Lipomatous variant of angiomyofibroblastoma-like tumor in male through fine needle aspiration

"*False positive just is in the eye of the beholder*"

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ABSTRACT

Angiomyofibroblastoma-like tumor of male is an extremely rare benign mesenchymal tumor. It commonly occurs in middle-age and is mainly localized in the inguinoscrotal region. It is originated from perivascular stem cells and has the capacity of lipoid and myofibroblastic differentiation that may allow the observation of lipomatous differentiation. We report a case of lipomatous variant of angiomyofibroblastoma-like tumor of male using fine needle aspiration biopsy from the scrotal region of a 53 years-old man. Differential diagnosis could be a real bane for the pathologist in this particular variant. This diagnose should be kept in mind to prevent overlapping with the malignant lipomatous tumors.

Key-words: Angiomyofibroblastoma-like tumor of male; cellular fibroma; lipomatous variant; fine needle aspiration

Variante lipomatosa do tumor tipo-angiomiofibroblastoma em homens através da citologia aspirativa por agulha fina

"*Falso positivo apenas aos olhos de quem o vê*"

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RESUMO

O tumor tipo-angiomiofibroblastoma é um tumor mesenquimatoso benigno extremamente raro. Este tipo de tumor afeta homens, geralmente na meia-idade, e localiza-se primordialmente na região inguino-escrotal. Esta neoplasia tem origem nas células estaminais perivasculares e tem a capacidade de diferenciação lipoide e miofibroblástica, o que poderá permitir a observação de um padrão lipomatoso diferenciado. Neste trabalho, reporta-se o primeiro caso de uma variante lipomatosa de um tumor tipo-angiomiofibroblastoma num homem de 53 anos de idade, diagnosticado a partir de uma biópsia aspirativa por agulha fina da região escrotal. O diagnóstico diferencial desta condição neoplásica pode revelar-se extremamente difícil para um patologista, em particular quando se trata da variante em causa. Assim, alerta-se para a necessidade do conhecimento deste tipo de tumor de forma a evitar a atribuição de um falso diagnóstico de tumor maligno lipomatoso.

Palavras-chave: Tumor tipo-angiomiofibroblastoma; variante lipomatosa; citologia aspirativa por agulha fina

INTRODUCTION

Cellular angiofibroma also known as angiomyofibroblastoma-like tumor (AMF-like) is a rare benign mesenchymal tumor; which is characterized by subcutaneous tissue of pelvic region of middle-aged female and male population[1]. This tumor is mainly composed of two distinct components: spindle cell and prominent blood vessels[1.] However, subtle and ordinary morphology may cause significantly perversive differential diagnosis. The precise histogenesis of AMF-like tumor is still elusive, nonetheless, perivascular pluripotential stem cells that reinforces the myoepithelial or fatty transformation is the most suggested theory[2-4]. Based on the theory, lipomatous variant is proposed by Laskin et al in 1997[3-4]. As a third component, lipomatous differentiation poses the differential diagnosis of AMF-like tumor even complicated.

This manuscript reports a false positive case focusing the difficulties on making the correct diagnosis in this rare cytological entity.

CASE REPORT

The patient was a 53 years old man presenting a subcutaneous mass of 12 cm in size that extended from right gluteus maximus to scrotum. Physical examination and ultrasonographic findings revealed a mobile vaguely delineated mass and the preliminary diagnosis made by radiologist and clinicians was “suspicious for sarcoma”.

Three samples were taken with fine needle aspiration cytology (FNAC) by using a 25 gauge needle, and adequacy was assessed with Diff-quik stain. The other slides were immediately fixed with alcohol solution and stained with PAP EA 50. The remaining material used for cell-block preparation.

Under microscopic evaluation, smears were cellular resembling a lipomatous neoplasm, with groups of adipocytes having a single peripherically located nucleus with a large inclusion and multiple vacuoles in the cytoplasm scalloping the nuclei with a thin capillary network (**Fig.1** and **2)**. Interestingly fibrotic and collagenous stromal fragments were remarkable in some areas of the slides (**Fig.3**).

