Sarcoma Research - International

Austin Publishing Group

Open Access Full Text Article

Case Report

A Spindle Cell Well Differentiated Liposarcoma / Atypical Lipomatous Tumor of the Neck: A Rare Case with Immunohistochemical Study

Khandeparkar SGS, Deshmukh SD*, Gaopande VP, Jadhav AB and Vaishya H

Department of Pathology, Shrimati Kashibai Navale medical college and general hospital, India

***Corresponding author:** Deshmukh SD, Department of Pathology, Shrimati Kashibai Navale Medical College and General Hospital, Westely Bypass ,PUNE, India, Mobile num: 9604791261; Email: siddhigsk@yahoo.co.in

Received: July 10, 2014; **Accepted:** Aug 04, 2014; **Published:** Aug 05, 2014

Abstract

Liposarcomas account for 20% of soft tissue sarcomas. It is rare in head and neck region particularly in the form of subcutaneous mass in the neck. There is paucity of information on head and neck subcutaneous liposarcomas of spindle cell atypical lipomatous tumor/well differentiated liposarcoma type (S-WDL/ALT). We encountered a case of S-WDL/ALT in 56 year old male who underwent surgical excision as a treatment modality and was kept for follow-up. At 1 year follow up the patient is free of disease. We present this case to highlight the fact that, this type of liposarcoma behaves in low grade less aggressive form along with review of literature.

Keywords: Spindle cell; Well differentiated liposarcoma; Neck; Atypical lipomatous tumor; WDL/ALT

Introduction

Sarcomas encountered in Head and Neck Region (HNS) show wide variety of histo-morphological types and grades. Liposarcoma of the head and neck represents approximately 1% of HNS [1]. They are a group of malignant neoplasms that affect critical structural units of head and neck 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 [2]. WDL/ALT tend to develop in the deep muscles of extremities (75%), retroperitoneum (20%) and other miscellaneous sites [3]. In this case, the tumor was located in the subcutis region of the neck which is very unusual. This tumor is composed of prominent spindle cell proliferation and varying sized adipocytes admixed 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 left side of the neck in 56 year old male. These tumors do not metastasize hence require less aggressive therapeutic management [2]. Accurate histopathological subtyping is absolutely essential as it impacts treatment strategies and outcome.

Case Report

A 56 year old male presented to surgery outpatient department with a progressively increasing left sided neck mass. This was noticed one year prior to presentation which was small initially and gradually increased to the present size. Local examination showed 8x7 cm, non-tender, ill defined, soft to firm, pseudofluctuant subcutaneous swelling in the neck. It was extending from midline to the angle of mandible. Overlying skin was unremarkable. Superiorly it extended up to the lower border of the mandible and inferiorly 2 cm above the clavicle. There was no previous history of lipomas in this case neither any remarkable family history associated. Systemic examination findings were non-contributory. With the provisional diagnosis of lipoma, Computed Tomography (CT) was done to know the nature and extent of disease process. CT of the neck showed a large ill- defined heterogenous lesion measuring 9.8x6.6x6.5cm, with predominantly fat densities and focal cystic spaces extending into the left parotid, submandibular, parapharyngeal and prevertebral region (Figure 1a). The mass reached inferiorly up to the thoracic inlet without intra-thoracic extension. Laterally it was abutting carotid vessels without compression. Left sternomastoid muscle was displaced posterolaterally. Esophagus was compressed. There was compression on the posterior aspect of the laryngopharyngeal airway. Laryngeal cartilages, thyroid laminae and thyroid gland appeared normal. Radiological diagnosis of lipoma or a neurofibroma was suggested. Fine Needle Aspiration Cytology (FNAC) was performed.

Fine Needle Aspiration Cytology (FNAC) revealed mild to moderately cellular smears composed of tissue fragments of cells with indistinct cell margins, moderate amount of eosinophilic cytoplasm and spindly nuclei of varying size. Few adipocytes were seen entrapped within the spindle cell fragments. The background was myxoid. Cytological report of low grade spindle cell neoplasm was offered and histopathological confirmation was advised.

