Full Length Research Article

A CASE OF SOFT TISSUE SARCOMA OF RIGHT FOREARM MANAGED BY GROIN FLAP

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ARTICLE INFO	ABSTRACT
	Soft tissue sarcomas constitute 1% of all adult malignancies. Of them 35% occurs in lower limbs and 15% occurs in trunks and 10% occurs in upper limbs .Here we present a middle patient with swelling over right forehand since 2 yrs, operated elsewhere 6 months back with recurrence. Wide local excision with a groin flap was done .Patient has been followed up regularly with adjuvant chemotherapy. A proper wide excision with reconstruction followed by physiotherapy can have better results with no functional deformity.

INTRODUCTION

Soft tissue sarcomas of the upper extremities are rare and hand surgeons typically encounter only one or two undiagnosed soft tissue sarcomas during their careers. It is incumbent on the surgeon to review repeatedly the characteristics of these tumors and remain suspicious, because these lesions typically are misdiagnosed and treatment is delayed. The most common soft tissue sarcomas of the upper extremity are the epithelioid sarcoma, synovial cell sarcoma, and malignant fibrous histiocytoma. Limb salvage surgery is the treatment of choice for soft tissue sarcomas to preserve upper extremity function.

Case report

Mr. Thimmarayappa, a 55years old male patient from kanakanagar presented with swelling over the right forearm since 2years. Patient was apparently normal 2years back when he noticed a swelling over the right forearm and he consulted a local doctor 6 months back and got it excised .The swelling recurred in the same region after 6 month insidious in onset gradually progressive, initially was of size of marble to the present size of orange. No h/o fever/loss weight/loss of appetite/skin changes/any other swelling. Past history- No h/o DM, HTN, TB in past. Family history -No h/o similar complaints in the family.

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General physical examination: A middle aged patient moderately built and nourished, conscious cooperative well oriented with time, place and person.

Pulse- 90bpm
BP- 120/80mm of hg, pallor present

Local Examination

A 5x3cm oval - swelling over the right forearm extensors aspect over the styloid process of ulnar with well- defined margin with hard in consistency and fixed, skin over swelling has scar of previous surgery. No visible pulsation /scars/ sinus / distended veins.

Wrist and elbow joint- normal.
No axillary lymph nodes palpable.

Systemic Examination

Respiratory system: Normal vesicular breath sound heard B/L, no added sounds.

Cardiovascular system: S1 andS2, no murmurs.
Per Abdomen: soft, non-tender, no organomegaly.
Central Nervous system: no focal neurological deficite.

A working diagnosis of soft tissue sarcoma /osteosarcoma was made and investigations were done.

Investigations: Fine Needle Aspiration Cytology-shows features suggestive of DERMATOFIBROSARCOMA PROTRUBERANS.

X RAY wrist AP and lateral- was normal and lesion was not involving the bone.

CT scan –mass extending into inter/intramuscular aspect of the extensor compartment of the forehand and not extending to the bone or involving the vessels, possibly-malignancy

