Bleeding soft-tissue sarcoma: rare presentation of upper extremity sarcoma

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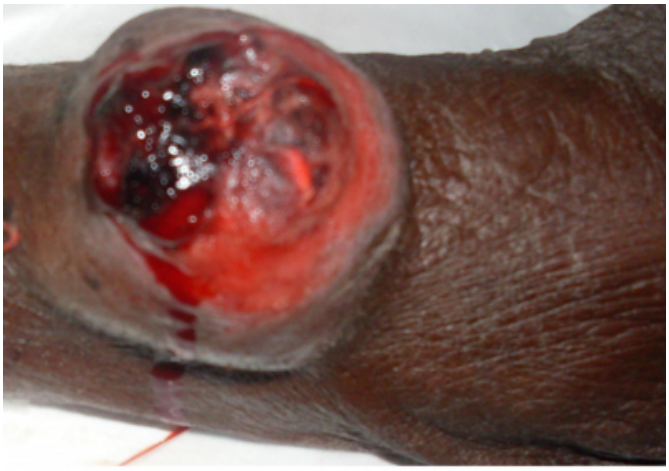
Abstract

Background:Soft-tissue sarcomas comprise <1% of all malignancies in the body. Common presentation is a painless large lump or swelling in the extremities or retroperitoneum. Bleeding is a rare presentation. Upper extremity sarcoma is a less common site. The mainstay of treatment is surgery.Method:A case report with review of literature is presented in brief.Result:Overall prognosis of soft-tissue sarcoma is not good.Conclusion:Nowadays, limb-sparing surgery is the gold standard for management of soft-tissue sarcoma. Amputation should be reserved for sarcoma involving bone, nerve or vessels.

CASE REPORT

An 80-year-old gentleman came to the surgical outpatient department with a swelling for the last 6 months, an ulcer over the swelling for the last month and bleeding for the last day on the left upper limb. No other significant history was available. On examination, there was an 8×8cm swelling which was rounded with a bleeding ulcer of about 6×6cm (photograph). The consistency of the swelling was soft with fixity to the skin, but without fixity to underlying structures. There was no evidence of axillary lymphadenopathy. The distal neurovascular bundle was normal. Respiratory and abdominal system examination did not reveal any evidence of distance metastases. X-ray of the local part and chest x-ray were normal. USG of the abdomen was normal. Four-quadrant biopsy taken from the ulcer showed high-grade malignant fibrous histiocytoma. Complete blood count, liver function and coagulation profile were normal on the day of admission. On the next day, hemoglobin dropped from 11g% to 6g%. In view of high grade, old age, size of more than 5cm, complex karyotype and close proximity to the elbow joint, an above-elbow amputation was done and post-operative recovery was uneventful.

Figure 1



Bleeding soft tissue sarcoma

DISCUSSION

Soft-tissue sarcoma is a term used for a heterogenous group of malignancy arising from the mesodermal cell. Soft-tissue and bone sarcoma account for <1% of cancer in the body[1]. Approximately 10,000 new cases have been registered in the United States every year, among which <50% die of the disease. Most soft-tissue sarcomas arise in the extremities, followed by retroperitoneum and other sites[2]. Most soft-tissue sarcomas have no clearly defined cause, genetic factors (like mutation in the p53 gene, association with neurofibromatosis), chemical factors (thorotrast, vinyl chloride etc.), ionizing irradiation and long standing lymph edema of the extremity following mastectomy have been

associated with development of lymphangiosarcoma (Stewart-Treves syndrome)[3]. The most common presentation of soft-tissue sarcoma is an asymptomatic swelling or lump, maybe associated with an episode of trauma. Rarely, it produces pain. The swelling of the proximal extremities can grow up to 10cm without producing symptoms. Differential diagnosis of soft tissue-sarcoma includes lipoma, sebaceous cyst, neurofibroma, myositis ossificans, squamous cell cancer, melanoma etc.

Evaluation of soft-tissue sarcoma includes evaluation of the primary site as well as of the potential sites for metastases. The optimal modality of imaging is controversial. CT and MRI can be done. Both CT and MRI provide valuable information on the primary tumor in relation to surrounding structures. The Radiology Diagnostic Oncology Group did not find any benefit of MRI over CT[4]. The most common site of metastases for soft-tissue sarcoma of extremities is the lung[5]. Biopsy is the cornerstone of the investigation. Lesions <5cm are best diagnosed by excisonal biopsy. Lesions >5cm are candidates for Tru-Cut or incisional biopsy. Incisional biopsy is more specific and sensitive as compared to Tru-Cut biopsy. The initial method of choice for diagnosis is Tru-Cut, an incisional biopsy is considered in patients with inconclusive report of Tru-Cut biopsy. Frozen section can also be used as method for obtaining diagnosis but is inferior to Tru-Cut and incisional biopsy for histological type and grading of tumor. Fine-needle aspiration is not reliable for diagnosis of soft-tissue sarcoma. The most common histologic variety is malignant fibrous histiocytoma followed by liposarcoma.

Nowaday's mainstay of therapy is surgery. Limb-sparing surgery is advocated if feasible. Amputation is reserved only for patients with vascular, neurologic or bone involvement. The principle of management is complete excision of tumor with negative surgical margin. With availability of multi-modality treatment, the current rate of amputation is <10%6. To optimize local control, surgery (limb-sparing) should be combined with radiotherapy in extremity sarcoma. The result of combination therapy is superior to that of surgery alone. Patients with low risk for local recurrence do not require radiotherapy7. The main cause of death in sarcoma is distant metastases. The role of chemotherapy in soft-tissue sarcoma still needs to be defined. A randomized controlled trial failed to show an improvement in disease-free and overall survival with postoperative chemotherapy[8].

CONCLUSION

Bleeding from soft-tissue sarcoma is a very rare presentation for extremity sarcoma. The most common location for soft-tissue sarcoma is in the extremities, particularly in the lower limb. The most common histological variety is malignant fibrous histiocytoma and the most common site for metastases is the lung. Biopsy is the mainstay of diagnosis and MRI does not offer any advantage over CT scan for local evaluation. Limb sparing surgery is nowaday's gold standard treatment for extremity sarcoma. Combination of radiation with surgery is advisable to optimize local control. The role of chemotherapy still needs to be defined.

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