Pleomorphic Liposarcoma of the Scapula in a Young Adult Female: A Rare Entity

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ABSTRACT

Pleomorphic liposarcoma of the scapula is one of the rarest entities occurring in a young adult female. We present a case of a 28-year-old female who presented with a large growing mass on the right scapula. Computed tomography and magnetic resonance imaging showed local invasion by the mass. Wide excision followed by adjuvant radiotherapy was done to treat the tumor successfully. A thorough histological analysis of all soft tissue masses should be done along with prompt diagnosis and intervention which can change the prognosis of this rare disease.

Keywords: Liposarcoma; Scapula; Pleomorphic; Magnetic Resonance Imaging; Radiotherapy

INTRODUCTION

Liposarcomas (LS) are malignant tumors of fat cells with distinct subtypes according to clinicopathologic and cytogenetic characteristics. Pleomorphic liposarcoma (PLS) is the rarest and most aggressive subtype and is characterized by the presence of pleomorphic lipoblasts [1]. It is usually present in lower and upper limbs and scapular presentation is extremely rare with only few cases reported so far in the literature [2]. We present a case of pleomorphic liposarcoma of the scapula in a young adult female in her late 20s, who was successfully treated by surgery and adjuvant radiotherapy.

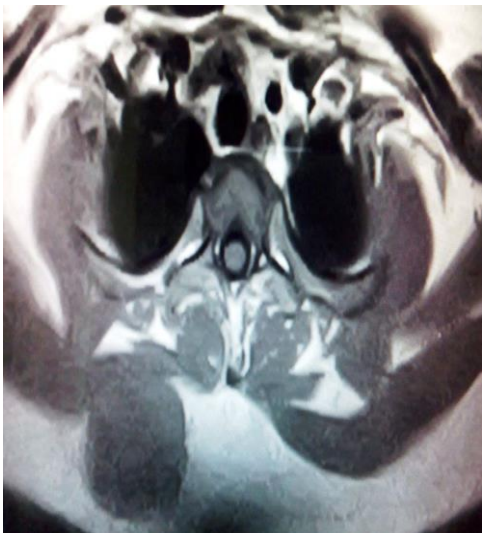
CASE REPORT

A 28-year-old female presented with a painless mass of approximately 10 x 5 cm on the right scapula. The mass was very small in size at the time of onset and had gradually increased in size for last 2 months before admission to our hospital. Examination revealed a firm, painless, palpable mass of approximately 10 cm in size with poorly-defined margins and adhesion to the underlying structures and skin. Overlying skin appeared normal in color. Laboratory findings including blood count, kidney function tests and liver function tests were in the normal range. Chest radiograph was normal and excluded any lung metastasis. Computed tomography and magnetic resonance imaging were done to rule out the possibility of lipoma but these studies did not show fat signal and showed loss of fat plane with underlying muscle (Figure 1). Fine needle aspiration cytology was done which showed presence of occasional lipoblasts. Wide excision of tumor was performed (Figures 2A, B) (Figure 3). Histopathological examination of the tumor revealed it to be 1 cm away from the overlying skin and microscopically, the tumor was situated in deep soft tissues with mitosis of approximately 40-45/10 HPF, necrosis, pleomorphism and high grade sarcoma. On immunohistochemistry, tumor cells were negative for CD34, desmin, caldesmon and S100. Tumor showed focal and weak positivity for SMA. Postoperative period was uneventful and patient was given adjuvant radiotherapy to avoid recurrence and the patient is doing well in the follow-up period after 9 months.

