Masson's tumor: a soft tissue tumor simulating a tendon cyst. Case report

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SUMMARY: Masson's tumor: a soft tissue tumor simulating a tendon cyst. Case report.

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Introduction. Intravascular papillary endothelial hyperplasia (Masson's hemangioma or Masson's tumor) is a benign vascular disease with an exuberant endothelial proliferation in normal blood vessels. Although relatively uncommon, its correct diagnosis is important because it can clinically be like both benign lesions and malignant neoplasms. We present a case of intravascular proliferative endothelial hyperplasia simulating a tendon cyst both clinically and on ultrasound.

Case report. A 74-year old Caucasian female presented with a 4-month history of soreness and swelling in the fourth finger of the right hand. Ultrasound showed an oval mass with fluid content, referred to a tendon cyst. A wide surgical excision was subsequently performed. The final histological diagnosis was Masson's tumor.

Discussion. The pathogenesis of intravascular papillary endothelial hyperplasia is still unclear but the exuberant endothelial cell proliferation might be stimulated by an autocrine loop of endothelial basic fibroblast growth factor (bFGF) secretion. There are three types of papillary endothelial hyperplasia: primary, or intravascular; secondary, or mixed; and extravascular. The main differential diagnosis is against pyogenic granuloma, Kaposi sarcoma, hemangioma, and angiosarcoma.

Conclusions. Masson's tumor can be like both benign lesions and malignant neoplasms clinically and on ultrasound. For this reason, the right diagnosis can be made only by histology, which reveals a papillary growth composed of hyperplastic endothelial cells supported by delicate fibrous stalks entirely confined within the vascular lumen.

RIASSUNTO: Tumore di Masson: tumore del tessuto molle che simula una cisti tendinea. Caso clinico.

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Introduzione. L'iperplasia endoteliale papillare intravascolare benigna (emangioma di Masson) è una neoplasia benigna caratterizzata da tessuto endoteliale esuberante che colpisce vasi altrimenti normali. È una patologia rara, importante da diagnosticare poiché simula numerose lesioni sia benigne che maligne. Obiettivo di questo lavoro è presentare un caso di tumore di Masson che clinicamente ed ecograficamente simulava una cisti tendinea.

Caso clinico. Donna caucasica di anni 74. Si presenta alla nostra osservazione per la presenza di una massa del quarto raggio della mano destra. L'ecografia dimostrava la presenza di una massa ovalare, a contenuto liquido, compatibile con la diagnosi di cisti tendinea. Pertanto si procedeva ad escissione chirurgica. L'esame istologico poneva diagnosi di tumore di Masson.

Discussione. La patogenesi dell'iperplasia endoteliale papillare non è ancora chiara ma sembra che il tessuto esuberante derivi dalla stimolazione autocrina delle cellule endoteliali dovuta alla secrezione di basic Fibroblast Growth Factor (bFGF). Esistono tre tipi di iperplasia endoteliale papillare: la forma primaria o intravascolare, la secondaria o mista e la terziaria o extravascolare. La diagnosi differenziale deve essere posta principalmente con granuloma piogenico, sarcoma di Kaposi, emangioma e angiosarcoma.

Conclusioni. Il tumore di Masson può simulare clinicamente ed ecograficamente numerose lesioni sia benigne che maligne. Per questa ragione la diagnosi di certezza può essere posta solo con l'esame istologico che dimostra la presenza di papille endoteliali, con un sottile stroma fibroso, che si aggettano nel lume vasale.

KEY WORDS: Masson's tumor - Masson's hemangioma - Intravascular papillary endothelial hyperplasia - Tendon cyst. Tumore di Masson - Emangioma di Masson - Iperplasia endoteliale papillare benigna - Cisti tendinea.

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Introduction

Intravascular papillary endothelial hyperplasia (Masson's hemangioma or Masson's tumor) is a benign vascular disease with an exuberant endothelial proliferation in normal blood vessels, due to the endothelium's differing histopathologic reaction pattern to different inputs (1).

Although relatively uncommon, its correct diagnosis is important because it can clinically resemble both benign lesions (e.g., mucocele, pyogenic granuloma, and hemangioma) and malignant neoplasms (angiosarcoma and Kaposi's sarcoma). The risk of misdiagnosis is higher for histologists unfamiliar with this lesion (2).

Masson's tumor is generally found in the head or upper and lower extremities. Its morphologic findings make differential diagnosis possible against a number of benign and malignant vascular proliferations. The key microscopic feature is the evidence of a papillary growth composed of hyperplastic endothelial cells supported by delicate fibrous stalks entirely confined within the vascular lumen. The lesion should not be mistaken for angiosarcoma, since its clinical behavior is always benign (3).

Case report

A 74-year old Caucasian female was referred to the Plastic Surgery Department with a 4-month history of soreness and swelling in the fourth finger of the right hand. She had no systemic complaints and no recent weight loss. There was no previous trauma to the area and the medical history was non-contributory.