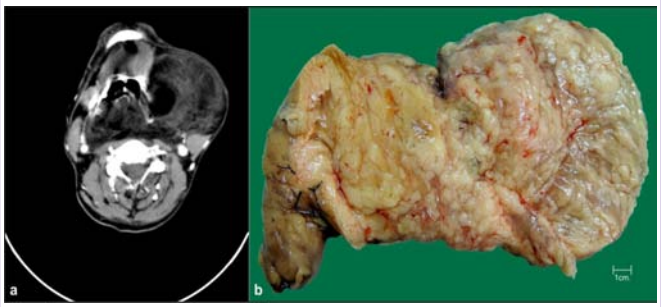

Figure 1: **a)** CT showing a large well defined heterogenous lesion measuring 9.8x6.6x6.5cm, with cystic and fat densities. **b)** Gross photograph: cut section shows a well circumscribed, yellowish lobulated tumor with whitish streaks measuring 21x12x2.5cm with myxoid areas.

Citation: Gaurish SKS, Sanjay D, Vandana PG, Amruta BJ and Vaishya H. A Spindle Cell Well Differentiated Liposarcoma / Atypical Lipomatous Tumor of the Neck: A Rare Case with Immunohistochemical Study. Sarcoma Res Int. 2014;1(1): 3.

In view of radiological findings and FNAC report, clinician chose to excise the lesion in toto. Left sided neck incision was given. A soft tissue mass measuring 15x10x10cm was viewed intraoperatively occupying the left side of the neck. The mass was seen pushing left carotid and internal jugular vein posteriorly. Superiorly it extended up to the skull bone. Medially it was seen extending through the prevertebral space into the opposite side of the neck exhibiting dumbbell appearance. So the incision was deepened and subplatysmal skin flap was raised. Mass was separated from all around. The sternocleidomastoid muscle was divided and removed along with the neck mass. The incision was later closed with drain inserted. What was the surgical approach?

On Gross examination, the tumor was circumscribed, lobulated and soft to firm measuring 21x12x2.5cm. On cut section the tumor appeared yellowish, lobulated, with myxoid areas (Figure 1b).

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 was seen. Stroma showed few atypical spindle cells, thin walled blood vessels, thick and thin fibrous septae with focal myxoid areas. (Figure 2a)The spindle cells revealed bland oval nuclei and moderate amount of eosinophilic cytoplasm. Few large atypical cells were noted (Figure 2a). No areas of necrosis and hemorrhage noted. On careful search, occasional lipoblasts were seen in the sections taken from the periphery of the tumor (Figure 2a inset, 2b). Based on histopathological appearance, atypical lipomatous tumor was considered and representative sections were subjected to immunohistochemical studies.

Immunohistochemistry (IHC) was performed with the following panel of antibodies viz. S-100 (Leica), CD-34 (clone QBEnd/10, Novacastra) desmin (clone33, Dako) and myogenin (clone Myf-4, Leica). Cell proliferative marker like ki-67 (clone MM1, dako) was also included. The lipogenic cells showed strong membrane immunoreactivity for S-100 (Figure 2c). Spindle shaped cells showed strong immunoreactivity for CD-34 (Figure 2d). The tumor cells showed Ki-67 labelling index of 7%. Desmin and myogenin were non-immunoreactive. With the above morphologic and IHC findings, a diagnosis of S-WDL/ALT of the neck was confirmed.

Since this tumor is known to have low grade behaviour and the margins of excision were free , no chemotherapy/ radiotherapy was given. However the patient was advised regular close follow up. The patient showed no signs of recurrence even after 1 year follow-up.

