

CASE REPORT

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Synovial Sarcoma of the Right Foot: A Case Report

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Abstract

Synovial sarcoma is one of the soft tissue sarcomas especially seen mostly in children and young adults. It is highly aggressive with significant chances of metastasis. It is the one of the soft tissue sarcomas caused due to chromosomal aberration i.e., translocation and most well-known "translocation-associated sarcoma". The present case study presents a patient of age 25yrs with a complaint of swelling in right lower leg with pain on walking and had a previous trauma to leg 3 years ago. On investigation, the biopsy report, microscopic evaluation, IHC, MRI, PET CT revealed the presence of synovial sarcoma. Syme amputation was performed, and patient is advised to attend regular follow-ups.

Keywords: Synovial sarcoma; Foot disease; Syme amputation

1 Introduction

Synovial sarcoma is a rare soft tissue sarcoma (STS) with uncertain differentiation. It accounts for 5-10% of all soft tissue sarcomas. Unlike other STS, synovial sarcoma affects both sexes equally and usually appears at a younger mean age of onset, occurring in adolescence and young adults (mean age of 39 years at diagnosis).¹

It is possibly the prototypical "translocation-associated sarcoma," as it was one of the first sarcomas to be identified by the presence of a specific chromosomal translocation that results in the development of the SS18-SSX

fusion oncogene, and its translocation remains unique to this type of tumor.² One pathognomonic feature of synovial sarcoma is the presence of t(X;18)(p11.2;q11.2).¹

Although synovial sarcoma has a variety of morphologic patterns, the two most common are the classic biphasic pattern, which includes solid or glandular epithelial structures with monomorphic spindle cells, and the monophasic pattern, which includes fascicles of spindle cells that only show ultrastructural or immunohistochemical evidence of epithelial differentiation.²

In general, this tumor is seen in the distal extremities, particularly the lower

limb. We present a rare case of synovial sarcoma of the right midfoot in a 23-year-old male treated with surgical resection and split skin graft.

2 Case Report

Study Place : R L Jalappa Hospital, Tamaka, Kolar, Karnataka

A 23Yr old male patient presented with history of trauma to his right foot came to hospital with a complaint of swelling. Trauma occurred 3years back following which he noticed swelling over dorsum of right foot.

Swelling is insidious in onset, gradually progressed to present state and size which is around 5x4x3cm. Skin is stretched over the swelling, surface is smooth and firm in consistency. Pain increased on walking and no relieving factor associated. Pricking type of pain is noticed with moderate in severity. History of weight loss noticed.

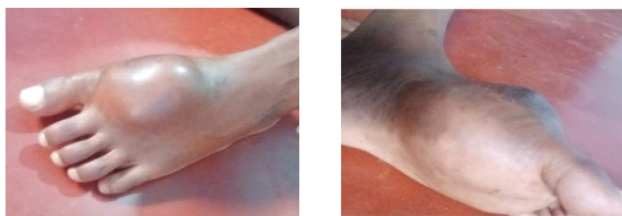


Fig 1. Clinical presentation of lesion

External surface - Grossly, skin appears to be free. Also noted tendons of extensor digatae, Calcaneus and talus bones. Cut surface is identified as well circumscribed and capsulated grey, white lesion measuring 8.5x8x7 cms. Grossly tumors appear to be involving cuneiform bone, 1st, 2nd, 3rd, 4th metatarsals, plantar muscles.

Tumor appears to be involving dermis. The tumor is 7.5 cms from posterior bony margin. The tumor is 3 cms from tip of toe (anterior margin). The tumor is 1 cm from plantar aspect (inferior margin). The tumor is 0.1 cm from superior margin (skin).

3 Investigations

3.1 Microscopic evaluation

On microscopic evaluation, tumor tissue consists of spindle cells having spindle shaped nucleus with blunt ends, cytoplasm is moderate and eosinophilic. At focal areas cell shows moderate to severe anisonucleosis with fine chromatin. Mitotic figures are 1-2/ HPE. These cells are arranged in whorled pattern and at places herring bone pattern is seen. The tumor tissue is invading the adjacent skeletal muscle. Focal areas show lymphocytic infiltrates within tumour tissue. Section studied from anterior skin margin, inferior margin, superior margin shows no evidence of tumor deposits.