The only clinical finding was a 10 x 9 mm subcutaneous mass in the fourth finger of the right hand (Fig. 1). Blood tests were normal. Ultrasound showed an oval mass with fluid content, referred to a tendon cyst (Fig. 2).

A wide surgical excision was subsequently performed. The specimen at the time of surgery was red to bluish in color and well encapsulated. Histological examination showed an intravascular proliferation of small papillae and the final pathologic diagnosis was Mas-

son's tumor. Standard hematoxylin-eosin staining (magnification x25) revealed free-floating papillary projections within the vascular lumen associated with an adjacent thrombus (Fig. 3).

Clinical follow-up at 8 months showed no evident complications or recurrence.

Discussion

Masson's tumor was first described in an ulcerated hemorrhoidal vein in 1923 by Pierre Masson and remains a rare disorder. It is a benign, generally intravascular tumor which is easily distinguished from an organizing thrombus. It has been given various names, including Masson's tumor, Masson's hemangioma, Masson's intravascular hemangioendothelioma, intravascular papillary endothelial hyperplasia, and reactive papillary endothelial hyperplasia (4).

Extracranial Masson's tumor is more common in females and the incidence peaks in the fourth decade. It can develop within a preexisting vascular malformation or thrombus. Multiple lesions may be examples of Masson's tumor within the setting of a systemic cavernous angiomatosis syndrome (5).

The pathogenesis of papillary endothelial hyperplasia is still unclear but the exuberant endothelial cell proliferation might be stimulated by an autocrine loop of endothelial basic Fibroblast Growth Factor (bFGF) secretion.

Hashimoto et al. have described three types of papillary endothelial hyperplasia. The primary or intravascular form arises in dilated blood vessels, usually within

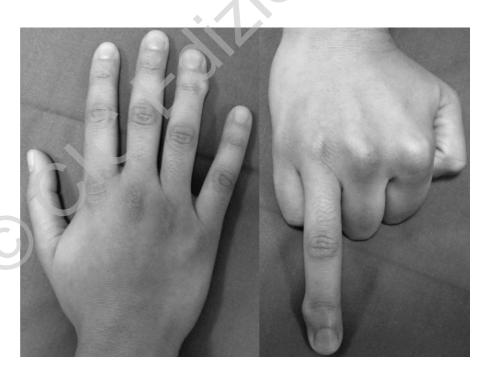


Fig. 1 - Masson's tumor. Subcutaneous mass in the fourth finger of the right hand.

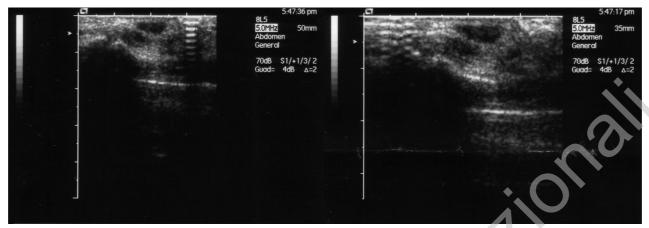


Fig. 2 - Masson's tumor. Ultrasound showing an oval mass with fluid content, referred to a tendon cyst.

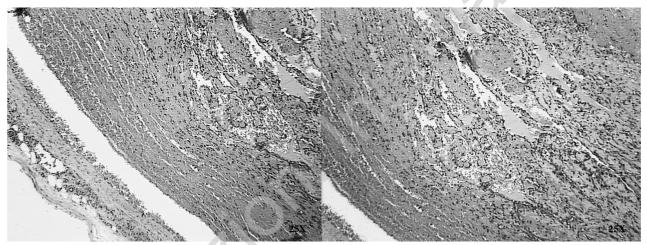


Fig. 3 - Masson's tumor. Standard hematoxylin-eosin staining (magnification x25) shows free-floating papillary projections within the vascular lumen associated with an adjacent thrombus.

veins, rarely within an artery. The secondary or mixed form grows within preexisting vascular structures such as aneurysm, hemangioma, arteriovenous malformation, lymphangioma, and pyogenic granuloma. The extravascular form occurs within hematomas (6).

Differential diagnosis should be carried out against pyogenic granuloma, Kaposi sarcoma, hemangioma, angioendothelioma, Kimura disease, and intravenous atypical vascular proliferation. Masson's tumor can be mistaken for angiosarcoma, which however virtually never occurs within the lumen of a vessel; in contrast, Masson's tumor typically grows and remains within the vessel lumen (5).

Our case of intravascular proliferative endothelial hyperplasia presented macroscopically as a soft tissue

tumor, clinically mimicking a tendon cyst. The main importance of this lesion lies in its possible misdiagnosis as angiosarcoma at the time of histological examination.

Conclusions

Masson's tumor clinically resembles benign lesions such as mucocele, pyogenic granuloma, and hemangioma, as well as malignant neoplasms including angiosarcoma and Kaposi's sarcoma. It can also be mistaken for other conditions, such as a tendon cyst, on ultrasound. For this reason the right diagnosis can only be made by histological examination.

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