Discussion

Liposarcomas are histologically divided into five subtypes: myxoid, pleomorphic, dedifferentiated, round cell, and ALT/WDL [4]. ALT/WDL are low grade, non-metastasizing, malignant neoplasms composed primarily of mature adipose tissue. 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 [5]. Presentations in other sites are uncommon. Especially in the head and neck. Individual case report defining cytological criteria for the diagnosis of /ALTWDL in the neck has been documented [6].

ALT/WDL is further subdivided in the adipocytic (lipoma-like), sclerosing, inflammatory, and spindle cell subtypes. S-ALT/WDL is a rarest variant of ALT/WDL [4]. It was first described in 1994. It is a distinct neoplasm which tends to occur in the subcutis of the shoulder region and extremities. Few cases have been described in the head and neck [3].

The size of the neoplasms ranges from 1.5 to 10cm. In our case, the tumor size was 21x12x2.5cm [6]. This gigantic size of S-ALT/WDL in the neck is so far not mentioned in the literature.

Histologically, this biphasic tumor 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 [3].

Imaging plays a major role in defining the extent of the tumor to nearby vital structures for treatment planning and deciding surgical approach decision. Discuss the choice of CT over MRI in the index patient Features that suggest malignancy include increased patient age, large lesion size, presence of thick septa, presence of nodular non-adipose mass like areas and decreased fatty areas better demonstrated on MRI. However in this case the patient underwent CT scan only due to financial constraints [7].

Cytological findings described in literature include mixture of adipocytes supported by fibro-vascular septa with hyperchromatic and enlarged nuclei within the fat and fibrous bands with one or two small nucleoli and scattered lipoblasts. Ultimately histopathology along with IHC studies clinches the diagnosis [8].

The differential diagnosis of S-ALT/WDL in the neck 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. Spindle cells constituting the tumor are bland showing no mitosis and are CD 34 positive. Collagenous stroma is prominent. Myxoid areas and prominent, thick walled, arborizing blood vessels may be found in the stroma. Lipoblasts

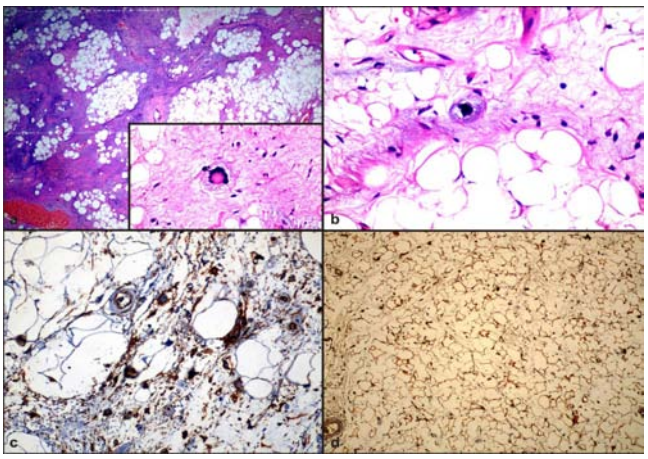

Figure 2: **a)** Photomicrograph of tumor: spindle cells arranged in short interlacing fascicles interspersed with lipogenic cells separated by thick and thin vascular fibrous septae. (x100), **inset**) Pleomorphic lipoblast **b)** Lipoblasts showing central hyperchromatic nucleus which is indented by multivacuolated cytoplasm. (x400) **c)** Spindle shaped cells showing strong immunoreactivity for CD-34.(x400) **d)**The lipogenic cells showing strong membrane immune reactivity for S-100.(x100).

are absent. Myxoid areas can pose diagnostic problems. However, myxoid liposarcoma shows chicken wire vascular patternwhich was absent in our case. Instead our case showed singly scattered large atypical nuclei consistent withspindle cell ALT/WDL [2].

Immunohistochemically, lipogenic cells in ALT/WDL show S-100 immunoreactivity and spindle cells show focal presence of CD 34 immunoreactivity [3].

