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Undifferentiated pleomorphic sarcomas of the proximal femur, A case study

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Abstract

Pleomorphic sarcomas are a subtype of soft tissue sarcomas occurring most commonly in the proximal thigh, with a male preponderance, common in 7th and 8th decades of life. It is a locally invasive tumour with a wide clinical presentation.

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Introduction

Soft tissue sarcomas are mesenchymal tumors, which arise from Connective tissue, which includes muscle, fat, blood vessels, deep skin tissues, nerves, bone, and cartilage. Undifferentiated Pleomorphic Sarcoma (UPS),which was previously called Malignant Fibrous Histiocytoma (MFH), is a subtype of a Soft Tissue Sarcoma (STS) that can occur anywhere in the body, but it usually occurs in the extremities (especially the lower extremities) or retroperitonealy. Undifferentiated pleomorphic sarcoma usually occurs in older adults, and often men account for two thirds of cases. Initially the nature of the tumor is benign, but soon becomes locally malignant leading to wide variety in its presentation, and thus a form of a diagnostic challenge. Although, no risk actors and causative agents are identified, it typically occurs on sites of prior radiation therapy, chronic sinuses, and exposure to certain chemicals but more often than not, it occurs in individuals without any prior risk factors [1].

Though the grade, and level of local invasion, metastasis, and post surgical margins guide the tumor management, treatment modality is basically subdivided into surgery chemotherapy and radiation therapy. Risk of recurrence is present as with any tumor, so regular follow up, and evaluations are recommended

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Case Report

This is a case report of a 49 year old male, who presented initially, with acute left thigh pain, for 3weeks, which was progressive. The pain as he described would often radiate around the left anterior thigh down to the knee and occasionally down the medial aspect of the lower leg and to the arch of the foot. A heavy construction worker by occupation, he initially attributed these symptoms to his posture, as the pain became progressive in the time course of 5 weeks, he decided to seek medical attention.

On a complete physical examination, there was weakness of hip flexion, and hip adduction. Loss of sensation over the anterior thigh and the medial aspect of the lower leg was noticed. A reduced Left knee Deep tendon reflex was observed. The patient was initially started on Folate, Vtamin B12, Vit D, 15000units per week and daily calcium tablets, along with regular physical therapy. Fifteen days later he returned to seek medical care after he had a fall, which led to "excruciating pain in his Proximal Left hip." Immediate X-rays taken showed us that he had a proximal femoral shaft fracture, along with a diaphyseal lesion. He had undergone internal fixation with medullary rods, and during this surgery, biopsy was taken from the fracture site, which showed, an unspecified histological grade 3 pleomorphic adenoma, with 40% necrosis, and areas of hemorrhage.

A Lower Extremity MRI With and Without Contrast, showed a mass originating from the distal femur which extends from 18.6cm in craniocaudal dimension along the medullary cavity and measures 3 x 4.2 cm where is breaks through the distal diaphyseal cortex extending into the soft tissues. The mass demonstrates low signal on T1-weighted images and heterogeneous but predominantly increased T2 signal/fluid signal on fluid sensitive sequences. No clear fat plane is identified between the soft tissue mass and the superficial femoral artery and vein. The MRI also showed that the mass abuts the sciatic nerve just proximal to the takeoff of the common peroneal nerve, without any clear fat between the soft tissue mass and the sciatic nerve. The central portion of the expansile soft tissue mass component demonstrated nonenhancement suggesting a necrotic center while the peripheral of the mass demonstrates irregular enhancement

A CT Angiography of the left Lower Extremity With IV Contrast showed an angulated and medially displaced distal femoral pathological fracture secondary to heterogeneous soft tissue mass originating from the distal femur with necrosis and demonstrating cortical break through and soft tissue extension, and more importantly a Mass effect on the posterior neurovascular structure with possible tumor involvement, with no lymphnodal metastasis on PET scabbing.

