ONLINE FIRST

CASE REPORT

A Giant Well-differentiated Liposarcoma: A Case Report

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ABSTRACT
We report a rare case of an enormous well-differentiated liposarcoma (WDLPS) in a patient aged <40 years. A 39 years old Saudi male patient presented with a swelling in the upper, medial aspect of the right thigh. It was a large swelling, about 15 x 15 cm. All blood investigations were unremarkable. CT scan revealed a huge (20.1 x 20 cm) well-defined lobulated heterogeneous, hypodense subcutaneous lesion on the medial aspect of the right upper thigh, extending superiorly to the subcutaneous area of the ipsilateral natal cleft and inferiorly down to the mid-thigh level. The lesion showed multiple internal septations with a solitary spot of tiny fatty component. The entire mass was excised en bloc with its capsule. It was huge, but encapsulated. Histopathology reported a multinodular mass with infiltrative margins, measuring approximately 24 x 20 x 13 cm in greatest dimensions. Cut surface was lobulated, gelatinous, and myxoid, with grey-white, firm nodules and focal areas of haemorrhage and necrosis. Microscopic sections of the mass revealed a malignant lipomatous tumour comprising mature adipocytes, atypical spindle cells, and multivacuolated lipoblasts, embedded in a loose myxoid stroma with focal dense fibrous stroma. Few scattered mitotic figures were seen. No infiltration of the overlying skin (about 2 cm margin) was seen, and no invasion of all other margins (< 1 cm) was found. No recurrence was seen in follow-up. Documentation of LS incidence among Saudis is scarce, but it is rising in the USA and Europe. With genetic predisposition in the White population, radiation or herbicides might be responsible for this increase.

KEYWORDS: Liposarcomas, LS, Well-differentiated liposarcoma, WDLPS, atypical lipomatous tumor, ALT, Saudi Arabia

INTRODUCTION

Liposarcomas (LS) are cancers derived from adipocytes or their precursor lipoblasts in soft tissues. They constitute 20% of soft tissue neoplasia[1]. Liposarcomas are rare mesenchymal sarcomas[2]. The types of liposarcomas described by WHO are atypical lipomatous tumor (ALT)/well-differentiated liposarcoma (WDLPS), dedifferentiated liposarcoma, myxoid/round cell liposarcoma (MRCLPS), and pleomorphic liposarcoma[1]. ALT/WDLPS can grow to an enormous size in limbs but seldom metastasize.

Limited data exist on atypical lipomatous tumor (ALT)/well-differentiated liposarcoma (WDL) in adults <40 years of age, as it is rare in this age group[4]. We report a case of ALT/WDLPS in a patient aged <40 years that grew to an enormous size and was managed and operated on at our institution.

CASE DESCRIPTION

A 39 years old Saudi male patient presented in the outpatient department, on 28 /12/2024, with a swelling in the upper, medial aspect of the right thigh that appeared about one and a half years ago. It slowly kept increasing in size. The patient did not seek any medical consultation for it. It was not associated with any pain or any other complaint. He had no underlying medical problem or any previous history of surgery.

On examination, a large, 15x15 cm swelling was noted on the upper medial aspect of the right thigh, with a broad base. The skin over it was intact, except for a very small area, less than 1 cm in diameter, with some skin excoriation. It was non-tender. There were no dilated veins over it. It was close to the right scrotum, but the scrotum was free and separate from the swelling. The skin over it was not involved. It was soft, with some cystic areas, was mobile over the deeper structures/muscles, and extended posteriorly towards the perineum and anal canal. The inguinal lymph nodes were not enlarged and not palpable. It was close to the anal canal, but anal and rectal examination was normal.

All blood investigations were unremarkable. Contrast-Enhanced Computed Tomography (CECT) scan revealed a huge (20.1 x 20 cm) well-defined lobulated heterogeneous, hypodense subcutaneous lesion on the medial aspect of the right upper thigh, extending superiorly to the subcutaneous area of the ipsilateral natal cleft and inferiorly down to the mid-thigh level. The lesion showed multiple internal septations with a solitary spot of tiny fatty component. There were no calcifications. The lesion caused displacement of adjacent thigh muscles, without definite invasion. The overlying subcutaneous tissue was intact. Adjacent bones were normal. There was no significant regional lymphadenopathy. CT scan of the chest, abdomen and pelvis was normal. The lungs, abdominal organs, para-aortic area, mesenteric lymph nodes, and urinary bladder were normal. A large saccular lesion, containing fluid in the medial part of the right thigh, appeared to be extending cranially.

On 30/12/2024, under spinal anesthesia, an elliptical incision was made over the mass, near its base, including ulcerated skin. It was to be excised, ensuring that the mass could be excised in toto without compromising the safety margin, and the skin could be closed later free of tension. The entire mass was excised en bloc with its capsule. It was huge, but encapsulated, extending deep between the muscles of the medial compartment of the thigh, away from major nerves and vessels, encroaching over the perineal muscles and the anal sphincter, but not involving them. Hemostasis was secured, and the wound was closed with a drain after closing the dead space with Vicryl 3/0 sutures.