Hb % - 9.5gm/dl   Serum urea – 22 mg/dl
WBC- 7700cells/cu.mm Serum creatinine – 0.6 mg/dl
Neutrophils – 70%   Serum uric acid – 3.1 mg/dl
Lymphocytes – 24%   Serum Electrolytes – Na+-140 meq /l
Eosinophils – 06%   K+ - 4.0 meq /l
Platelet count – 2.8 lakhs.
Total Bilirubin – 1.0 mg/dl   SGOT – 22IU/L Total
Protein – 6.7gm/dl   Direct Bilirubin – 0.5 mg/dl
SGPT – 14IU/L   Albumin - 3.2gm/dl
Indirect Bilirubin – 0.5 mg/dl   ALP – 27IU/L
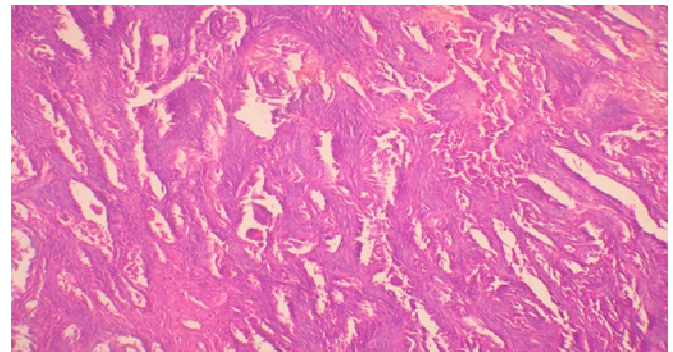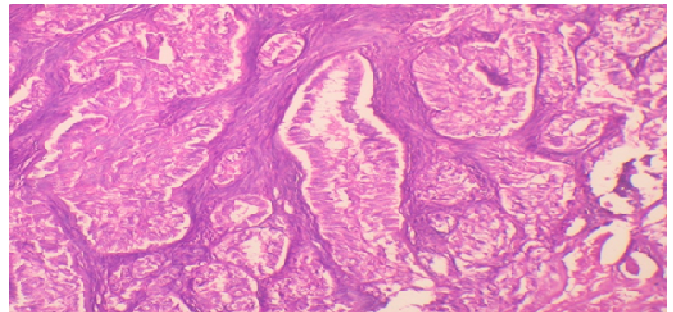
Metastatic work up (ultrasound abdomen and chest xray PA view) was normal.

Treatment-operation was done in 2 stages and 2nd stage was done 6 weeks after the 1st stage.

1st stage-Under GA and Brachial plexus block wide local excision of the tumour with 1 cm margin and pedicle groin flap was applied over the defect. The excised specimen was sent for HPE.

2nd stage-under GA,groin flap release and SSG over the contracted abdominal wound was done

Histopathology

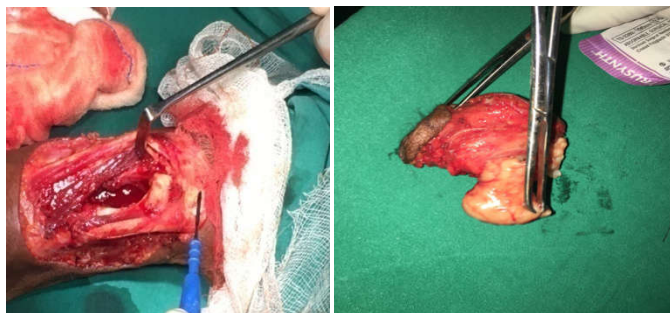Microscopy – skin with epidermis and dermis. Epidermis is un remarkable. Dermis shows a well circumscribed lesion composed of 2 components – epithelial and stromal components. Epithelial cells are large, round to oval, vesicular nuclei with abundant pale staining cytoplasm with distinct cellular border. Some areas show glandular structures containing granular – homogenous eosinophilic secretions. Margins for the tumour cells are negative. Impression – Excision biopsy dorsum of forearm shows histologic features consistent with a synovial sarcoma – monophasic type with epithelial components predominance.

Patient was given 3 cycles of adjuvant VAC (vincristine, Adriamycin and cyclophosphamide) chemotherapy and patient underwent physiotherapy for elbow and wrist joint movements.

