Case Report

Undifferentiated pleomorphic sarcoma (UPS) or Malignant fibrous histiocytoma (MFH)

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Abstract

Malignant fibrous histiocytoma (MFH), currently classified as undifferentiated pleomorphic sarcoma, is the most frequent soft tissue sarcoma in adulthood, but it is not as common as a primary skin tumour. We report a case of 50-year-old male cultivator with no significant previous medical history who presented with a solitary painless ulcerated tumour on the right knee. Histopathological findings of ulcer showed pleomorphic cells in the dermis, areas of necrosis and mitotic figures and histopathology of lymph nodes showed tumour metastasis. Probably we are reporting this type of case for the first time in Bangladesh.

Key words: Undifferentiated pleomorphic sarcoma; Malignant fibrous histiocytoma

Introduction:

Undifferentiated pleomorphic sarcoma (UPS), previously known as malignant fibrous histiocytoma (MFH), was first described in 1960 by O'Brien and Stout and histological features of UPS were first described by Kempson and Kyriakos[1].UPS usually occurs in older adults, men account for two-thirds of cases and the Caucasian population is more commonly affected. It can occurs anywhere in the body, typically involves the extremities (70-75%) with lower extremities accounting for 59% of cases and less commonly the retroperitoneal spaces, abdominal cavity or other sites such as visceral organs[2]. Soft tissue sarcomas generally manifest as deep lesions, but they may affect the skin and subcutaneous tissue[3]. The most common clinical presentation is large solitary painless soft-tissue mass in the lower extremities, typically 5-10 cm in diameter. The majority of tumours are intramuscular. At gross pathologic examination, the lesion is multi-lobulated with necrosis, degeneration or extensive haemorrhage can be present. Well defined border of the tumour may appear. Microscopically, a malignant cell with variably pleomorphic nuclei are characteristic.[4] UPS is often diagnosed in advanced stages, with a tendency to local recurrence and systemic metastasis. Since tumour thickness and size are identified as major prognostic factors, early recognition becomes crucial to improve prognosis. The mainstays of treatment for UPS are complete surgical excision most often supplemented with adjuvant radiation therapy.

Case Report:

A 50-year-male cultivator presented with a solitary painless ulcerated tumour on the right knee for 2 years. The ulcer was small to larger within the last

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few months, size about 5 cm x 6 cm. The patient's medical history was unremarkable. On examination, the patient was a healthy individual with normal vital signs. The ulcer was localized, deep, firm, non-tender; base was edematous, weepy; the floor was covered by inflammatory exudates, serosanguinous fluid and necrotic tissue; edge was raised, circular, well demarcated but irregular (Figure-1). The colour and temperature of the surrounding skin and rest of the leg were normal. No cyanosis and gangrene was present

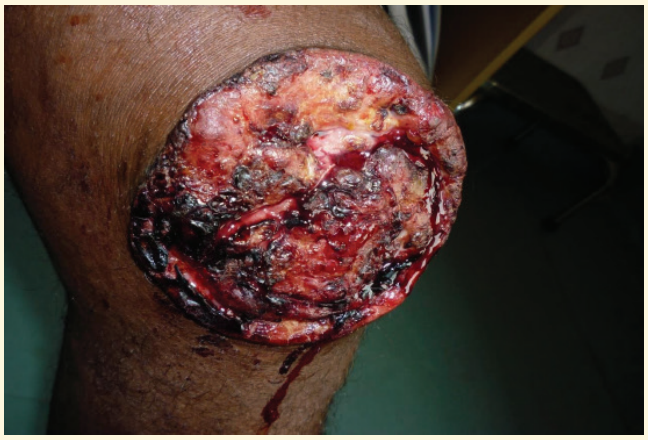

Figure 1: Clinical image of lesion shows solitary ulcerated plaque, size about 5 cm x 6 cm

