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Lipofibromatosis Presenting as a Neck Mass in Eight-Years Old Boy – a Case Report

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Lipofibromatosis is a very rare pediatric neoplasm with a histologically distinctive fibrofatty pattern. Some of the authors believe that this term should encompass several rare soft tissue tumors of childhood and that tumors of childhood which contain fat as an integral component warrant new classification. We present a case report of a neck tumor in eight-years old boy which shares clinical and morphological features similar to those originally described by Fetch et al. with peaceable with literature results of immunohistochemical stains.

Introduction

In 2000 Fetch et al. first described a series of distinctive soft tissue tumors which they termed lipofibromastoses. Lipofibromatosis is a rare entity, to our knowledge about 50 cases have been reported in the literature. Those lesions appeared exclusively in children from birth to early second decade of life and in some cases have been congenital also. There is an over 2:1 male predominance and predilection for distal extremities. Some authors believe that lipofibromatosis encompasses several misinterpreted rare soft tissue pediatric neoplasm previously diagnosed as infantile or juvenile fibromatosis, a variant of fibrous hamartoma of infancy, and a fibrosing lipoblastoma. Currently according WHO classification of soft tissue tumors lipofibromatosis is so-called infantile fibromatosis, nondesmoid type [3, 6].

Clinical History

The 8-years old boy was diagnosed due to the presence of neck mass observed from two months. In physical examination a solid tumor without accompanied lymphadenopathy was noticed. There was no other abnormalities in physical examination or laboratory tests. The MR revealed the intramuscular pathologic mass (from C2 to C7) 25x20x47mm in its diameter, with focal differences of density and without surrounding soft tissues, vessels or vertebras inflitration. Radiologist made a suspicion of cavernous haemangioma or teratoma.

Materials and Methods

Described case was identified for our studies from the consulting files of the Department of Pathology of the Age of Development and Department of Pathology Konopnicka Memorial Hospital Medical University of Łódź. From tumor tissue samples paraffin blocks about the thickness 3-4 of micrometers were prepared and stained with hematoxylin and eosin (H&E). Slides were examined with computer image analysis system (Multi Scan Base v. 8.08 - Computer Scanning System, Ltd.). Microscopic pictures (Nikon Microphot FXA) were transferred to the computer by camera (CC2OP). We performed immunohistochemical stains with DAKO antibodies: Smooth Muscle Actin (SMA), S-100 Protein, CD34, Desmin (DES), MyoD1, Myogenin, CD99, Bcl-2, NSE, EMA, Ki-

67 and with use of immunoperoxidase reaction according to Hsu (EnVision + the System, Peroxidase – DAB). The antibodies, their clones and descriptions are showed in Table1. Appropriate positive and negative controls were used throughout. Clinical data were obtained from the clinician and reviewed.

Results

Histopathological findings

Grossly the lesion was 45x22x20mm with yellow-whitish fatty or lipoma like look, focally more firm but without macroscopically identifiable areas of necrosis or hemorrhages and with ill-defined margins. The lesion was excised with overlying normal epidermis.

In microscopic examination the fatty component, which appear to predominate in most of the picture, was composed of mature adipocytes disrupted by spindled cells (Fig. 1 and 2). Spindled fibroblastic element had focal fascicular growth and involved the fat septa. The tumor entrapped surrounding skeletal muscles and was excised incompletely. There was no solid fibrous growth or primitive nodular fibromyxoid component. Mitoses or features of nuclear atypia were absent. Histologic examination excluded the diagnosis of haemangioma or teratoma.

Immunohistochemical study

The CD99, SMA, BCL-2, NSE and S-100 expression with typical patterns were observed in the spindled fibroblastic cells. MyoD1, Myogenin, Desmin, EMA and CD34 were negative. Ki-67 antibody was detected in single cell nuclei only. Details of immunohistochemical stains results are showed in Table1.

TABLE 1
Results of immunohistochemical examination of the lesion

No.	Antibody	Clone	Description	Result
1.	CD99 (MIC2)	12E7	Monoclonal-Mouse Anti-Human	Positive, focal
2.	S-100	-	Polyclonal-Rabbit Anti-S 100	Positive, dispersed
3.	BCL-2 Oncoprotein	124	Monoclonal-Mouse Anti-Human	Positive, focal
4.	NSE	BBS/NC/VI-H14	Monoclonal-Mouse Anti-Human	Positive, focal
5.	SMA	1A4	Monoclonal-Mouse Anti-Human	Positive, focal
6.	Ki-67	MIB-1	Monoclonal-Mouse Anti-Human	Positive, in single nuclei (index = 3)
7.	MyoD1	5.8A	Monoclonal-Mouse Anti-MyoD1	Negative
8.	Myogenin	F5D	Monoclonal-Mouse Anti-Myogenin	Negative
9.	Desmin	D33	Monoclonal-Mouse Anti-Human	Negative
10.	EMA	E29	Monoclonal-Mouse Anti-Human	Negative
11.	CD34	QBEnd 10	Monoclonal-Mouse Anti-Human	Negative