Section studied from anterior and posterior bony margin are negative for tumor involvement. Section studied from 1st, 2nd and 3rd cuneiform bones show tumor involvement.

Features are of Mesenchymal tumor possibility of:

1. Synovial sarcoma (Monophasic variant, spindle cell variant).
2. Low - Grade fibrosarcoma.

3.2 MRI

Multipplanar multiecho MRI of foot has been performed. Large lobulated heterogenous intensity mass lesion is noted along with the plantar as well as dorsal aspect of medial midfoot extending into first and second web foot spaces. MRI findings are suggestive of soft tissue neoplasm.

3.3 IHC reports

IHC report is done with satisfactory controls. The neoplastic cells are diffusely positive for TLE1, CD99. They are negative for SMA, desmins, S100, CD 34 and CK. Ki 67 index is 20 %. Immunohistopathological features are suggestive of synovial sarcoma, biopsy of right foot.

3.4 PET CT Scan

PET scan was advised and the reports showed a large lobulated heterogeneously enhancing soft tissue lesion with low-grade, FDG uptake is seen along the medial aspect of right foot predominantly in the region of the mid foot showing extension into the dorsum as well as plantar aspects measuring -9.0 x 6.4 x 7.6 cm (Max SUV 3.3); it is predominantly involving the intra/ intermuscular planes with extension into the subcutaneous plane; anteriorly, it is extending medial to the great to and into the first and second web spaces; a tiny calcific focus is seen along the superior aspect in the dorsum closely abutting the skin; bulging of the skin with surface contour deformity is seen more in the dorsum of foot; few subtle cortical erosion are seen in the 1st, 2nd and 3rd metatarsals at places; rest of the surrounding bones in the right foot appear unremarkable.



Fig 2. Gross lesion after amputation of leg

The patient underwent Syme Amputation of Right Foot. After the operation, the patient continues to attend for follow up.



Fig 3. Amputation of right leg

4 Discussion

Synovial Sarcoma was termed due to its frequent presence in the soft tissue surrounding large joints, particularly the knee, and was thought to be caused by synovial cells.² The most common location for synovial sarcomas is the lower extremities. Patients appear with a palpable, slow-growing, and occasionally painful mass. Owing to the subtle onset, diagnosis is frequently delayed.^{1,3}

Histologically, synovial sarcoma can be divided into four types: monophasic spindle-cell, monophasic spindle-cell, poorly differentiated, and biphasic spindle-cell. The weakly differentiated spindle-cell type has the worst prognosis, and biphasic and monophasic spindle-cell types predominate.

Cytogenetic and molecular genetic studies have conclusively shown that translocation of t(X; 18) is only related with synovial sarcoma. The cause is yet unknown, though.

Synovial Sarcoma are multinodular masses that range in size from less than 1 cm to 15 cm in diameter. The cut surface can be solid, firm, and fleshy smooth and glistening or tan, gritty, and calcified. Calcification is frequent, yet it might be difficult to detect.²

In a study by Bakri A et al³, CT often shows a noninfiltrative, well-circumscribed mass, often with punctate, peripheral calcifications, which is consistent with the current study's PET CT results.

Complete excision and surgical resection, with or without radiation, have been shown to be beneficial in establishing tumor control locally. Certainly, tumor response has occurred

with first-line chemotherapy regimens that include ifosfamide (with or without doxorubicin).³ However, surgical excision with negative margins remains the primary treatment for synovial sarcoma, with the addition of radiation and/or chemotherapy depending on the patient and tumor features.

In the past, patients were typically treated with amputation, but developments in adjuvant therapy and cross-sectional imaging have enabled the majority of patients to be treated with limb-salvage surgery. Negative surgical margins are extremely important because they predict both local recurrence and overall survival.³

A Syme amputation involves disarticulation of the tibio-talar joint and resection of the malleoli. The amputation was first performed in 1842 by James Syme (1799-1870). The typical Syme amputation is a one-stage surgery. Later, a two-stage treatment was developed, with the first stage used for debridement in the event of infection with merely a loose closure, and the second stage for malleoli trimming and skin flap revision.⁴

Reamputations at a higher level were performed in 0% of children and 20% of adults during the follow-up periods of the included studies.

The objective information derived from this investigation supports the opinion that patients who underwent Syme's ankle disarticulation amputation appear to fare better than similar patients with transtibial amputation.^{5,6}

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