The patient underwent, a tumour extraction, a wide circumfrential excision, along with revision surgery of his early pathological fracture. The patient was immediately started on neoadjuvent chemotherapy consisting of Adriamycin and Cisplatin, but on testing his surgical margins, the medial margin was positive by 0.5mm, so he has been started on and local irradiation the patient as of now is responding well, no new masses .on surveillance MRIs, CT and PET scan, except for feelings of malaise, and falling WBC profile, for which Filigastrim was initiated. He will be monitored, with regular irradiation and continuing cycles of chemotherapy.

Discussion

The incidence of soft tissue sarcomas is relatively low at 2 to 3 per 100 000 per year, and for undifferentiated pleomorphic sarcoma in particular, which the 4th most common soft tissue sarcoma, the incidence is 1 per 100 000 per year. The most common sites of an undifferentiated pleomorphic sarcoma include proximal femur, proximal humerus and the retroperitoneum, with a high preponderance for appearance of this tumour in latter life, especially in the 7th and 8th decades of life [2].

Quite opposite of what one thinks of sarcoma, which is a group of masses with a uniform presentation, the term sarcoma is coined for tumours, with histological distinct subgroups, arising from a group of totipotent cells. The most frequent sarcomas in adults are liposarcoma, fibrosarcoma, and pleomorphic sarcoma (previously known as malignant fibrous histiocytoma [3]. The most commonly encountered sarcomas inclue liposarcoma, fibrosarcoma and pleomorphc sarcoma.

Though mostly slow growing, problems with these types of soft tissue mesenchymal tumours arise when they either become too large to cause discomfort to the patient or they turn malignant, which is a risk with any unchecked growth in a group of cells. The most common presentation is when patients "discover" the mass after a trivial trauma.

Clinical features of a long bone tumour like a pleomorphic sarcoma, has wide range of presentation, and the symptoms don't show up until the late disease process, ranging from deep aching, gnawing bone pain, to involvement of the nerves, most commonly on the posterolateral surface resulting in numbness and tingling in the posterior aspect of the thigh, calf and foot, or "sciatica", to pathological fractures, which often lead to a quick discovery of the tumour.

Once a diagnosis of an undifferentiated pleomorphic sarcoma is made, hereditary syndromes such as neurofibromatosis, Gardner syndrome, Li-Fraumeni syndrome, and retinoblastoma, should be ruled out, as these conditions include a myriad of tumours [4].

Diagnosis is typically made using, T1 and T2 weighted MRIs, which is the gold standard, and an ultrasound core biopsy to identify cell populations, percentage of atypical cells irregular mitotic figures and regions of haemorrhage and necrosis. Immunohistochemcal staining is also required, for stains such as HMB 45, S-100, cytokeratin, to guide treatment options and to assess prognosis.

The treatment modalities are three fold, as in all cases of a bone malignancies, consist of excision, with wide margins the rest of treatment including chemotherapy and radiation therapy depends on the grade of the tumour, local invasion and metastasis. Lymph node involvement must be carefully assessed, not only for dissection but also to make sure no spread has occurred. Recurrence occurs in about half of the patients, common within the first 2 years of treatment. Increased risk of recurrence includes factors such -a mass size greater than 5 cm, a high-grade histology, a deep anatomical location of the tumour, poor resection of the original tumour [4].

Like in our case, the occurrence of pleomorphic sarcoma, albeit in the most common area, but in a comparatively younger, middle age group, without any predisposing factors and broad clinical presentation makes it a formidable challenge, both in terms of a timely diagnosis and treatment.

conclusion

Undifferentiated pleomorphic sarcomas of the lower extremity, commonly occur in 7th or 8th decade, with a male predominance. Due to its initial slow growing nature, extensive local invasions, a wide variety of symptoms are common, particularly involving the muscle, superficial sift tissue, and the neurovascular bundle, hence a timely diagnosis, is often challenging. The gold standard for diagnosing a pleomorphic sarcoma is an MRI. Even after appropriate treatment, the risk of recurrence is high, especially within a period of 2 years; therefore, regular follow-ups with imaging such as an MRI are mandatory.

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