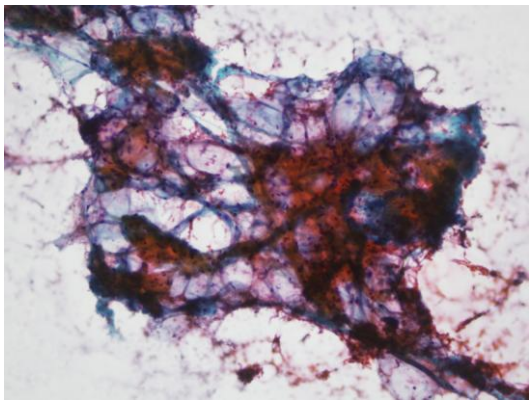

Fig1. Admixture of lipocytes and multivacuolated lypoblasts with a thin capillary network. x400. PAP EA50 stain.

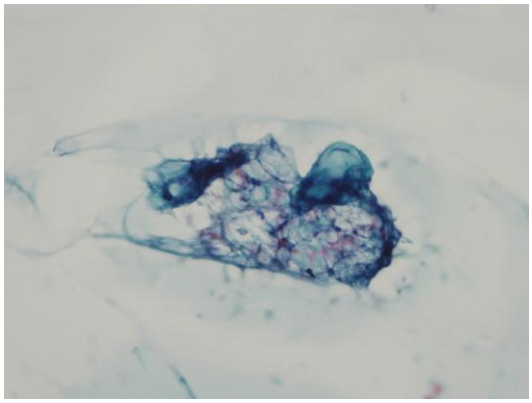

Fig2. Note the atypical lipoblasts shows scalloped, hyperchromatic prominent nuclei with a variable size multivacuolation in cytoplasm. x600. PAP EA50 stain.

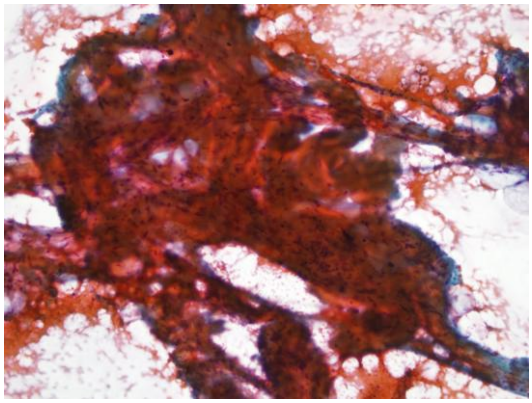

Fig3. Significant amount of fibrotic, collagenous stromal fragments were seen in the background. x400. PAP EA50 stain.

Cell-block preparations consisted of blood cloth and no lesional cells were observed. FNAC diagnosis was “atypical lipomatous tumor” based on the cell population, clinical/radiologic findings and superficial localization.

Surgical excision was performed. Macroscopically the tumor was well-circumscribed, showing multinodular appearance of edematous and collagenous stroma. The tumor includes two types of components: short spindle cell fascicles lacking any particular pattern and evenly distributed vessels without any branching pattern (**Fig.4**).

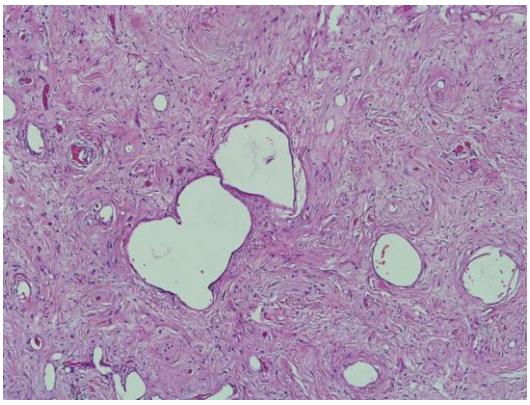

Fig4. Spindle cell bundles with a variable size of sleave-like vasculature were observed in pathology specimen. x600. H&E stain.