DISCUSSION

Synovial sarcomas are malignant, high-grade, soft-tissue neoplasms that are estimated to represent between 5% and 10% of all soft-tissue sarcomas (Siegel, 2007 and Weitz, 2003). The estimated incidence of this tumor in the general population is 2.75 per 100,000 (Siegel, 2007). In fact, in adults, synovial sarcoma is the fourth most common type of sarcoma after malignant fibrous histiocytoma, liposarcoma and rhabdomyosarcoma (Siegel, 2007). In children, synovial sarcoma incidence is second only to rhabdomyosarcoma in terms of soft-tissue sarcoma (Sultan, 2009), Approximately one-third of synovial sarcomas occur in the first two decades of life (Siegel, 2007; Weitz, 2003 and Sultan, 2009). This tumor is most prevalent in adolescents and adults between 15 and 40 years of age (Siegel, 2007; Weitz, 2003 and Sultan, 2009) Concerning gender incidence, the male:female ratio is 1.2:1, with males being more frequently affected. Synovial sarcoma has a similar incidence in all ethnic groups (Athanasian, 2011; Siegel, 2007; Weitz, 2003 and Sultan, 2009). This malignancy is usually located close to the large joints of the extremities, especially the lower extremities and in particular around the knee and ankle (Athanasian, 2011; Siegel, 2007; Weitz, 2003 and Sultan, 2009). Other joints that are commonly affected are the shoulder and the hip. Most often it arises in the para-articular regions, usually in close association with tendon sheaths, bursae and joint capsules (Athanasian, 2011; Siegel, 2007; Weitz, 2003 and Sultan, 2009). However, it seldom involves the joints themselves (Siegel, 2007). In addition, contrary to what its name might suggest, synovial sarcoma also occurs in areas with no apparent relation to synovial structures, such as the heart, pericardium, pleura, lung, mediastinum, larynx, peritoneal cavity and abdominal wall (Siegel, 2007). In the hand, this tumor is more frequently found in the carpal region than in the fingers (Athanasian, 2011; Siegel, 2007; Weitz, 2003 and Sultan, 2009).

According to most authors, delay in diagnosis is very frequent (Siegel, 2007). In the majority of cases, the presence of a clinically detectable tumor prior to surgery is estimated to range from 2 to 4 years, but an insidiously growing mass or pain at the tumor site has been noted for as long as 20 years prior to initiation of proper treatment (Siegel, 2007 and Andrassy, 2001). Recently, it has been shown that the occurrence of long-standing pain at the tumor site preceding the development of a bulge is significantly more common with synovial sarcomas than with other sarcomas (De Silva, 2003). The imaging appearance is nonspecific, and in all cases a biopsy is necessary to confirm the diagnosis (Siegel, 2007; Wong, 2001 and Nakajima, 1997). Histologically, synovial sarcoma is typically characterized by epithelium-like and/or spindle cell components arranged in a biphasic or monophasic pattern, although a poorly differentiated variant has also been described recently (Ferrari, 2004). The biphasic pattern is considered the "classic" type and is generally recognizable by the coexistence of morphologically different but genetically similar epithelial cells and fibroblast-like spindle cells (Siegel, 2007). The monophasic type is closely related to the biphasic type and represents merely one extreme of its morphological spectrum, sharing phenotypical features identical to the spindle-cell portion or the epithelium-like component, corresponding to the monophasic fibrous variant or to the monophasic epithelial variant, respectively (Siegel, 2007). Histologically, the poorly differentiated type is composed mostly of small, solidly packed, oval or spindle-shaped cells that seem to have an intermediate phenotype between epithelial and spindle cells, often with scant differentiation, simulating other neoplasms, namely, angiosarcoma or small-cell carcinoma (Siegel, 2007).

Immunohistochemically, the majority of synovial sarcomas express cytokeratins, epithelial membrane antigen, calponin, B-cell lymphoma 2 (BCL-2) and CD-99. Vimentin can also be found in the spindle cells of these tumors. These markers can help differentiate synovial sarcomas from other sarcomas (Miettinen, 1984 and Beck, 2010). Although microscopic resemblance to the developing synovium was initially suggested, its origin from preformed synovial tissues remains to be proven (Ferrari, 2004; Miettinen, 1984). Owing to the similarity between synovial sarcoma tumor cells and primitive synoviocytes, the term synovial sarcoma was coined (Ferrari, 2004; Miettinen, 1984). However, most of these tumors occur outside the joints themselves and bear no resemblance to synovial structures either ultrastructurally or immunohisto chemically (Ferrari, 2004). It has been proposed that synovial sarcoma arises from the pluripotential mesenchyme of the limb bud (Siegel, 2007).

