University Journal of Surgery and Surgical Specialities

ISSN 2455-2860 2018, Vol. 4(1)

Double Primary and Multiple Metachronous Primary Sarcomas

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Abstract : Soft tissue sarcomas are very rare, with median age of presentation being 51 to 80. Most common site of sarcomas are in lower extremities followed by head and neck region. The incidenceof multiple metachronous sarcoma is rarer.Patients and methods A review of the departmental sarcoma database following the presentation of this index case in the clinic. Results and discussion A 68 year old male who had been previously treated with surgery for degenerated schwannoma over left infra-scapular region, developed Fibrosarcoma right thigh after 1 year of first surgery, for which he was surgically treated. 7 months after second surgery, he developed soft tissue sarcoma (MFH) of right leg for which he was operated upon. Subsequently he developed Soft tissue sarcoma of back. A review of the Indian literature and departmental sarcoma database, restricted to soft tissue sarcomas and schwannoma, could not identify any other such case. The purpose of this report is to increase awareness amongst clinicians regarding such a possibility of multiple primary tumours.

Keyword : Metachronous, Multiple Primary , Schwannoma , Sarcoma

DOUBLE PRIMARY AND MULTIPLE METACHRONOUS PRIMARY SARCOMAS.

INTRODUCTION

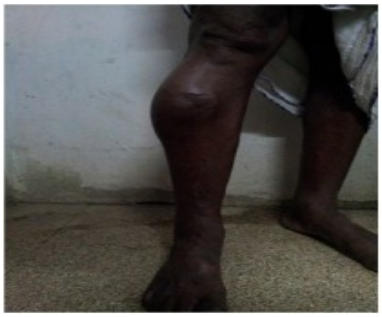
Soft tissue sarcomas (STS) are very rare[4.] It's incidence in general population is 3.2 per 10000 with median age of presentation being 51 to 80[1]. Most common site of sarcomas are in lower extremities followed by head and neck region.[8] The incidence of multiple metachronous sarcoma is 4 per 10000. 1,3

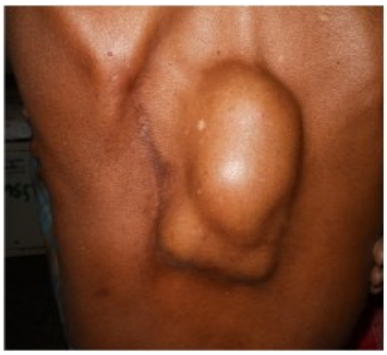
CASE REPORT:

68yrs male presented with 15 x 10 cm swelling in posterolateral aspect of the right thigh for 1 year in the subcutaneous plane without muscle involvement. Trucut biopsy was suggestive of Fibrosarcoma. Wide excision and split skin grafting was done, followed by adjuvant chemotherapy.

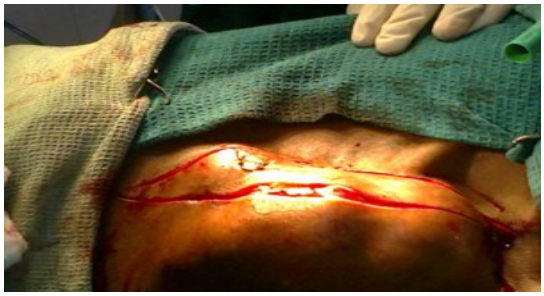
Patient had been previously treated for swelling in the left side of the back (infrascapular region) which was excised, histopathological examination of which revealed degenerative Schwannoma.

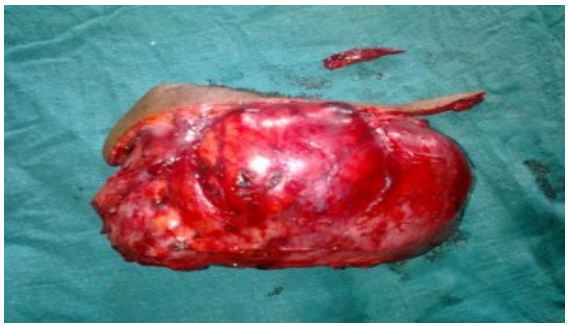
After 7 months he came came back again with complaints of swelling in the lateral aspect of the right leg of 5 months duration. On examination there was a 10 x 7 cm size soft tissue swelling without invoving underlying structures. Tru-cut biopsy revealed to be malignant fibrous histiocytoma- pleomorphic type. Wide excision and split skin grafting was done.showed normal lungs. Trucut biopsy revealed malignant fibrous histiocytoma-infiltrating type. Wide excision with primary closure was done



