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CASE REPORT

Spindle Cell Liposarcoma: A Rare Variant of Liposarcoma Arising in Forearm with Ulceration

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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 sclerosingliposarcoma, 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) is rarely documented in literature.[1] WDL/ALT tend to develop in the deep muscles of extremities (75%), retroperitoneum (20%) and other

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miscellaneous sites.[2] 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.[1] 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 lipomasn or 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**).

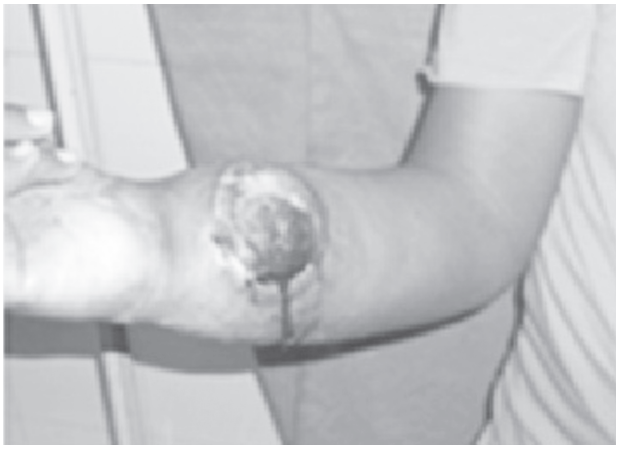

Figure 1 Spindle-cell Liposarcoma Presenting with Ulceration over Right Forearm

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**).

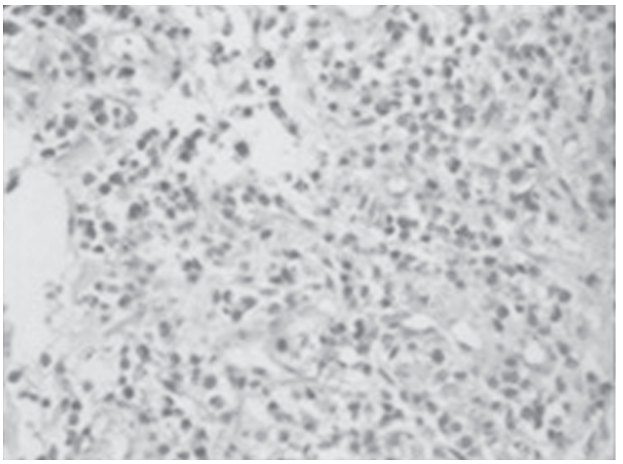

Figure 2 HPE Shows spindled and stellate shaped cells in a myxoid background and many curved vessels suggesting myxoid and round cell liposarcoma (40X)

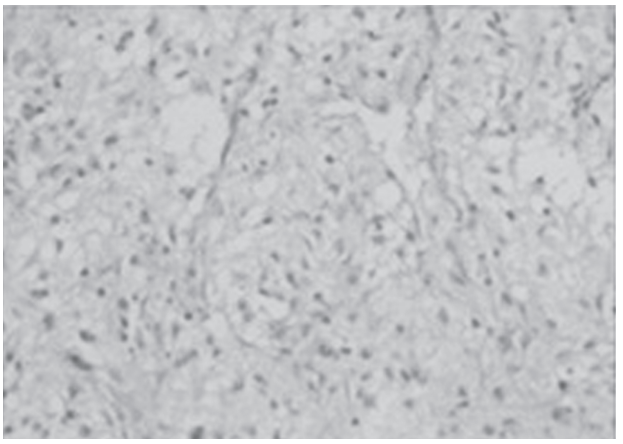

Figure 3 HPE demonstrate cells with uniform round nuclei in a myxoid background (40X)

