ABSTRACT

Liposarcoma is the most common soft tissue sarcoma accounting for 20% of all mesenchymal malignancies. Spindle cell liposarcoma is a rare histological variant of liposarcoma. Spindle Cell Liposarcoma presenting with ulceration is a rare entity in the history of literature. We report a case of spindle cell liposarcoma arising from the right forearm presenting with ulceration. A 14-year-old young boy presented with a slow growing ulcer over the right forearm. Histological examination of biopsy showed spindled and stellate shaped cells in a myxoid background and many curved vessels suggesting myxoid and round cell liposarcoma suggesting spindle cell liposarcoma. Based on this, a histological diagnosis of spindle cell liposarcoma was made. Spindle cell liposarcoma is a rare variant of well-differentiated liposarcoma characterized by prominent spindle cell component. In previously reported cases spindle cell liposarcoma was demonstrated in the subcutaneous tissues of limbs, trunk, shoulder girdle, buttock presenting a mass without ulceration. Main differential diagnoses include benign lesions such as spindle cell lipoma, diffuse neurofibroma as well as dermatofibrosarcoma protuberans and other malignancies such as sclerosing liposarcoma, myxofibrosarcoma, malignant peripheral nerve sheath tumor and fibromyxoid sarcoma. Spindle cell Liposarcomas tend to recur locally and may have a potential for metastasis. Wide excision and long term follow up looking for recurrence and metastasis is necessary. To our knowledge, Liposarcoma presenting with ulceration at forearm has not been observed in literature. So, our case is a rare variant of liposarcoma arising at forearm with ulceration.

Keywords: Spindle cell Liposarcoma; leg; atypical Lipomatous Tumor; well differentiated Liposarcomas; Spindle cell

INTRODUCTION

Sarcomas encountered in limbs show wide variety of histo-morphological types and grades. Liposarcoma of the leg represents approximately 1% of sarcoma of limbs. They are a group of malignant neoplasms that affect critical structural units of legs that can result in grave consequences if they are not diagnosed and managed properly. Amongst the group of Well Differentiated Liposarcomas/ Atypical Lipomatous Tumor (WDL/ALT), spindle cell variant (S-WDL/ALT) tend to develop in the deep muscles of extremities (75%), retroperitonaeum (20%) and other

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In this case, the tumor was located in right forearm with ulceration as an unusual condition. This tumor is composed of prominent spindle cell proliferation and varying sized adipocytes and mixed with few lipoblasts set in a fibrous and/or myxoid background. It tends to occur in adults. Here, we report a case of S-WDL/ALT in the right forearm in a 14 year young boy. These tumors do not metastasize hence require less aggressive therapeutic management. Accurate histopathological subtyping is absolutely essential as it impacts treatment strategies and outcome.

CASE HISTORY

A 14 year young male presented with a progressively increasing ulcerated growth over the right forearm. This was noticed three months prior to presentation, which was small initially and gradually, increased to the present size. In this case there was neither any previous history of lipomas nor any remarkable associated family history.

FINDINGS

Local examination showed 8 x 7 cm, non-tender, well-defined, ulcer over the upper part of the right forearm (Figure 1).

Findings of systemic examination were non-contributory. Routine hematological examination was normal except low hemoglobin and high ESR. Routine biochemistry investigations, viz., Random Blood Sugar, Serum Creatinine, Liver and Kidney function test showed no abnormality. X-ray of chest and right forearm were normal. Ultrasonography of abdomen neither showed hepatosplenomegaly nor adenopathy. Routine examination of urine was normal. On microscopic examination, circumscribed tumor composed of spindle cells arranged in short interlacing fascicles interspersed with single and at places collections of varying sized groups of lipogenic cells were seen (Figure 2 and 3).

Figure 1 Spindle-cell Liposarcoma Presenting with Ulceration over Right Forearm

Figure 2 HPE Shows spindled and stellate shaped cells in a myxoid background and many curved vessels suggesting myxoid and round cell liposarcoma (40X)

Figure 3 HPE demonstrate cells with uniform round nuclei in a myxoid background (40X)

Stroma showed few atypical spindle cells; thin walled blood vessels, thick and thin fibrous septae with focal myxoid areas. The spindle cells revealed bland oval nuclei and moderate amount of eosinophilic cytoplasm. Few large atypical cells were noted. No areas of necrosis and
hemorrhage noted. On careful search, occasional lipoblasts were seen in the sections taken from the periphery of the tumor. Based on histopathological appearance, spindle cell liposarcoma tumor was considered.