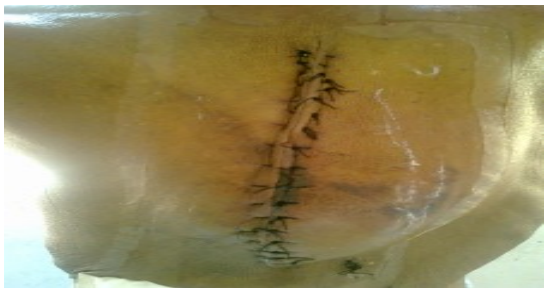


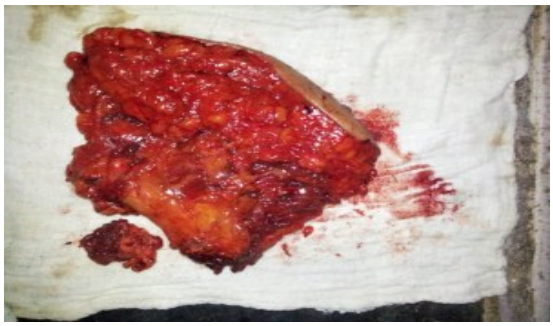
MFH of Right Leg
MFH of Back soft tissue sarcoma back



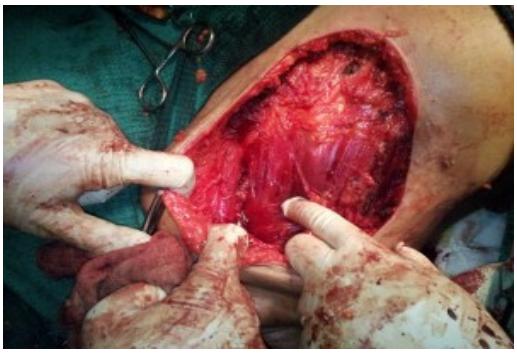


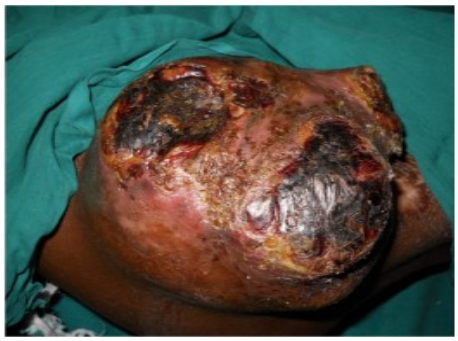
soft tissue sarcoma back soft tissue sarcoma back



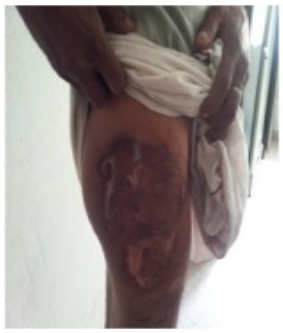


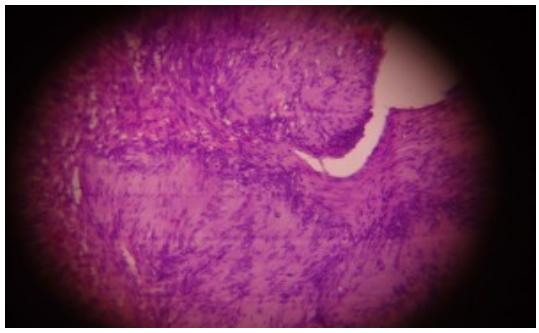
soft tissue sarcoma leg sft tissue sarcoma leg