Cytogenetic and molecular genetic studies contribute substantially inorder to detect mostsubtypesof lipomatous tumors. ALT/WDL shows amplification of the MDM2 and/or CDK4 genes or an immunohistochemically detectable expression of MDM2 and/or CDK4 in most cases. This is not apparent in case of most S-ALT/WDL. Monosomy of chromosome 7 and absence of 12q amplification has been reported in S-ALT/WDL.ALT/WDL is characterized by supernumery ring and/or giant marker chromosomes containing amplified material of the q13–15 regions of chromosome 12.Interestingly, S-ALT/WDL is found to show deletion of material of the long arm of chromosome 13. This Rb-1 deletion is a characteristic finding in spindle cell lipoma. Given the similarities of SCL and S-ALT/WDL, it can be speculated that S-ALT/WDL represents the atypical/low-grade counterpart of SCL 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 S-ALT/WDL [3].

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 [3].

Wide local excision is not always achievable in most of head and neck sarcomas due to the complex anatomy and close proximity of major vital structures to primary tumor. When the surgical margins are not adequately free, post-operative radiotherapy and / or chemotherapy should be considered. Multidisciplinary management of soft tissue sarcomas in the neck requires establishment and execution of detailed treatment plan in a coordinated timely manner for successful local regional control with minor functional and/or cosmetic deficits as result of treatment [1].These tumors do not metastasize hence require less aggressive therapeutic management. In this case, the patient was disease free on 1 year follow up [9]. Accurate histopathological subtyping is absolutely essential as it impacts treatment strategies and outcome [3].

Our experience with present case highlights that this subtype of lipogenic tumor needs careful histopathological evaluation for accurate diagnosis. These patients may still require aggressive management with a long term follow up as a result of difficulty in achieving free surgical margin especially when located in the neck.

References

1. Aljabab AS, Nason RW, Kazi R, Pathak KA. Head and neck soft tissue sarcoma. See comment in PubMed Commons below Indian J Surg Oncol. 2011; 2: 286-290.
2. Mentzel T, Palmedo G, Kuhnen C. 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.
3. Weiss SW, Goldblum JR. Benign lipomatous tumors and Liposarcoma. 2nd edn. In: Weiss SW, Goldblum JR, editors. Enzinger and Weiss's Soft Tissue Tumors, Philadelphia, Mosby; 1998; 429-516.
4. Laurino L, Furlanetto A, Orvieto E, Dei Tos AP. Well-differentiated liposarcoma (atypical lipomatous tumors). See comment in PubMed Commons below Semin Diagn Pathol. 2001; 18: 258-262.
5. 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". See comment in PubMed Commons below Am J Surg Pathol. 1992; 16: 1051-1058.
6. Buluş H, Günbey E, Simşek GG, Coşkun A, Morkavuk B. Giant atypical lipomatous tumor/well-differentiated liposarcoma of the neck. See comment in PubMed Commons below J Craniofac Surg. 2011; 22: 1122-1124.
7. Kransdorf MJ, Bancroft LW, Peterson JJ, Murphey MD, Foster WC, Temple HT. Imaging of fatty tumors: distinction of lipoma and well-differentiated liposarcoma. See comment in PubMed Commons below Radiology. 2002; 224: 99-104.
8. 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. See comment in PubMed Commons below Acta Cytol. 1999; 43: 452-456.
9. Kim YB, Leem DH, Baek JA, Ko SO. Atypical lipomatous tumor/well-differentiated liposarcoma of the gingiva: a case report and review of literature. See comment in PubMed Commons below J Oral Maxillofac Surg. 2014; 72: 431-439.

Sarcoma Res Int - Volume 1 Issue 1 - 2014
Submit your Manuscript | www.austinpublishinggroup.com

Citation: Gaurish SKS, Sanjay D, Vandana PG, Amruta BJ and Vaishya H. A Spindle Cell Well Differentiated Liposarcoma / Atypical Lipomatous Tumor of the Neck: A Rare Case with Immunohistochemical Study. Sarcoma Res Int. 2014;1(1): 3.