CASE REPORT

A PLUM ON THUMB: A RARE PRESENTATION OF SYNOVIAL SARCOMA

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Synovial sarcoma (SS) is a rare and aggressive mesenchymal tumour accounting for around 5–10% soft tissue neoplasms usually found in joints of upper and lower extremities. A 35years old healthy looking male patient from Afghanistan presented with swelling on palmar side of base of thumb from last one year. Seven months back excisional biopsy was taken report of which showed neurofibroma/dermatofibroma with. No evidence of malignancy seen. From last 5months mass reappeared and gradually increased in size with itching sensation and mild pain. On local examination there was 5×4×5 cm reddish mass on palmar surface of base of thumb with extension into mid thenar eminence with diffuse margins. X-ray showed soft tissue density mass with spikes of calcification. Ultrasound showed 4.2×4×4.5 cm heterogeneous solid lesion on anteromedial surface of root of right thumb without any remarkable intralesional calcification and remarkable intralesional vasculature. MRI reported lobulated well defined soft tissue mass eliciting low to intermediate signal on T1 and WIs and bright signal on T2and STIR Vividly enhancing mass. Case was operated mass was excised and biopsy sent. Post op status was unremarkable. Biopsy reported poorly differentiated biphasic synovial sarcoma. No recurrence seen till 3months.

Keywords: Synovial sarcoma; Mesenchymal tumour; Soft tissue neoplasms

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INTRODUCTION

Synovial sarcoma (SS) is a rare and aggressive tumour accounting for around 5–10% soft tissue neoplasms.[1] Thought to arise from mesenchymal or specialized arthrogenic tissue.[2,3] It is usually found in joints of upper and lower extremities. With or without synovial membrane involvement.[4] Diagnosis depends on histological and immunohistochemical analysis. Based on histopathology it has four subtypes, with biphasic SS as the most common one.[5] Treatment of choice is surgical excision with safe wide margins whenever possible, as effectiveness of chemo and radiotherapy yet need to be established.[6] Late diagnosis leads to poor prognosis in terms of mortality and morbidity. [7]

CASE REPORT

A 35years old healthy looking male patient from Afghanistan, *Mawlana* by profession presented to orthopaedic OPD with swelling on palmar side of base of thumb from last one year. According to patient he had on off pain from last 10 years on same site which was mild and rarely need any pain killers. A year back mass appeared which increases in size and was asymptomatic. Seven months back excisional biopsy was taken report of which showed Neurofibroma/dermatofibroma (Sections of benign tumour composed of bundles of nerve fibres showing wavy nuclei. No evidence of malignancy seen). From last 5months again mass appeared and gradually increased in size with itching sensation and mild pain. Past, family history was not significant with no smoking history. No co morbidities. General physical examination was normal, no anaemia or lymphadenopathy. On local examination there was 5×4×5 cm reddish plum like mass on palmar surface of base of thumb with extension into mid thenar eminence with diffuse margins. It was soft in consistency temperature comparable with surrounding, non-tender and adherent to underlying tissue. Inhibiting thumb movement because of its size. Rest of wrist and other fingers movements were normal with normal distal neuromuscular status. Trans illumination was positive. No axillary or trochlear lymph nodes were palpable. Systemic examination was unremarkable. HbsAg, anti HCV, Anti-AIDS screening was negative. Haemoglobin was 15.5g/l, platelets 295000, ESR and CRP level were in normal range. Xray showed soft tissue density mass on anterior and medial aspect of proximal phalanx of right thumb have spikes of calcification. High resolution B scan local ultrasound showed 4.2×4×4.5cm heterogeneous solid lesion on anteromedial surface of root of right thumb without any remarkable intralesional calcification. Colour Doppler showed remarkable intralesional vasculature. There is no intralesional cystic component seen. Erosion of adjacent bone could not be assessed. MRI reported lobulated well defined soft tissue mass (4.7×5.4×4.7 cm in TR×AP×CC) eliciting low to intermediate signal on T1 and WIs and bright signal on T2and STIR Vividly enhancing mass right hand involving mainly volar aspect and to less extent dorsal aspect of first interdigital space

extending into first inter metacarpal space, synovial sarcoma cannot be ruled out and histological correlation was recommended. Case was operated excision of mass was done and biopsy sent. Post op status was unremarkable. Biopsy reported poorly differentiated biphasic synovial sarcoma. On follow-up visits no recurrence seen till 3-months.

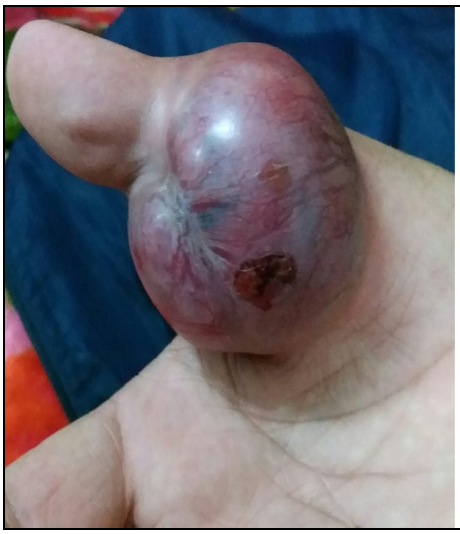

Figure-1: Gross appearance of thumb lesion (Anterior)