The lymph node was palpable on the right inguinal region, 3 to 4 in number, isolated but grouped, 1.5 cm x 1.5 cm in size approximately, firm, non-tender, not fixed with the overlying skin and underlying structure. The differential diagnosis included an atypical fibroxanthoma and a pleomorphic dermal sarcoma, leiomyosarcoma, myxofibrosarcoma. Basic investigations including complete blood count with ESR, blood sugar, chest x-ray (P/A view), ultra-sonogram of the whole abdomen, thyroid, renal and liver function tests were all normal. Histopathological examination of the ulcer revealed the dermis and subcutis present a tumour made of spindle-shaped cells. The tumour cells show moderate pleomorphism. Areas of necrosis and mitotic figures are seen. The surgical margin is involved by the tumour (Figure-2, 3)

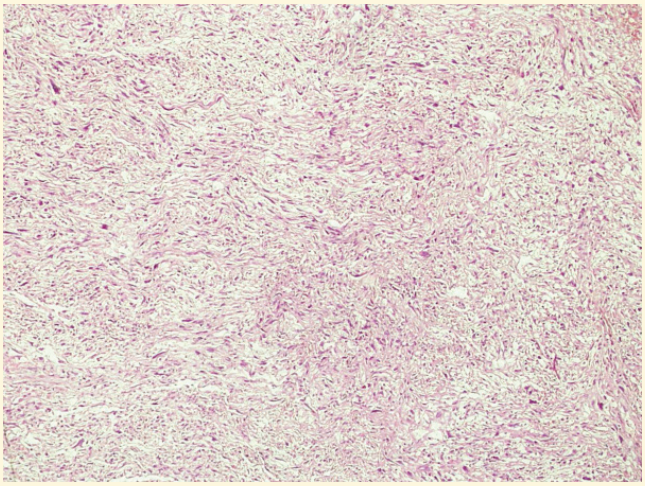

Figure 2: Neoplastic proliferation of spindle-shaped cells (H and E)

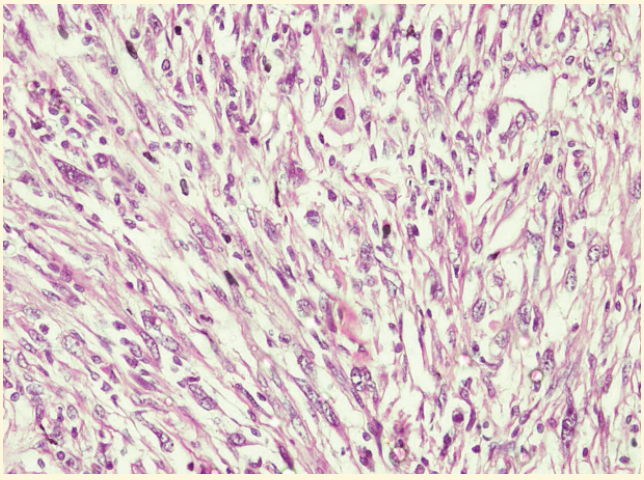

Figure 3: Mitotic figures in malignant cells (H and E)

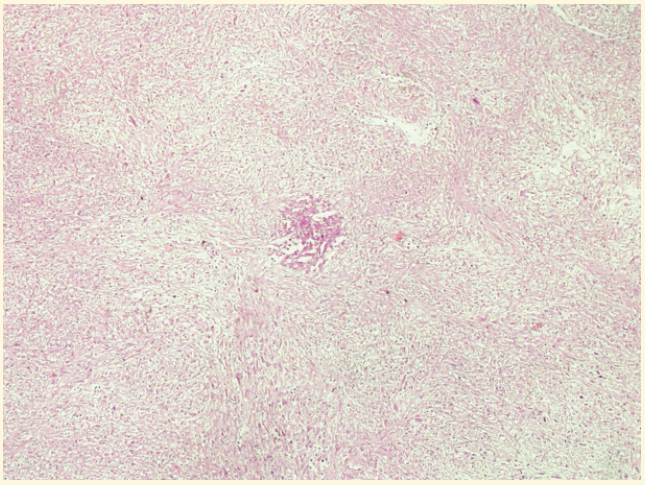

Figure 4: Metastatic tumour in lymph node (H and E)

Histopathology of lymph nodes showed tumour metastasis (Figure-4)

The diagnosis of undifferentiated pleomorphic sarcoma (UPS) was done based on clinical history, lesion morphology and pathologic findings.