The mass was sent for histopathology. It was reported as a multinodular mass with infiltrative margins, measuring approximately 24 x 20 x 13 cm in greatest dimensions. Cut surface was lobulated, gelatinous, and myxoid, with grey-white, firm nodules and focal areas of haemorrhage and necrosis. The overlying skin ellipse measured 17 x 12 cm and was completely separate from the mass. Microscopic sections of the mass revealed a malignant lipomatous tumour comprising mature adipocytes, atypical spindle cells, and multivacuolated lipoblasts, embedded in a loose myxoid stroma with focal dense fibrous stroma. Few scattered mitotic figures were seen. No infiltration of the overlying skin (about 2 cm margin) was seen, and no invasion of all other margins (< 1 cm) was found. The diagnosis was well-differentiated liposarcoma.

The patient was referred to medical oncology for further management. He was followed up in the surgical OPD for four months. The wound initially had some serous discharge. A few clips were removed to drain the serous discharge, and the wound was treated conservatively. After drainage of the initial serous discharge, it healed well. Surgical clips were removed after 2 weeks. All investigations were repeated, including CBC, biochemistry, and LFTs, and the results were normal. He was reinvestigated by the oncologist with a total-body scan, local MRI, and PET scan, and was found to be disease-free.

Seven months later, the patient had an appointment in the King Faisal Specialist Hospital in Riyadh, Oncology Department, where they declared him disease-free.

DISCUSSION

Liposarcomas (LS) are rare tumors. They are malignant mesenchymal sarcomas originating from soft tissues, and are believed to arise from cells in the lipocyte lineage[2]. They constitute about 13-20% of all soft-tissue sarcomas and are the most common soft-tissue sarcomas globally[2]. They mostly originate in the lower limbs, but rarely in the retroperitoneum, abdominal wall, or subcutaneous tissues[1]. In most cases, they appear in the lower limbs, particularly in the thigh, and usually measure> 11 cm [8,9]. Our patient also presented with this sarcoma in the thigh with similar dimensions.

In the USA, the incidence of LS is increasing, as data from 2001-2016 showed a 19% increase in LS diagnoses. Their incidence in Europe is four to five per 100,000 per year[2]. However, there is a gap in the epidemiological recognition of soft tissue sarcomas in Saudi Arabia. Although they are relatively uncommon as compared to other cancers, the documentation of the incidence and characteristics of these sarcomas in the Saudi population is deficient[3v]

In literature, the average age at diagnosis of LS was found to be 50 years of age[2,5-9]. The retroperitoneal liposarcomas usually present at a median age of 56 years; however, they can occur at any age[10]. Our patient, however, was quite young, i.e., 39 years old.

According to the literature, men account for about 60% of cases[2,5,7,8]. The gender distribution of retroperitoneal sarcomas is approximately equal, although some studies have reported female predominance[10]. In our case, the patient was a male.

In more than 80% of cases, the patients are Caucasians, less than 10% are African Americans, and the remaining belong to other races[2,7]. Our patient was a Saudi Arab national. One study assessed the incidence of soft-tissue sarcomas in Saudi Arabiax[11]. The data were extracted from the Saudi Cancer Registry. This study included all the patients with primary soft-tissue sarcomas of the limbs and pelvis between January 2006 and December 2015. Out of these 659 patients, 20.5% had liposarcomas. Patients with liposarcomas had the longest mean survival, i.e., 7.74 years. There are no widely accepted risk factors for liposarcomas[2].

They are almost always deep-seated[8]. The tumor of our patient also extended deeper into the thigh and towards the perineum. It was huge and extended deep between the muscles of the medial compartment of the thigh; it was away from major nerves and vessels. It encroached on the perineal muscles and the anal sphincter, but did not involve them.

Treatment of WDLPS is mostly restricted to surgical resection along with negative surgical margins[12]. They mostly do not require radiotherapy or chemotherapy. Our patient's entire tumor mass was excised en bloc with its capsule. No radiotherapy or chemotherapy was required.

On histopathological examination, these liposarcomas are usually composed of lobules of mature adipocytes, variable in size and divided into compartments by thick, fibrous bands[2,8]. The histopathology report of our patient showed mature adipocytes, atypical spindle cells, and multivacuolated lipoblasts embedded in a loose myxoid stroma, along with focal dense fibrous stroma, typical of WDLPS. No local or distant metastasis was found in our patient, which is typical of this type of LS[7,12].

There are no widely accepted risk factors for liposarcomasx[2]. Because the etiology and risk factors are unknown, the alarming increase in liposarcoma incidence is concerning. An increase in exposure to radiation or herbicides can be a possible explanation[13]. As a greater risk has been found among whites and males, there might be some genetic predisposition to liposarcoma.

CONCLUSION

The documentation of the incidence of LS in the Saudi population is deficient. However, their incidence has been found to rise in the USA and Europe. Along with genetic predisposition in the White population, radiation or herbicides might be responsible for this rise.

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AUTHOR CONTRIBUTION

Hashmi WA: Concept, revision
Hashmi A: Design, drafting
Hashmi AW: Concept, revision
Malik MK: Design, drafting

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