Case Report

Synovial Sarcoma of Foot: A Case Report

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Abstract

Synovial sarcoma (SS) is a soft tissue tumor with high malignant potential. It is a stem cell malignancy. It is usually found on the extremity. More than 90% of the SS have characteristic translocation i.e. t(X;18) (p11.2; q11.2). Types of SS include monophasic, biphasic and poorly differentiated type. Imaging, histology, immunochemical staining and chromosomal analysis is the main modality of diagnosis. Surgery with pre- or post-operative irradiation is the main modality of its treatment. However the role of chemotherapy is still controversial. 5 – year survival of this locally aggressive tumor is poor. Prognosis depends upon age, size, and histological type of tumor.

Key Words: Synovial sarcoma, monophasic, biphasic.

Case Report

An 18 year old lady presented to our outpatient department (OPD) with chief complaint of a mass present on the dorsum of foot for last one year. According to her she has insidious onset of reddish discoloration of skin on the dorsum of left foot 2 years back. This was followed by formation of a nodule. The patient was managed at other centre with excision of the nodule. There appeared a sprouting mass with discharging sinus approximately 1 month after the excision of nodule. The patient also developed pain and was unable to bear weight on her left foot. The mass was again excised 1 year prior to presentation to usat other centre with wide margin where the histopathology confirmed it to be a monophasic synovial sarcoma. The patient has now presented to us for definitive management. There is no history of fever, appearance of mass in

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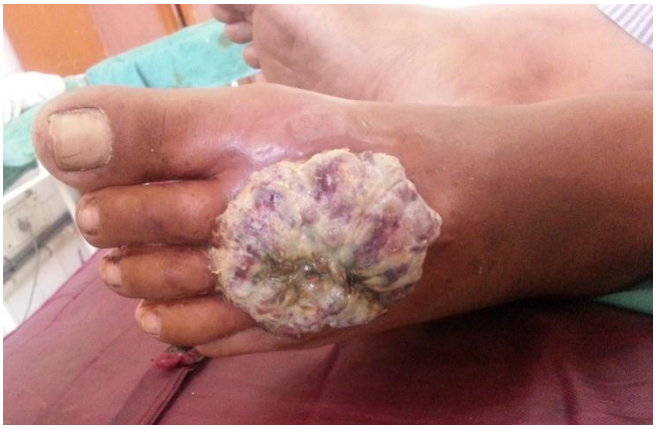

Fig. 1: Mass on the dorsum of left foot with edema.

other part of body and recent significant body weight loss.

On examination hergeneral condition was fair with normal body built and no pallor, icterus, edema and lymphadenopathy. Her pulse was 88beats per minute, respiratory rate of 18 cycles per minute, blood pressure of 110/80 mm of Hg and temperature of 38.8°c.

On local examination there was a mass of 8 × 8 cm on the dorsum of left forefoot with pus present at the centre. It was soft in consistency and was friable. The surrounding skin was erythematous and edematous. There was local rise of temperature and the mass was tender. The mass was fixed with underlying structures. The dorsalis paedis artery was palpable and the movements of metatarsophalyngeal and interphalyngeal joints were restricted.

The abdomen and chest were within normal limit.

Her erythrocyte sedimentation rate was increased to 44 in 1 hour with normal total count.

X-ray of the foot showed thinning of the cortex of 2nd and 3rdmetatarsal (MT).

MRI of the foot showed an irregular extensive infiltrating mass arising from dorsum of left foot encasing the 2nd and 3rd MT completely and 60 to 70% circumference of 4th MT. the mass is also infiltrating the marrow of involved MT. The medial margin of the mass is encroaching the cortex of 1st MT however its marrow is not involved. The mass is of 7.5 × 6 × 5.8 cm. its proximal limit is up to the bases / proximal limits of MT/ cuneiforms. On plantar aspect it involves deep muscle layer namely lumbricals and interossi however the tendons of flexor digitorum superficialis is normally visualized.

