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RESEARCH ARTICLE

SYNOVIAL SARCOMA IN THE NAPE OF NECK: A RARE CASE WITH LITERATURE REVIEW

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ABSTRACT

Background: Soft tissue sarcoma are rare among which synovial sarcoma account for 5-10% of all soft tissue sarcoma**. Case report**: This case corresponds to a 55 year old male patient presented to surgery OPD with swelling over nape of neck for 1 year. Contrast CT showed well defined heterogeneously enhancing lobulated mass lesion of size 38x51x60mm. FNAC revealed malignant spindle cell lesion. Histopathology of excised specimen showed malignant round/spindle cell neoplasm. Immunohistochemistry was done and was suggestive of synovial sarcoma of nape of neck. **Discussion:** Numerous spindle cell lesion can present along nape of neck. Surgery is considered mainstay of management of synovial sarcoma. More advanced diseases requires combined treatment with adjuvant radiotherapy.In this casepatient is now undertaking adjuvant external beam radiotherapy.

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INTRODUCTION

Soft tissue sarcomas (STS) are rare, malignant mesenchymal tumors that includeover 70 different and highly diverse histological subtypes (Sultan, 2005). This heterogenous group of lesions includes those of neural, fibroblastic, myofibroblastic, myogenic, epithelial and vascular tumours (Chan, 1997). The annual incidence of soft tissue sarcoma is estimated to be between1.4 to 5 cases per 100000 (DeVita, 2015). Most soft tissue sarcoma are believed to be sporadic and have no clearly defined cause. In small proportion ofcases some predisposing factors have been identified which include genetic factor, lymphedema, prior radiation therapy and carcinogens. Soft tissue sarcomas become more common with increased age and median age at diagnosisis 65 years. Soft tissue sarcoma can occur in any site throughout body. A total 45%are located in extremities with 30% occurring in lower limb (most commonly in the thigh 38% are intra-abdominal, 10% truncal and 5% are head and neck) (DeVita, 2015). Synovial sarcoma (SS) accounts for 5–10% of all Soft tissue sarcomas (Corey, 2014). Synovial sarcoma is an intriguing disease and unlike the majority of soft tissue sarcomas, it can occur at any age and everywhere in the body.

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The peak incidence is in the 30s and it often presents in the extremities (Vlenterie, 2018). Synovial sarcoma was initially described as a biphasic neoplasm comprising of both epithelial and uniform spindle cell components. Spindle cells are of mesenchymal origin and constitute a part of the body's connective tissue. On cytological examination these cells appear elongated with a fusiform or ovoid nucleus (Wikivet, 2018; Åkerman, 2003). The tissue oforigin can be determined based on evidence of collagen, cartilage, bone, fat ormyxomatous material formed by the tumour cells (Wikivet, 2018). Patients usually present with a deep-seated mass that has been present for several years. Haematogenousspread is typical for sarcoma. In most cases, metastases are localized in the lung (80%), although metastases can arise in lymph nodes (up to 20%), bone(9.9%), and liver (4.5%).[8] Most soft tissue tumors are benign and are usually cured by simple surgicalexcison. The cornerstone of treatment for synovial sarcomas, as with other soft tissue sarcomas, is complete surgical resection (Eilber, 2008). This fact is explainable by the location and natural history of the disease synovial and as synovial sarcoma are very rare so weare discussingthe outcomes of this case here.

CASE REPORT

A 55 years old male patient presented to surgery OPD with swelling over nape ofneck for 1 year measuring around 8x5 cm (Fig 1). Patient had no history of any similarcomplaint in family or any history of trauma.