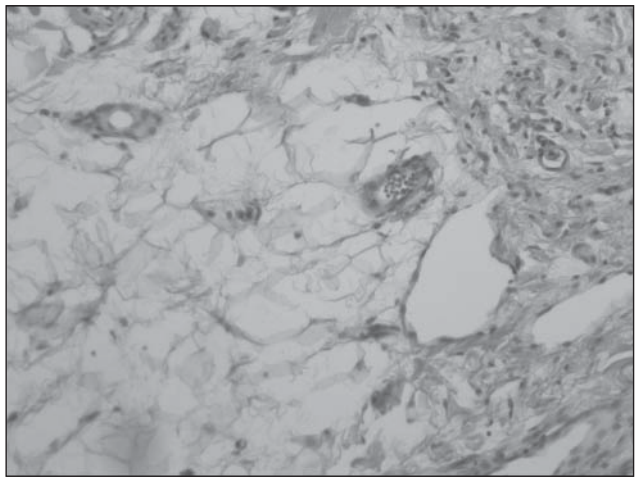

Fig. 1. Lipofibromatosis – mature fatty tissue as an integral part of the lesion, H&E. Oryg.magn. 400x.

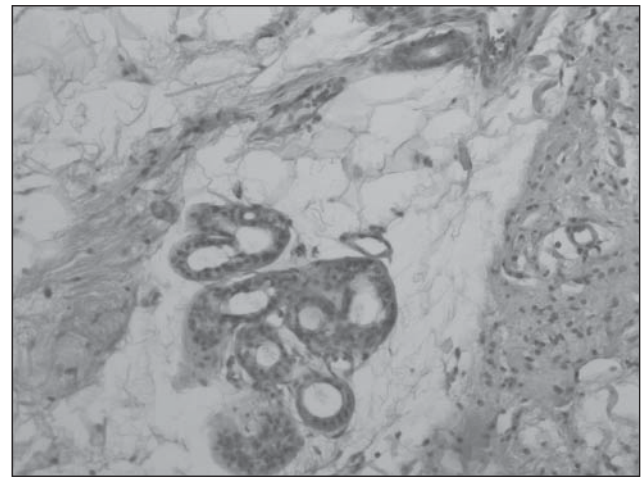

Fig. 2. Lipofibromatosis – adipocytes disrupted by a spindled cell component, H&E. Oryg. magn. 400x.

Discussion

We presented herein the clinicopathological picture of the lesion, which shares very similar clinical and morphological features to those originally described by Fetch et al. [3]. Typically lipofibromatosis appears in children below 12-years old, mostly males. So-called infantile fibromatosis, non-desmoid type demonstrates a predilection to for the distal extremities (the hand, arm, leg or foot), trunk and head [1, 3, 7, 8]. Lipofibromatosis cases exhibit slow growth and, as the result rarely exceed 5 cm, with the median size of 2 cm. Grossly lesions are poorly circumscribed and entrap surrounding skeletal muscles, skin adnexa, vessels or nerves. Immunohistochemical examination was similar to the initial study, either. The most common immunoprofile of lipofibromatosis is focal staining of the spindle cells with CD99, SMA, BCL-2 and less frequent positivity for S-100 protein and EMA. Typically negative is stain with desmin [3, 5].

Based on macroscopic view of the prominent fat component with more firm areas and poorly-defined margins, a number of adipocytic tumors enter the differential diagnosis. Lipomatous or fibro-lipomatous lesions range from benign to locally aggressive and to tumors with metastatic potential and include: angiomyolipoma, lipoma, (pleomorphic, spindle cell), atypical lipomatous tumor, well-differentiated liposarcoma, hibernoma, lipoblastoma/lipoblastomatosis, hemosyderotic fibrolipomatous lesion, fibrous hamartoma of infancy and fibrohistiocytic lipoma. Morphological features found in microscopic examination: the same size, mature adipocytes, lack of nuclear atypia, necrosis and mitotic activity (confirmed by low Ki-67 index) allow to exclude malignancy. Simple morphological pattern with the prominent mature fat component with spindled cells involved fatty tissue septa without primitive mesenchymal component, exclude most of the non-malignant conditions listed above, specially lipomatosis, lipoblastoma/lipoblastomatosis and fibrous hamartoma of infancy [6].

Spindle cell lipoma, based on both morphological characteristics and anatomical location should be taken into consideration. Those tumors typically occur around the upper back/neck region but they are usually circumscribed and are composed of collagen bundles, mast cells and myxoid stroma. Listed in the differential diagnosis fibrohistiocytic lipoma was excluded due to cells origin and pattern. At the end, examined lesion appeared differ from heamosiderotic fibrolipomatous tumor which is rare but distinctive entity which shares morphological features with lipofibromatosis. Spindle cells found in this lesion are less primitive and form whorls which is generally not seen in lipofibromatosis. In addition hemosiderin deposites absent in examined by us case, even found in lipofibromatosis are focal and inconspicious opposite to heamosiderotic fibrolipomatous tumor [2]. In summary, the hallmark of examined by us lesion was the mixture of mature adipocytes and fibroblastic spindle cells which involved fatty tissue septa and the absence of primitive nodular fibromyxoid pattern, solid fibrous growth or nuclear atypia. However diagnosed as a lipofibromatosis lesion probably comprises part of the spectrum of rare and easy to misdiagnosed tumors of childhood which contain fat as an integral component and which probably warrant separate classification.

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