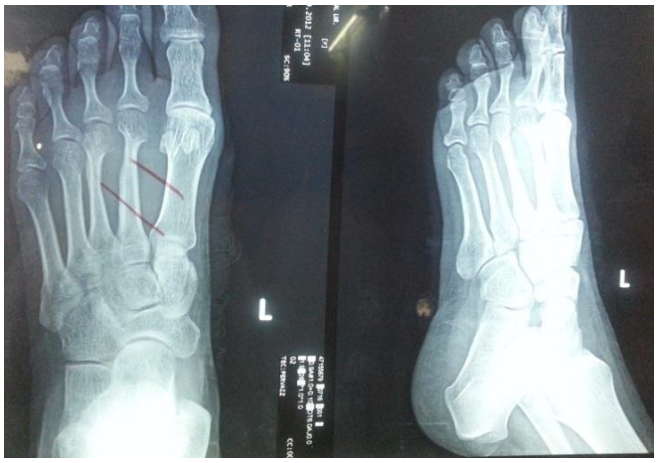

Fig. 2: X-rays AP and oblique view of left foot: Thinning of the cortex of 2nd and 3rd MT.

Computerised tomography (CT) of chest and abdomen showed no metastasis.

Radionuclide skeletal scintigraphy showed slightly increased uptake of radionuclide involving distal part of 2nd and 3rd MT of left foot. Rest of the bone scan was normal.

Histopathology showed spindle cell neoplasm favoring synovial sarcoma.

Immunohistochemical staining showed cytokeratin positive in epitheloid areas, Bcl_2 and CD_{99} was positive in tumor cells and CD_{34} was negative.

The patient was diagnosed as a synovial sarcoma of left foot and was managed with forefoot amputation.

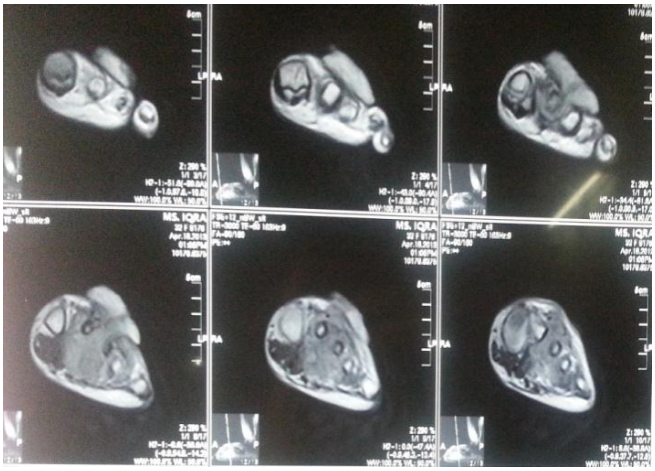

Fig. 3: T_2 weighted transverse section of left foot: mass involving 2nd, 3rd and 4th MT with soft tissue involvement.

Discussion

Synovial sarcoma (SS) is a malignant soft tissue tumor. It is a misnomer as it has no component from synovium rather it is a stem cell malignancy.[1]

SS comprises 6-9% of all adult soft tissue sarcoma. It comprises 12 – 15% of soft tissue tumor of extremity. It is common between 3rd and 4th decade of life.[2-5] There is no predisposition for either sex. No known genetic conditions or etiological agents for this tumor.

71% of its primary site of origin is from lower extremity, 16% from upper extremity and 13% are from other areas. Most common extra-extremity sites

of primary tumors are trunk (13%), retroperitoneal (rare), and head and neck (rare).[6] Like other soft tissue sarcoma, SS with its primary origin from extremity mostly metastasizes to lungs and intra-abdominal metastasis are rare. Its nodal involvement frequency is higher than other soft tissue sarcoma.

Histological SS can be divided into monophasic, biphasic and poorly differentiated type. Monophasic SS is entirely composed of ovoid spindle shaped cells while that of biphasic SS is composed of both spindle cells and epithelial cells. Poorly differentiated type consists of cells resembling small round blue cell tumor. One third of the tumor shows calcification. Cystic changes and necrosis may also be seen.[7,8]

The patient usually presents in their thirties with a mass on extremity which is spontaneous in onset, progressively increasing in size and associated with pain. The patient may present in earlier or late stage of the disease depending on the health awareness of the community. 82% presents with localized disease and 18% with metastatic disease at the time of presentation.[6] There are situations where the patient may present with secondaries first and the primary later.[9]

Imaging, histology, immunohistochemical staining and chromosomal analysis are the main tools to reach to a diagnosis.

