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CASE REPORT

MALIGNANT FIBROUS HISTIOCYTOMA OF THE EXTREMITY: REVIEW OF LITERATURE & CASE REPORT

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ABSTRACT: Malignant fibrous histiocytoma is a soft tissue sarcoma found in young & elderly and rarely in children. It is rarely confined exclusively to skin and subcutis. In most cases it is only diagnosed after excision and analysis of the tumour. It is aggressive presenting a high degree of local recurrence and metastasis. This article reports a case of malignant fibrous histiocytoma on an extremity of 73 year old patient.

KEY WORDS: Histiocytoma; Excision: Radiotherapy; Recurrence.

INTRODUCTION: MFH of soft tissue typically presents in a patient that is approximately 50 to 70 years of age though it can appear at any age. MFH is very rare in persons less than 20 years old. There is a slight male predominance. Soft tissue MFH can arise in any part of the body but most commonly in the lower extremity, especially the thigh. Other common locations include the upper extremity and retroperitoneum. Patients often complain of a mass or lump that has arisen over a short period of time ranging from weeks to months [1]. It most commonly spreads (metastasizes) to the lungs, but can also invade the lymph nodes and bone. Studies suggest association of genetic abnormality on the short arm of chromosome 19 that may give rise to this disease. Malignant fibrous histiocytoma (MFH), a type of sarcoma, is a malignant neoplasm of uncertain origin that arises both in soft tissue and bone. It was first introduced in 1961 by Kauffman and Stout. MFH manifests a broad range of histologic appearances with four sub-types described as Storiform-pleomorphic, Myxoid, Giant cell & Inflammatory. Of these, the storiform-pleomorphic is the most common type, accounting for up to 70% of most cases; the myxoid variant is the second most common accounting for approximately 20% of cases. The different modalities in management of MFH include surgery, radiotherapy and chemotherapy. The main stay of treatment is radical surgery in past to limb sparing surgery in recent time. The purpose of radiation is to improve local tumor control by killing residual microscopic disease. Radiation has been clearly shown to improve the incidence of local recurrence and has become an integral part of the treatment for MFH. The most commonly used form of radiation is external beam radiation which can be given pre-operatively, intra-operatively, post-operatively, or in some combination. More recently, clinical trials incorporating ifosfamide and doxorubicin have demonstrated an improvement in disease-free survival. This work reports the case 73 year old female with cutaneous malignant fibrous histiocytoma and underscores the importance of early diagnosis of this neoplasia.

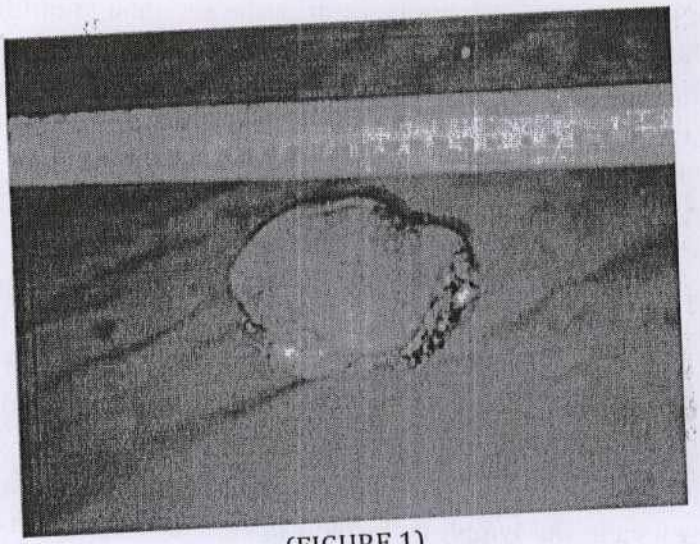
CASE REPORT: A 73 year old female presented with history of swelling in right lateral aspect of thigh since four months. She also reported that one year back patient had similar complaints and it was excised and then lost the followup.

At clinical examination she presented with a solitary swelling of size 4*3 cm in the right lateral aspect of thigh, with previous surgical linear scar, non tender, mobile, superficial to muscle plane. Suggesting a clinical diagnosis as soft tissue sarcoma.

FNAC C/1400/10: reported as Malignant spindle cell tumour.

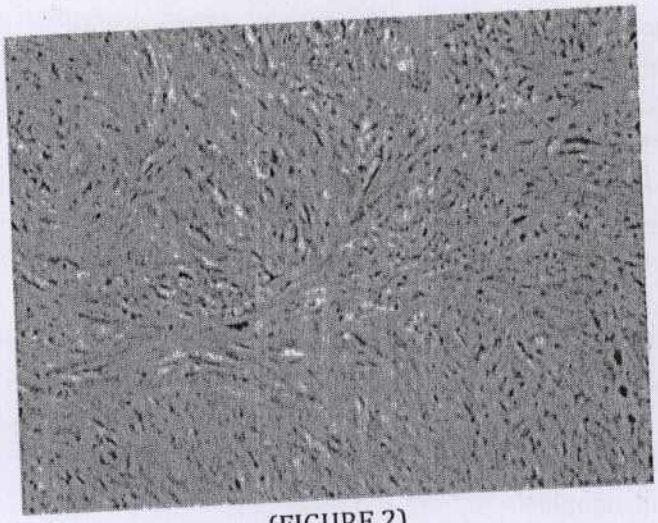
Blood count, urine summary, transaminases, bilirubin, urea, creatinine, CT of skull, chest x ray and ultrasound scan of total abdomen were normal.

Wide excision of tumour tissue was done with primary closure of the wound on 7/6/2010.



(FIGURE 1)

Histopathology report (B/1355-10) revealing Malignant fibrous histiocytoma –giant cell type.



(FIGURE 2)

FIGURE 2 - A highly cellular tumour consisting of spindle to round polygonal cells with hyperchromatic nucleus with coarse chromatin and also multi nucleate giant cells are seen. After surgery patient is disease free till date and she is on radiotherapy.

DISCUSSION: It is a sarcoma with a high degree of polymorphism and capacity for producing collagen, but without other defined characteristic [2]. Approximately two thirds of these tumors are located in the skeletal muscle.

According to their location they can be classified into superficial and profound. The superficial form is very rare and is confined to the skin and to the subcutaneous tissue; it may be adhered to the fascia. The profound form extends from the skin along the fascia until the muscle, or it can be located entirely within the muscle [3].

Ample excision is recommended, due to the high degree of local recurrence (approximately 44%) and metastasis, most commonly in the lungs (approximately 42% of cases) [4]. The presence of metastasis is usually associated with a poor prognostic. Primary MFH of the skin can have a more favorable prognostic than homologous tumor originated in more profound and retroperitoneal soft tissue [5].

Wide excision of the tumour tissue was performed due to great extension of the lesion involving the greater part and according to reports in the literature due to the high risk of local recurrence and metastasis.

Nevertheless, in this case it might have been possible to avoid recurrence if the patient had followed up to us with histopathological report of the previous surgery showing malignant fibrous histiocytoma for which wide excision was not done, hence the recurrence. As these histiocytomas can be precisely diagnosed only by histopathological examination the excised tumour tissue should be subjected to histological examination so that no lesion is missed and recurrence is avoided.

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