Intranuclear inclusions and longitudinal grooves were noted in the spindle cell component. The cytoplasm of these cells was mostly pale-eosinophilic. Thick and thin-capillary sized vessels were striking and representing hemangiopericytoma-like appearance due to the perivascular fibrosis and hyaline degeneration (**Fig.5**).

Perivascular lymphocytic aggregates were also prominent. Tumor represents hypocellular and hypercellular zones in some areas. Admixture of mature adipocytes was distributed in the lesion.

Immunohistochemically, spindle cells extensively and intensely expressed CD34 and had focal weak staining with SMA. There was no immunoreactivity with S100, CD117, EMA and desmin antibodies. A weak positivity in the

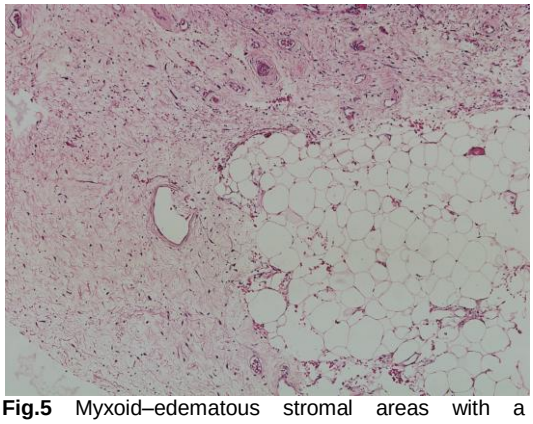

Fig.5 Myxoid–edematous stromal areas with a lipomatous differentiation. x400. Diff-Quik stain.

nucleus was seen with immunostaining for estrogen and progesterone receptors (**Fig.6**).

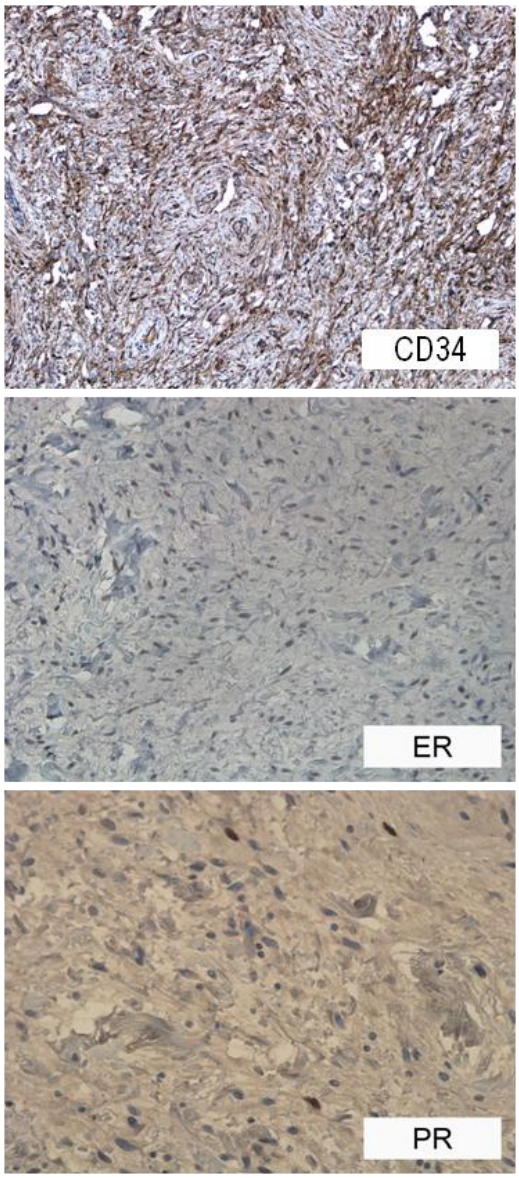

Fig.6 Immunohistochemistry method show typical positivity for CD34, ER and PR antigens. x400.