Calcification may be seen on the plain x-ray films. On CT, peripheral "punctuate" pattern of calcification is seen in 42.5% of synovial sarcoma.[10] MRI is the major modality of imaging for the detection and staging of soft – tissue tumors. Imaging features associated with a poor prognosis include a large tumor size of .10 cm, absence of calcification and hemorrhagic appearance.[11]

Immunohistochemical assist in diagnosing SS. Epithelia CAM 5.2 and EMA are most helpful. Others like Bcl_2 is also positive in SS. CD99 and S100 is positive in 60% and 40% respectively. Immunohistochemical stain helps SS to differentiate from malignant peripheral nerve sheath tumor, leiomyosarcoma and Ewing's tumor.[8] Fluorescence in situ hybridization (FISH) and Reverse – Transcription – Polymerase Chain Reaction (RT – PCR) is highly sensitive and specific test.[8]

More than 90% of the SS have characteristic translocation between chromosome 18 and chromosome X. This translocation i.e. t(X;18) (p11.2; q11.2) involves SYT gene on chromosome 18 and SSX1, SSX_2 and SSX_4 on chromosome X. SYT_1 / SSX_1 is the most common translocation occurring in biphasic type of SS.[8] Rare translocation of SYT / SSX_4 occurs in monophasic and poorly differentiated SS. These transcripts can be detected by PCR thus aiding in diagnosis. This X; 18 translocation being unique to synovial sarcoma, its presence provides a definitive diagnosis, even in cases with unusual clinical or histologic features.[12-14] Expression of Transducin – Like Enhancer 1 (TLE_1) is significantly correlated with t(X;18) and may serve as a new robust diagnostic biomarker in synovial sarcomas and potential therapeutic target.[15]

Surgery, radiotherapy and chemotherapy remain the method for treatment of synovial sarcoma.

Surgical treatment involves resection of tumor with wide margin and maximal preservation of function. Different investigators has suggested adequate margin of 1 to 3 cm.[16] Un-resectable tumors are followed by amputation of the limb with preservation of its maximum function. Patient with secondaries in lungs can be treated with metastasectomy. If complete resection can be achieved, pulmonary metastasectomy may be associated with improved survival in pediatric / adolescent patients with SS with pulmonary metastases.[17]

Adjuvant radiation therapy is used in patients with tumors $\geq$ 5 cm in size.[18] Radiation depends on the primary site and size of the tumor. Radiation field includes tumor along with 2 to 3 cm of tumor margin.[16,19] Adjuvant radiation therapy can be administered in a number of ways such as external beam therapy (neoadjuvant or adjuvant), brachytherapy, and intensity modulated radiation therapy (IMRT). Radiotherapy has good role in local control of tumor. Irradiation of the tumor can be done either preoperatively or postoperatively. Regardless of the type of radiation therapy employed, it has been proven to improve the local control rate in patients with high grade sarcomas, such as synovial sarcoma.[20]

Ifosfamide based chemotherapy can be used for SS associated with metastasis.[8] Different other medications based adjuvant and neoadjuvant chemotherapy has been devised but because of their toxicity, its use is controversial.[21-24]

Age, size, histology, and use of radiotherapy influence prognosis.[21] Doubtful benefit for survival benefit with chemotherapy has been seen in many series.[21-24] Synovial sarcoma can adequately be controlled at the primary site by conservation surgery and RT. Monophasic has better prognosis than biphasic. Rates of development of distant metastasis and subsequent death from disease remain high.[21,25] 21% of patients with primary extremity sarcoma who survive for 5 years will die of disease within 5 years. Despite abse-

nce of metastasis by 5 years, 9% will die of disease within 5 years. Patients with positive microscopic margins are at risk for post – 5 – year disease – specific mortality and therefore require long-term follow.[4]

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