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).[3] 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.[4] 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 rarest variant of ALT/WDL.[3] 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.[2, 5] 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.[2] 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.[6] Ultimately histopathology along with immunohistochemistry studies clinches the diagnosis.[7] A few cases of atypical lipomatous tumor/well-differentiated liposarcoma of the gingival has also been recorded in literature.[8] The differential diagnosis of S-ALT/WDL in the limbs includes Spindle Cell Lipoma (SCL) and spindle cell myxoid liposarcoma. Spindle cell lipoma occurs subcutaneously in the posterior neck, upper back and shoulder region. 90% of the lesion is made up of uniform appearing mature adipocytes. Immunohistochemically, lipogenic cells in ALT/WDL show S-100 immunoreactivity and spindle cells show focal presence of CD 34 immunoreactivity.[9] These tumors do not metastasize hence require less aggressive therapeutic management. Accurate histopathological subtyping is absolutely essential at the earliest as it impacts both treatment strategies and outcome.[2] Recently, cytogenetic and molecular studies have highlighted that WDL/ALT are characterized by giant marker and ring chromosomes containing ampliûed sequences of 12q13-15 which is the site for several genes including MDM2, CDK4, GL1, SAS and HMGIC.[1, 2]

However, WDSCL lacks the MDM2 and CDK4 gene ampliûcations and instead show monosomy of chromosome.[1, 2, 9] Another study depicted that unlike epithelial neoplasms, the malignant transformation of a pre-existing mesenchymal tumor has been questioned for a long time.[10] 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.[11] 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[10, 12], 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.[1] 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.[2] Previous literature shows that liposrcoma 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

REFERENCES

1. Mentzel T, Palmedo G, Kuhnen C et al. Well-differentiated spindle cell liposarcoma (‘atypical spindle cell lipomatous tumor’) does not belong to the spectrum of atypical lipomatous tumor but has a close relationship to spindle cell lipoma: clinicopathologic, immunohistochemical, and molecular analysis of six cases. Mod Pathol 2010;23:729-736.
2. Weiss SW, Goldblum JR. Benign lipomatous tumors and Liposarcoma. Enzinger and Weiss’s Soft Tissue Tumors 1998 2nd ed. p. 429-516.
3. Laurino L, Furlanetto A, Orvieto E, Dei Tos AP. Well-differentiated liposarcoma (atypical lipomatous tumors). Semin Diagn Pathol 2001;18:258-262.
4. Weiss SW, Rao VK. Well-differentiated liposarcoma (atypical lipoma) of deep soft tissue of the extremities, retroperitoneum, and miscellaneous sites. A follow-up study of 92 cases with analysis of the incidence of “dedifferentiation”. Am J Surg Pathol 1992;16:1051-1058.
5. Bulus H, Günbey E, Simsek GG, Coskun A, Morkavuk B. Giant atypical lipomatous tumor/well-differentiated liposarcoma of the neck. J Craniofac Surg 2011;22:1122-1124.
6. Kransdorf MJ, Bancroft LW, Peterson JJ, Murphey MD, Foster WC, Temple HT. Imaging of fatty tumors: distinction of lipoma and well-differentiated liposarcoma. Radiology 2002;224:99-104.
7. Collins BT, Gossner G, Martin DS, Boyd JH. Fine needle aspiration biopsy of well-differentiated liposarcoma of the neck in a young female. A case report. Acta Cytol 1999;43:452-456.
8. Kim YB, Leem DH, Baek JA, Ko SO. Atypical lipomatous tumor/well-differentiated liposarcoma of the gingiva: a case report and review of literature. J Oral Maxillofac Surg 2014;72:431-439.
9. Italiano A, Chambonniere ML, Attias R, Chibon F, Coindre JM, Pedeutour F, Monosomy 7 and absence of 12q ampliûcation in two cases of spindle cell liposarcomas. Cancer Genet Cytogenet 2008;15:99-104.
10. Mentzel T. Biological continuum of benign, atypical, and malignant mesenchymal neoplasms-does it exist? J Pathol 2000;190:523–525.
11. Dei Tos AP, Mentzel T, Newman PL et al. Spindle cell liposarcoma, a hitherto unrecognized variant of liposarcoma. Analysis of six cases. Am J SurgPathol 1994;18:913-921.
12. Dei Tos AP, Doglioni C, Piccinin S, *et al.* Coordinated expression and amplification of the MDM2, CDK, and HMGI-C genes in atypical lipomatoustumours. J Pathol 2000;190:531-536.