**DISCUSSION**

Histologically, Liposarcomas are divided into five different subtypes: myxoid, pleomorphic, dedifferentiated, round cell and Atypical Lypomatous Tumors (ALT)/Well differentiated Lyposarcomas (WDL). ALT/WDL is regarded as low grade, non-metastasizing, malignant neoplasms composed primarily of mature adipose tissue. Statistically, approximately 75% develop in the deep soft tissue of the limbs, followed by 20% in the retroperitoneum and a much smaller percentage in the inguinal region. Furthermore, ALT/WDL is further subdivided into the adipocytic (lipoma-like), sclerosing, inflammatory and spindle cell subtypes. Spindle cell- ALT (S-ALT)/WDL is a rare variant of ALT/WDL. It is a distinct neoplasm which tends to occur in the subcutis of the shoulder region and extremities without presenting with ulceration. Few cases have been described in the head and neck. Histologically, this biphasic tumour is composed of prominent spindle cell proliferation and varying sized adipocytes admixed with few lipoblasts set in a fibrous and/or myxoid background. Lipoblasts show central or peripheral hyperchromatic nucleus which is indented by univacuolated or multivacuolated cytoplasm. As described in the literature, Cytological findings include mixture of adipocytes supported by fibro-vascular septa with hyper chromatic and enlarged nuclei within the fat and fibrous bands with one or two small nucleoli and scattered lipoblasts. Distinction of lipoma and well-differentiated liposarcoma can also be made using imaging techniques. Ultimately histopathology along with immunohistochemistry studies clinches the diagnosis. A few cases of atypical lipomatous tumor/well-differentiated liposarcoma of the gingival has also been recorded in few cases of atypical lipomatous tumor/well-differentiated liposarcoma of the gingival has also been recorded in few cases of atypical lipomatous tumor/well-differentiated liposarcoma of the gingival has also been recorded in few cases of atypical lipomatous tumor/well-differentiated liposarcoma of the gingival has also been recorded in few cases of atypical lipomatous tumor/well-differentiated liposarcoma of the gingival. The differential diagnosis of S-ALT/WDL is characterized by giant marker and ring chromosomes containing amplified sequences of 12q13-15 which is the site for several genes including MDM2, CDK4, GL1, SAS and HMGIC. However, WDSCL lacks the MDM2 and CDK4 gene amplifications and instead show monosomy of chromosome. Another study depicted that unlike epithelial neoplasms, the malignant transformation of a pre-existing mesenchymal tumor has been questioned for a long time. Based on the similarities of clinical, histologic, immunohistochemical, and molecular findings in spindle cell lipoma and well-differentiated spindle cell liposarcoma, it can be speculated that well-differentiated spindle cell liposarcoma represents the atypical/low-grade counterpart of spindle cell lipoma, that the Rb-1 deletion represents an early event in the development of both neoplasms, and that additional genetic changes are necessary for the development of well-differentiated spindle cell liposarcoma. Another hypothesis is the transformation of a pre-existing spindle cell lipoma to a well-differentiated spindle cell liposarcoma, and in some cases of well-differentiated spindle cell liposarcoma, a recent enlargement of a long-standing neoplasm has been reported. A few studies have demonstrated that in striking contrast to epithelial neoplasms, a malignant transformation of a pre-existing benign mesenchymal neoplasm has been questioned for a long time with the exception of the transformation of a neurofibroma to a malignant peripheral nerve sheath tumor in the setting of a neurofibromatosis. However, it has been nicely demonstrated that a biologic continuum of benign, atypical, and malignant lipogenic neoplasms exists, and probably some cases of well-differentiated spindle cell liposarcoma arise in a long-standing spindle cell lipoma similarly to cases of malignant peripheral nerve sheath tumors arising in pre-existing neurofibromas; however, this hypothesis has to be substantiated in further studies. Collagenous stroma is prominent. Myxoid areas and prominent, thick walled, arborizing blood vessels may be found in the stroma. Lipoblasts are absent. Myxoid areas can pose diagnostic problems. However, myxoid liposarcoma shows chicken wire vascular pattern which was absent in our case. Instead our case showed singly scattered large atypical nuclei consistent with spindle cell ALT/WDL. In striking contrast to epithelial neoplasms,
a malignant transformation of a pre-existing benign mesenchymal neoplasm has been questioned for a long time. However, it has been shown that a biologic continuum of benign, atypical, and malignant lipogenic neoplasms exists and probably some cases of S-ALT/WDL arise in a long-standing SCL. However, this hypothesis has to be substantiated in further studies. Previous literature shows that liposarcoma is common in elderly male. Our case is a 14 years old male. Spindle cell liposarcoma is a rare variant of liposarcoma. To our knowledge, Sindle Cell Liposarcoma presenting with ulceration over forearm has not been encountered in previous literature.

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Conflict of Interest: None

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