A particular chromosomal translocation t(X;18) has been noted in over 90% of cases, both in adults and in children (Ferrari, 2014 and Beck, 2010). Although synovial sarcomas can be graded histologically according to mitotic index, percentage of necrosis and tumor differentiation, almost all authors believe these tumors should always be regarded as high-grade sarcomas (Ferrari, 2014 and Beck, 2010). Synovial sarcomas not only are locally aggressive but also have a higher metastatic potential than most other soft-tissue sarcomas. Hence, the overall prognosis for synovial sarcoma patients is poor (Athanasian, 2011, Sultan, 2009 and Ferrari, 2004). In fact, according to most reports, notwithstanding intensive multimodal therapy, including surgery, chemotherapy and radiotherapy, the outcomes of these patients have changed little in the past two decades (Siegel, 2007 and Nakajima, 1997). According to the literature, local recurrence and/or metastatic disease are found in nearly 80% of patients (Athanasian, 2011; Sultan, 2009; Ferrari, 2004; Shi, 2012). Several factors have been associated with a higher recurrence risk. These factors include older age, larger tumor size (> 5cm), truncal location or proximal tumors in the limbs, male sex, bone or neurovascular invasion, incomplete excision on pathological examination, p53 overexpression, high proliferative index and, more recently, specific SYT-SSX fusion types (Wolden, 2010 and Eilber, 2008). The most common site for metastasis is the lung (Andrassy, 2001; Okcu, 2001). Lymph node involvement has been reported to occur in as many as 27% of patients (Okcu, 2001). Surgical resection is the definitive choice of treatment for primary synovial sarcomas and has been shown to both control local recurrence and prevent systemic dissemination (Athanasian, 2011; Sultan, 2009; and Ferrari, 2004). Unfortunately, the minimal acceptable margin has not been clearly established, and the surgeon must be aware of the possibility of microscopic infiltration of tumor cells into the pseudocapsule of the tumor (Murray, 2004; Siegel, 2007 and Wong, 2001). Many investigators have suggested 1 cm to 2cm resection margins (Murray, 2004; Athanasian, 2011; Siegel, 2007; Wong, 2001). Because of proximity to the joints, the ablation can consist of either tenosynovectomy and/or post-operative radiotherapy and/or chemotherapy, or simply extremity segment amputation

(Siegel, 2007). If ablation and tenosynovectomy are selected to retain maximal function, there might be a compromise of soft-tissue cover over tendons and neurovascular pedicles (Siegel, 2007). When proximity to critical anatomical structures and patient desire do not allow the surgeon to obtain adequate surgical margins, isolated limb perfusion and radiotherapy must be considered, as they can potentially prevent amputation. In all cases, multidisciplinary discussion of adjuvant therapies that may prevent amputation must occur prior to surgery (Murray, 2004; Athanasian, 2011; Siegel, 2007; Wong, 2001 and Okcu, 2001). Flaps are also an important option to bear in mind, as they can provide coverage of vital anatomical structures, as well as minimize the effects of radiation injury on these structures (Talbot, 2008). Discussion of the most adequate curative procedure, knowledge of the available reconstructive options, consideration of possible comorbidities and patient wishes for limb preservation must all be taken into account before surgery (Talbot, 2008). The efficacy of adjuvant chemotherapy is still a matter of intense debate (Okcu, 2001). Similarly, radiotherapy is associated with a higher rate of local disease control, but not with better survival rates (Miettinen, 1984 and Talbot, 2008). The presence of metastasis is considered the major cause of poor outcome, and several reports describing the results of current therapy showed a 5-year survival rate of around 27% to 55% (Athanasian, 2011; Sultan, 2009; Ferrari, 2004). Factors determining a worse prognosis include tumor diameter > 5cm, inadequate surgical resection, local recurrence, patient age over 20 years, monophasic variant and high mitotic activity (Athanasian, 2011; Deshmukh, 2004; Sultan, 2005; Shi, 2012).

Conclusion

Awareness of this rare tumor by surgeons dealing with hand pathology can hasten diagnosis, and this, in turn, can potentially increase survival.[9] Therefore, a high index of suspicion for this disease should be kept in mind, particularly when evaluating young people, as this is the most commonly affected group [3,7–9].

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