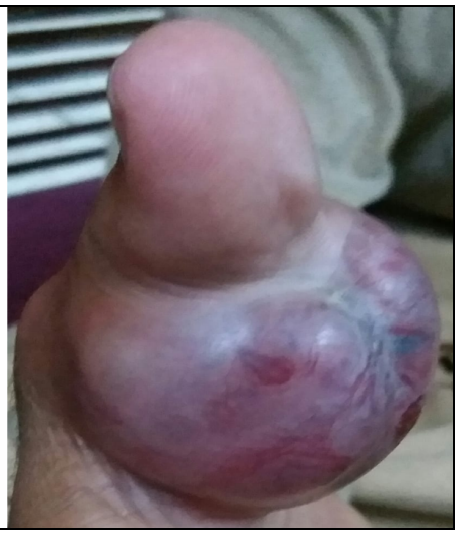

Figure-2: Gross appearance of thumb lesion (Lateral)

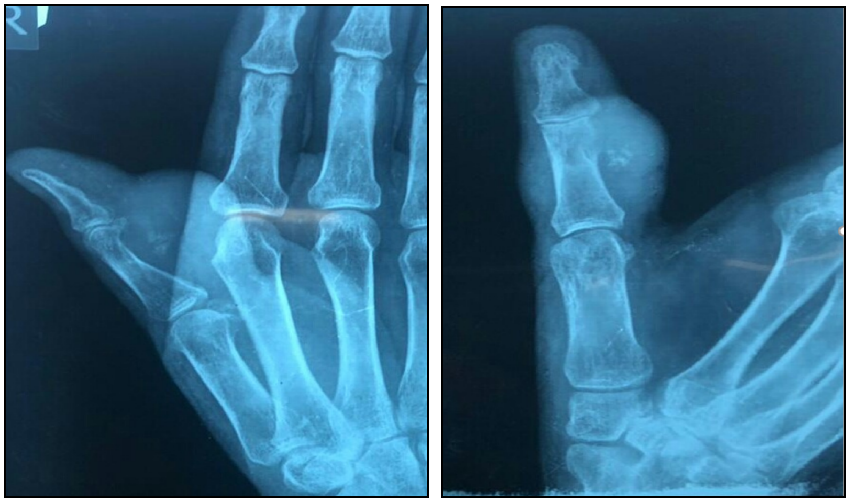

Figure-3: X ray of thumb lesion (Anterio-posterior and lateral view)

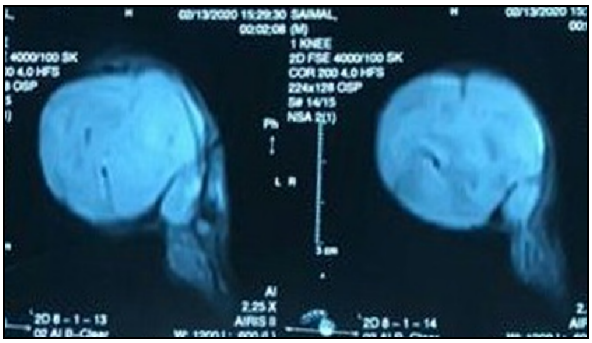

Figure-4: MRI of thumb lesion coronal section

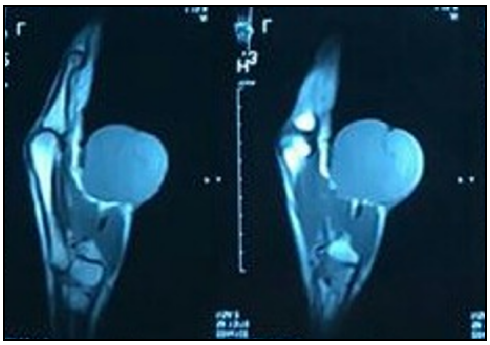

Figure-5: MRI of thumb lesion sagittal section

DISCUSSION

Soft tissue sarcoma of the hand is rare and the most common histological type is synovial sarcoma. The common age of involvement is 15–40 year.[8] Lower limb is the most affected area about 60% followed by upper limb 23% and the head and neck 9%.[9] Typical presentation is of slow growing mass.[10] Location of synovial sarcoma on the palmer aspect of hand and digit is very rare[11] and is the reason 38–95% of these cases are treated by unplanned excision[10]. Absence of distant metastasis at presentation shows favourable outcomes. Local or distant recurrence is very common accounting for about 80% of the case.[11,12]

Factors for higher recurrence rate are old age, male gender, size >5 cm, trunkal location or proximally located on the limb, bone or neurovascular involvement, incomplete excision, p53 over expression and SYT-SSX fusion types.[11,13] The standard treatment protocol is wide excision followed by adjuvant radio and adjuvant chemo.[12] Regional lymph node dissection is not required for synovial sarcoma of hand.[12] One to two cm resection margins should be taken into consideration.[13] The role of adjuvant chemotherapy is debatable.[11] Adjuvant radiotherapy has high rate of local control but it is of no benefit in the outcome of survival, 5 year survival rate is 27–55%.[11,13,14]

In our case unplanned excision done and the histopathological report was showing the diagnosis of neurofibrom/dermatofibroma followed by recurrence of mass within two months. This early recurrence can be attributed to the unplanned surgery and incomplete excision. Three months follow up showed good result in term of function of the hand and there is no recurrence of mass showing that properly planned surgery with clear margins are very imp to prevent recurrence and in case of suspicion of mass to be synovial sarcoma the patient should be refer to orthopaedic oncologist or hand surgeon that have expertise in the hand tumour cases as we did, surgery been performed by hand surgeon.

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