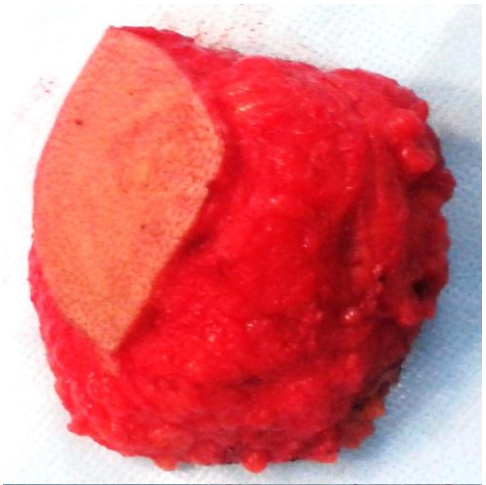
DISCUSSION

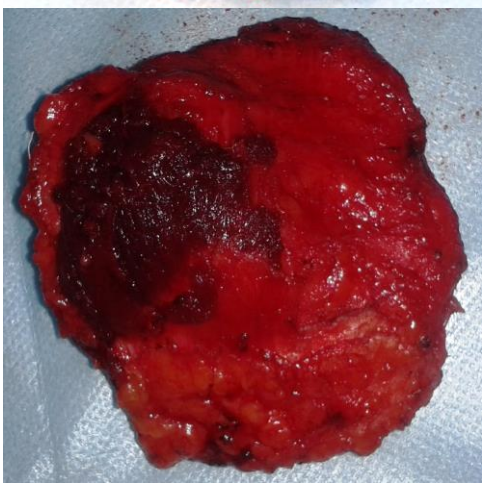
Liposarcoma (LS) is a malignant disorder of fat cells and accounts for up to 18% of all soft tissue sarcomas. It usually affects adults of 40 to 60 years age and can occur in any part of the body but most of them involve the thigh and abdominal cavity. However, in our case, it is present in scapula and in a female of 20 year age group [3]. There are four subtypes of liposarcoma, well-differentiated, myxoid, pleomorphic and dedifferentiated, each of these has its own unique characteristics and behaviors.

Figure 1: MRI showing tumor invasion into deep muscles



Figures 2A, B: Anterior and posterior views of the excised tumor specimen showing muscle invasion





Pleomorphic liposarcoma (PLS) is the rarest and also the most aggressive among all these types with cells that look very different from normal cells. It is so uncommon that there is limited data in the literature regarding its clinical and pathologic spectrum.

The etiology of this rarest entity is unknown; however, it is characterized by highly complex chromosomal alterations and usually presents as a painless, deep-seated mass. Histological recognition of PLS requires careful examination for the presence of pleomorphic lipoblasts and immunohistochemistry usually has no significant role in its diagnosis. However, S-100 protein immunoreactivity is seen in up to 48% of lipogenic areas and SMA in 49% of non-lipogenic areas of PLS [4, 5].

Being a high-grade sarcoma, PLS has high rate of recurrence and metastasizes especially to lungs. The treatment of PLS requires wide and deep surgical excision along with local radiation [6]. However, it has poor prognosis with a 5-year survival rate of about 50% as they usually recur and metastasize [7].

The differential diagnosis of PLS includes pleomorphic leiomyosarcoma, pleomorphic rhabdomyosarcoma, pleomorphic malignant peripheral nerve sheath tumor, and malignant fibrous histiocytomas [8]. It is important to bear in mind that the recognition of pleomorphic lipoblasts is most important for identification of PLS whereas immunohistochemistry is more helpful in excluding other types of pleomorphic sarcomas than confirming a diagnosis of PLS as already discussed.

CONCLUSION

PLS is a rare high-grade sarcoma of adulthood that occurs most commonly in the deep soft tissues. It frequently metastasizes to the lungs with poor prognosis and histologically, the presence of pleomorphic lipoblasts remain the mainstay in clinching the diagnosis.

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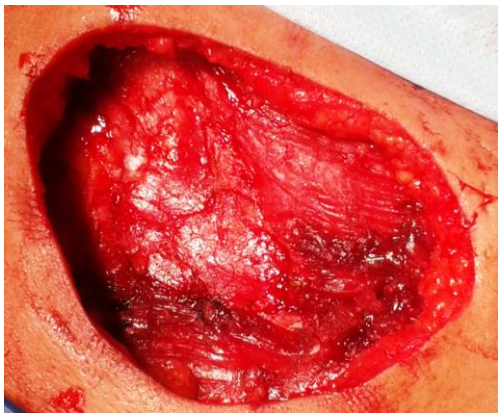
Figure 3: Area left after wide excision of the tumor



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