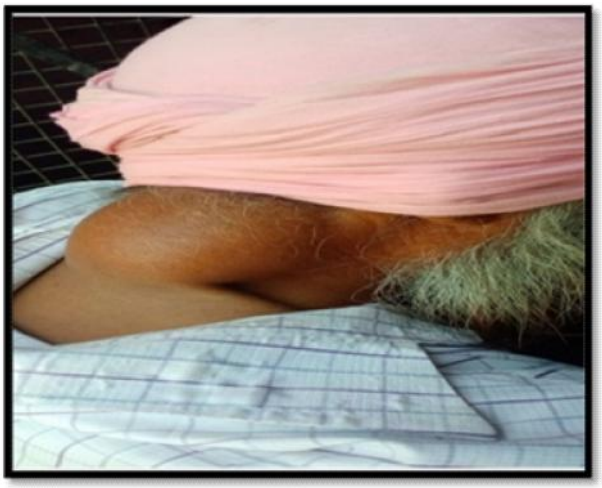

Figure: 1

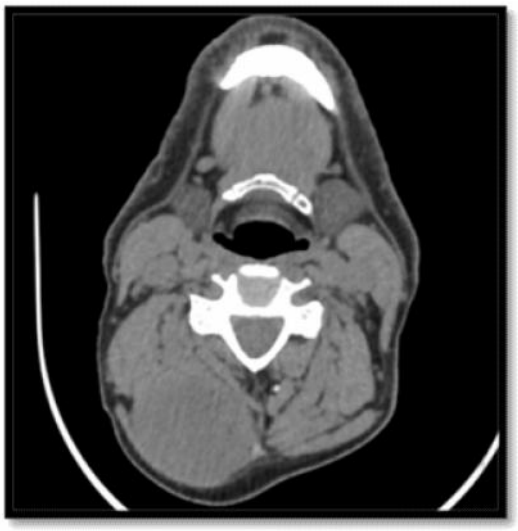

Fig. 2

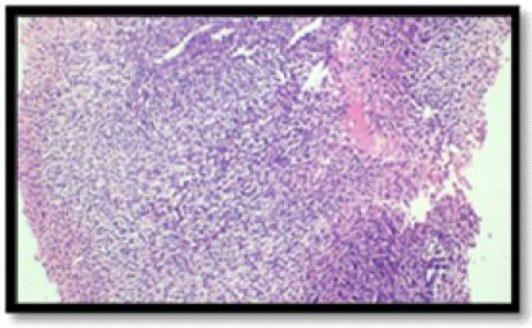

Fig. 3 A

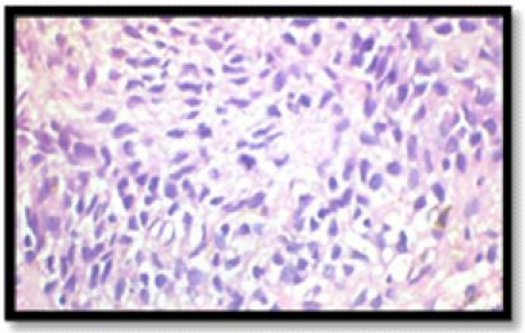

Fig. 3 B

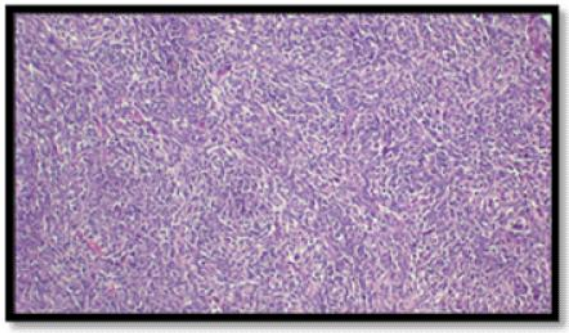

Fig. 4A

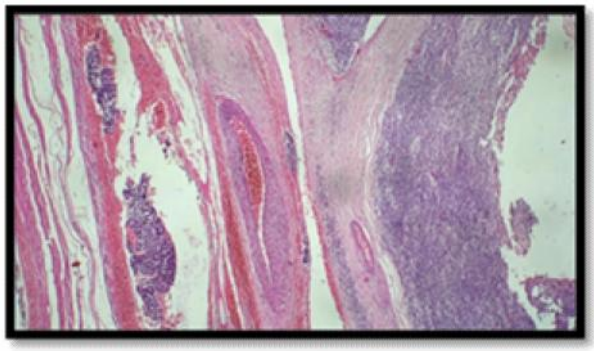

Fig. 4B Figures title missing

On examination swelling was firm, oval, non-tender, with restricted mobility and irregular borders. Contrast CT(Fig-2)of neck showed well defined heterogeneously enhancing lobulated mass lesion 38x51x60mm, involving right trapezius, splenius, capitis, semispinalis capitis and obliqus capitis inferior muscle extending from C2-C3 IV disc level up to C6 vertebral level. Areas of necrosis were seen within mass lesion. Few small sub-centimeter sized satellite nodule are seen adjacent to lesion. Overlying skin and subcutaneous tissue appeared normal. FNAC was done (Fig.3A & 3B) from nape of neck swelling smears were prepared and processed routinely stained with Leishman Giemsa stain. Smears of marked cellularity shows oval to spindle shaped cells in small groups and singly dispersed having round to oval shaped nucleus, fine chromatin, scanty to moderate cytoplasm. Findings were suggestive of malignant spindle cell lesion.

Patient was planned for surgery under prone position in general an aesthesia and excisional biopsy was done and sent for examination. Histopathological examination showed fibromuscular connective tissue and tumor tissue composed of diffusely infiltrative as well as sheets round to spindle shaped cells having N:C ratio, hyperchromatic nucleus, moderately nuclear polymorphism, inconspicuous nucleoli, fine chromatin, scanty to moderate vacuolated cytoplasm, karyorrhectic debris, mitosis and necrosis (Fig. 4A & Fig 4B). The inked capsule surface was uninvolved by tumor. At one place tumor breached the capsule but did not invade the skeletal muscle. This was suggestive. Of malignant round cell/spindle cell neoplasm (pTNM–pT3Nx) Immunohistochemistry was done in whichvimentin and BCL2 were diffuse strongly positive, Pan CK and CK 7 were intermediate to strong focal positive, synaptophysin and calretinin were weak to intermediate focal positive while CD45, Tdt, desmin, CD34 and S-100 were negative.

This was suggestive of synovial sarcoma-nape of neck withFNLCC grade3and lymphovascularinvasion. Patient is now undertaking adjuvant external beam radiotherapy.

DISCUSSION

There are numerous atypical spindle cell lesions which can present along the nape of neck. Because it is so rare synovial sarcoma should be strongly investigated with attention to the clinical scenario, careful evaluation the H&E morphologic features, and judicious use of immunostains, one can work through these difficult cases to arrive at the correct diagnosis. The cornerstone of treatment for synovial sarcomas is complete surgical resection. This fact is explainable by the location and natural history of the disease. In this situation, awide local excision is a radical treatment. The outcome of surgery alone for early synovial sarcoma is excellent. Synovial sarcoma (SS) is a rare, yet highly malignant, type of soft tissue sarcoma (STS), for which survival has not improved significantly during the past years. Compared to other Soft tissue sarcoma, synovial sarcoma is relatively chemo sensitive (Desar, 2018). Ifosfamide and ifosfamidecombinations are active in different lines of treatment. In high-risk extremity and chest wall STS, neoadjuvant doxorubicin and ifosfamide has shown as much activity as high-doseifosfamide. There are indications that combination chemotherapy with doxorubicin and ifosfamide in this setting improves outcome (López-Pousa *et al.*, 2016). More advanced disease requires combined treatment with adjuvant radiotherapy. The role of radiotherapy as primary treatment did not alter the prognosis. In this cases the outcome was not favourable.

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