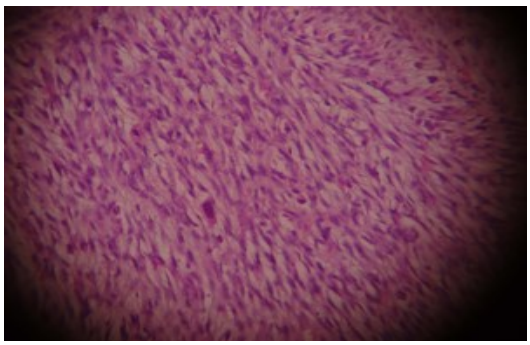


Fibrosarcoma Right Thigh Post-op Right Thigh





Schwannoma Malignant Fibrous- Histiocytoma



REVIEW OF LITERATURE Sarcoma means FISH FLESH or fleshy growth i.e. the tumour's tendency to feel fleshy when palpated.soft tissue sarcomas are rare[4]. Its incidence in general population is 3.2 per 10000[1.] Affected pts have a increased risk of developing a second primary malignancy, particularly another primary STS[4, 5.] Distinction between metastases and second primary STS is difficult[5]. Incidence of multiple metachronous soft tissue sarcomas are 4 per 10000[1, 3]. Median age of pt at the time of presentation with sarcomas was 51-80years[1].Median interval between two sarcomas for diagnoses is 4 months to 9 ½ year[2].most common site of primary STS are in lower extremities followed by head and neck region[8]. But metachronous multiple soft tissue sarcomas at extremity is very rare[2]. Most primary tumours that are greater than 10cm size are of high grade malignancy.[6]

Classification based on tissue of origin:

1. Fibrous tissue: Fibrosarcoma.
2 Fibrohistocytes: Malignant fibrous histiocytoma.
3. Adipose tissue: Liposarcoma.
4. Smooth muscle: Leiomyosarcoma.
5. Striated muscle: Rhabdomyosarcoma.
6. Blood vessels: Haemangiosarcoma.
Kaposi sarcoma. Malignant glomus tumour.
7. Lymph vessels: Lymphangiosarcoma.
8. Synovial tissue: Synovial sarcoma.
9. Mesothelial tissue: Mesothelioma.
10. Peripheral nerves: Malignant schwanoma. Neuroepithelioma.
11. Autonomic ganglion: Neuroblastoma.
12. Tumour of Pleuri-potential mesenchyme: Malignant mesenchymoma.

Sarcomas spread through blood by embolic phenomenon or from intraoperative manipulation of neoplasm to lungs, liver, bone6. Lymph node and brain metestasis is very rare 7, 8.

MALIGNANT FIBROUS HISTIOCYTOMA:

This is the most common sarcoma in adult.50 -75% occur in extremities especially lower extremities. Other sites are shoulder, trunk, retroperitoneum. Mean age 5th and 6th decade.

TYPES: Giant cell, fibrous, myxoid, inflammatory-
, angiomatoid, pleomorphic.

FIBROSARCOMA:

It constitutes 14% of all soft tissue sarcomas. Peak age of incidence is from 4th and 5th decade. A slowly growing tumour arising from fascial connective tissue, inter and intramuscular fibrous septae. 50 -60% in lower extremity. Thigh is the most common site. It is related to trauma, burns, scars, injection sites, ulcers, post-irradiations.

TYPES:

1.Congenital and infantile fibrosarcoma.
2. Adult fibrosarcoma.
3. Inflammatory sarcoma.
4. Cicatricial, post irradiation.

SCHWANNOMA:

Schwannoma is one the few truly encapsulated neoplasms. It's most common locations are flexor surface of extremities, neck, mediastinum, retroperitoneum, cerebello-pontine angle. Since this is a benign neoplasm, it rarely recurs locally. Great majority of cases occur sporadically, small percentages of cases are associated with NF type2.

ANCIENT SCHWANNOMA:

It is a variant of schwannoma with Isolated cells with bizarre hyper-chromatic nuclei. Mitoses are usually absent or extremely scanty.

INVESTIGATIONS:

X-ray of the part: may show soft tissue shadow, any calcification or any erosions of underlying metastasis if at all any.
X-ray chest- to rule out lung metastasis.
FNAC useful only in recurrent sarcomas.
Trucut biopsy may be more useful than FNAC.
Excisional biopsy if<3cm.
Incisional biopsy if>3cm.
CT and MRI: accurately delineate the muscle compartment or anatomic structure involved by disease.

Chest CT as part of staging evaluation in sarcomas rule out lung metastasis9.
Surgery is the treatment of choice.
Wide excision with 2-3 cm margin clearance is done with or without split skin graft.

CONCLUSION:

Multiple primary and multiple metachronous sarcomas are unusual in occurrence. Patients who have history of soft tissue sarcoma are at an increased risk for the development of second primary STS All soft tissue sarcomas need regular follow up and metachronous sarcomas should be treated adequately and aggressively for better results and salvage.

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University Journal of Surgery and Surgical Specialities