DISCUSSION

Cellular angiofibroma was introduced by Nucci *et al*. in 1997 and it was accepted as a distinct entity from angiomyofibroma in female[3]. The term AMF-like tumor in male was endorsed by Laskin et al[4]. However, Iwasa and Fletcher supported that both cellular angiofibroma and AMF-like tumor are the same entities[1].

Although AMF-like tumor is a benign soft tissue neoplasm, there is a small group of cases of sarcomatous transformation reported in the literature[5,6]. The tumor occurs in vulvo-vaginal region in women and inguino-scrotal/paratesticular region in men[1]. Genetically, AMF-like tumor shows loss of chromosome 13 and 16 as in spindle cell lipoma and mammary-type myofibroblastoma which is considered a close kinship in all these entities[7]. The presence of lipomatous component was reported in few case reports[2,3].

Differential diagnosis encompasses AMF, spindle cell lipoma, solitary fibrous tumor, mammary-type myofibroblastoma and aggressive angiomixoma[1]. However, in the current case all main spectrum of lipomatous tumors must be included in the differential diagnosis instead of the entities written above. Lipomatous tumors on cytology were assessed such as lipoma, lipoblastoma and atypical lipoma/liposarcoma[8]. All these entities have numerous variants as well as the non-lipomatous tumors which may also have lipomatous variants, being aware of the variants with academic concern instead of clinical significance, like in the present case. Well-differentiated liposarcoma/atypical lipomatous tumor is a malignant, non-metastazing tumor that if superficially localized it is named atypical lipomatous tumor, and if deeply-sited it is referred as well-differentiated liposarcoma[9]. Well-differentiated liposarcoma/ atypical lipomatous tumor peak incidence is 60-70 years old and the most common sites are limbs and retroperitoneal region[9]. Moreover, scrotal and paratesticular liposarcomas were also reported in the literature[10,11]. Cytomorphology of atypical lipomatous tumor is a real challenge even in resection specimens, where the yield is generally poor and the stromal fragments are hardly seen in FNAC specimens[12]. Mature adipocytes can easily be observed as well as a limited number of lipoblasts. Prominent nuclear atypia and arborizing vasculature are unusual findings[12]. Spindle cell lipoma also should be considered in the differential diagnosis[1]. This entity is seen in subcutaneous tissue of neck and shoulder area, and cytology shows bland cells, but might have nuclear irregularities or scarce intranuclear inclusions[12]. Lack of atypical lipoblasts and capillary network as well as the absence of floret cells, ropy collagen fibers and mast cells helped in eliminating this entity[8,12].

Lipoblastoma is a benign neoplasm of embryonal fat tissue that usually occurs at 3 years of age, with a male predominance, and that can be seen in pelvic area. It is very uncommon in adults. Angiolipoma is a tumor of the second decade of life and it is commonly localized in forearm[12]. Suggested characteristics of angiomyolipoma such as pleomorphic endothelial cells and mast cells[12] were not detected in the current case.

Immunohistochemically, AMF-like tumor expresses CD 34 in 30%-60% of the cases. SMA and desmin antigens are not necessarily expressed[9]. Almost all cases of AMF-like tumor of male are negative for S100 and keratins immunostaining and positive for ER and PR immunostaining[9].

In the current case, atypical lipoblasts and size of the subcutaneous lesion suggested atypical lipomatous tumor. After a detailed histopathology and immunohistochemical analysis of pathology specimens, diagnosis was turn out to AMF-like tumor of male.

The present case does nicely remind us a phenomenon paper "*The unpredictable fatty tumors*" of Sawyer et al that was published in 1968[13]. Although experienced evaluation of pathologist/cytopathologist, the diagnostic accuracy of lipomatous lesions still might be limited, as in the current example. To the best of our knowledge, this is the first case of lipomatous variant of AMF-like tumor of male on FNA that should help us to keep in mind that the variant of soft tissue tumors is evaluated as academic concern. However, considering the possibilities and clinical findings, even in the absence of molecular analysis, it might still be a great help to prevent the false positive for a giant mass as benign tumor, instead of sarcoma as in the current case.

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