Discussions:

The term "Malignant Fibrous Histiocytoma (MFH)" has been changed by the WHO to Undifferentiated Pleomorphic Sarcoma (UPS) not otherwise specified.[5] According to the histology MFH is classified into five types—storiform/pleomorphic, giant cell, inflammatory, myxoid (myxofibrosarcoma) and angiomatoid.[2] In such cases, an alternate name of undifferentiated high-grade pleomorphic sarcoma is being advocated by the WHO in its 2002 classification of soft tissue tumours. The diagnosis of storiform-pleomorphic MFH is one of exclusion when no line of differentiation is identified. Pleomorphic is the most commonly seen histological variant of tumour and our case also belonged to this subtype.[5] However, undifferentiated pleomorphic sarcoma (UPS) is a rare entity, which is a member of spindle cell neoplasms arising from the skin or soft tissue and can show variable clinical courses, despite being histopathologically similar.[6] Dermal UPS is called by different names in the literature, such as atypical fibroxanthoma (AFX), superficial malignant fibrous histiocytoma (MFH), superficial UPS or pleomorphic dermal sarcoma (PDS).[7-9] However, undifferentiated pleomorphic sarcoma not only encompasses skin neoplasms but also a heterogeneous group of soft tissue malignant neoplasms of the internal organ, retroperitoneal, and osteoid origins.[10-11] The mutation of telomerase reverse transcriptase (TERT) is present in 76% of the skin UPS cases and appears to be associated with UV damage.[7] Thus, the increased incidence in the sun-exposed or UV-damaged skin areas such as head and neck can be explained by TERT mutation.[13] On the other hand, the presence of a rapidly growing aggressive lesion on the non-UV-exposed skin areas, particularly in elderly patients, it is recommended to rule out other possible malignancies before diagnose UPS due to its rarity.[13] USP may also arise at sites of chronic ulceration.[4] USP usually affects the proximal limbs, although it has been described in the cephalic location. It usually presents as a slow-growing tumourous lesion, though no clinical characteristic allows differentiation from other sarcomas. Tumour size and depth have been identified as the main prognostic factors. Therefore, establishing an accurate and early diagnosis is crucial in improving the tumour prognosis.[1] In the present report, the case was a painless deep seated ulcerated plaque on the right knee with a slow-growing tumourous lesion with no significant previous medical history. The diagnosis of USP is clinically difficult to identify, especially in cases where the tumour is deep and is usually diagnosed after the removal[10] . The definitive diagnosis of UPS relies on histological studies and showed malignant cells with nuclei pleomorphism and atypical mitotic figures. There is no characteristic immunohistochemical profile. Immunohistochemical analysis is used to help xclude other tumours that may have a pleomorphic appearance.[4] In our case report, the diagnosis of UPS was confirmed by a combination of clinical history, clinical examination and histologically.

UPS treatment options vary based on factors such as: (i) whether the UPS is localized or metastatic, (ii) location of the tumour, (iii) patient's age and general health. In general, treatment options for UPS may involve surgery and chemotherapy. Treatment of localized UPS may include chemotherapy followed by surgery to remove the tumour and surrounding tissue. Prognosis of Undifferentiated pleomorphic Sarcoma (USP) is influenced by the depth of tumour infiltration into the surrounding tissue, the mass of the tumour and anatomic location of the tumours.

Conclusions:

UPS is a rare entity with relatively difficult to diagnose. So, the final diagnosis should be made based on the correlation of the clinical history, lesion morphology, and pathologic findings.

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