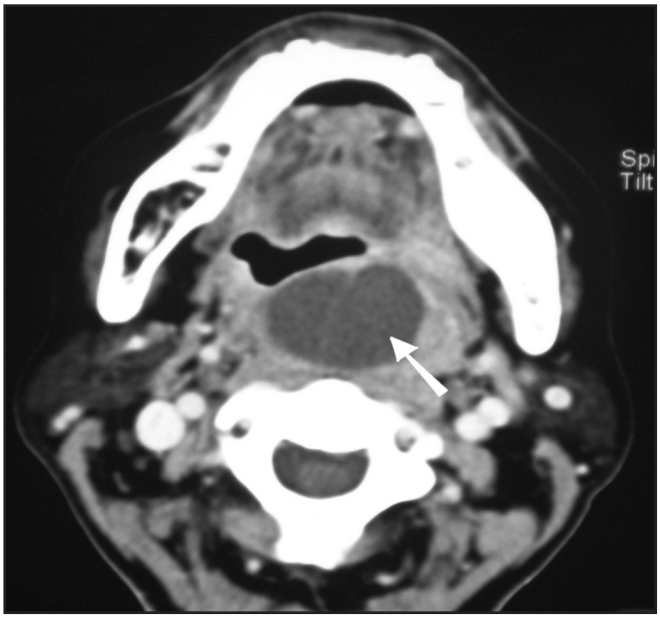

Fig. 1: CT scan of the Neck (contrasted) showing large lobulated mass with solid and cystic component measuring 5x4x2 cm arising from the left parapharyngeal space.

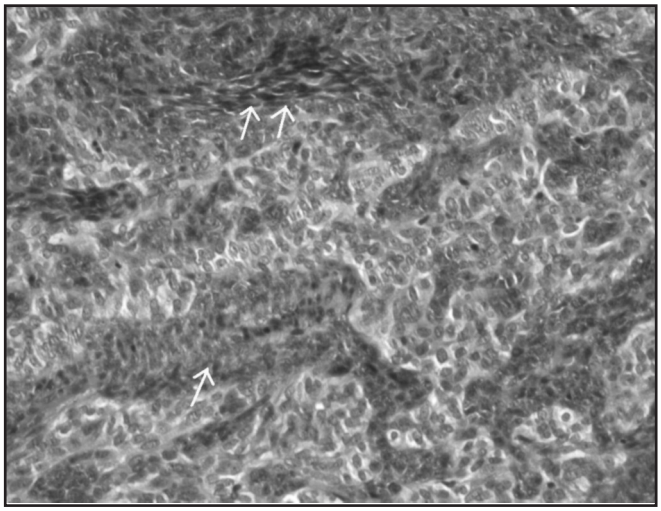

Fig. 2: Microscopic examination of the specimen demonstrating Biphasic tumour consisting of both epithelial (single arrow) and spindle cell (double arrow). ((hematoxylin-eosin stain, original magnification X 10).

because can be it is easily confused with other mesenchymal or epithelioid neoplasms [1]. Special immunohistochemical stains and cytogenetic evaluation can help in confirming the diagnosis [4].

Although characterized by its slow growth, synovial sarcoma results in poor long-term survival because of its tendency for local recurrence and distant metastasis, especially to the lungs[1]. Local recurrence and lung metastasis lowers survival to 40% at 5 years and 25% at 10 years [3].

The rarity of this tumour means that there is no ideal treatment regimen. Even with the probability of high local recurrence, of 40–80% in adults and reaching 90% in childhood, surgery remains the treatment of choice [3, 5], with or without adjuvant radiotherapy [5].

Patients treated with surgery and adjuvant radiotherapy have higher survival rates and lower recurrence rates than those treated with surgery alone or surgery with chemoradiotherapy [1]. Although, adjuvant treatment decreases the local recurrence, there no improvement in long term survival, the 5 years survival rate ranging only 25-55% [4] . Our patient could have benefitted from an early combined modality treatment despite adequate wide excision and clear surgical margin.

The important prognostic factors in synovial sarcoma are the size of the initial tumor and its extent at the time of primary treatment. Tumour size > 5 cm was associated with poor outcome [1-2]. Early diagnosis, younger age (<20 years) and wide surgical excision were found to be favourable prognostic factors for better survival [3-4].

CONCLUSION

Synovial sarcoma of the head and neck is a rare entity; therefore there is limited information on the treatment of this malignant disease. Given the loco-regional recurrence as seen in our case, combined modalities of treatment should be considered at the onset of treatment despite having good surgical margins.

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