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Myopericytoma of the Leg: A Rare Lesion

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Abstract **Case Report**

Myopericytomas are rare, slow-growing benign perivascular tumors most commonly arising within the superficial subcutaneous soft tissues of the lower extremity. They represent one of several related perivascular tumors of myoid lineage with similar morphology and shared immunohistochemical profile including positive staining for smooth muscle actin. Histologically, myopericytoma exhibit concentric, perivascular proliferation of spindledmyoid cells with bland elongated nuclei and associated blood vessels. The present study reports a case of myopericytomas found in the leg and foot of a patient. The masses were surgically excised, and on pathologic and immunohistochemical examination, the diagnosis of myopericytoma was made.

Keywords: Myopericytomas, perivascular tumors, morphology, blood vessels, immunohistochemical examination.

INTRODUCTION

Myopericytoma is a rare type of benign perivascular soft tissue tumor typically presenting with well-circumscribed, slow growing, and painless firm mass [1]. It can occur anywhere in the body, but more common in subcutaneous tissues and dermis of the extremities [2,3] and is often misdiagnosed.It is characterized histologically by the proliferation of oval to plump or spindle-shaped myoid perivascular cells, called myopericytes, which are typically arranged in a concentric and multilayered pattern around the vascular spaces [4]. The tumor is mostly diagnosed based on the histopathologic findings and immunohistochemistry (IHC) staining is done for the confirmations [5]. Standard guidelines regarding the management are still not available, and complete surgical excision of the affected area is the preferred and potentially curative method [6]. This paper reports a case of myopericytoma found in the external aspect of the distal right leg in a 47-year-old male.

CASE REPORT

A 47-year-old male presented to Hospital Mohammed VI in April 2022 with two slowly growing masses on the inner side of the right leg and on the feet which had been growing slowly for a few months. He did not have any relevant genetic history. He is a non-smoker. He was not under any medications. The patient's history was marked by multiple surgical excisions (2012-2013-2014-2017) which histopathological study was in favour of a glomus tumour. His physical examination revealed about 2 cm mass, firm, non-compressible, fixed, painless on the lower third of the right leg associated to a 3 cm mass fixed with no overlying skin changes and only midly tender to palpation.The Magnetic resonance imaging (MRI) showed multiple nodules in the soft tissues of the lower third of the right leg, ankle and feet that are variable in size and shape.

The largest measuring 21x26mm and 48x21mm. The patient underwent a complete monobloc surgical excision of the tumor located at the medial malleolus and of a tumor on the external aspect of the upper 1/3 of the right leg. Histopathologically, the tumor was composed of spindleshaped eosinophilic cells, which showed striking multi-layered, concentric growth around blood vessels. Immunohistochemically, tumor cells showed positivity for smooth muscle actin, caldesmon, and CD34 but they were negative for cytokeratin, S100, and CD31.Based on these findings the diagnosis of myopericytoma was made.

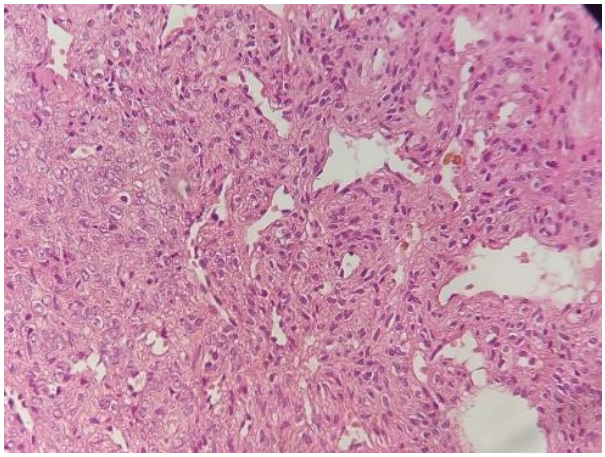

Figure 1: Histological appearance of the concentric perivascular arrangement of lesional spindle shaped cells with around blood vessel walls

DISCUSSION

Myopericytoma is a relatively new disease entity described by Granter and colleagues in 1998 [7]. In the 2013 World Health Organization classification of tumors, myopericytoma is classified into the category of pericytic/perivascular tumor along with glomus tumor, myofibroma, and angioleiomyoma [8]. Myopericytomas are benign solitary tumors, mostly found in middleaged men. They are most commonly seen in the dermis and subcutaneous soft tissue of the peripheral extremities. They can occur in other locations, including proximal extremities, head, neck, lungs, muscle, and bone. They are usually painless, but the intravascular subtype tends to be painful [9,10]. MRI and color Doppler ultrasound show hypervascularity .Excisional biopsy is standard for diagnosis and treatment.

Clinically, myopericytoma most often presents as a solitary, well-demarcated, slow-growing, and non painful palpable nodule or soft tissue mass present for several years [7,4]. Alternatively, less common cases of multiple small clustered nodules histologically appearing as numerous discrete perivascular nodules infiltrating the dermal or subcutaneous tissues have been termed myopericytomatosis by Fletcher and colleagues [8]. Most have an excellent prognosis following complete surgical resection, but local tumor recurrence may occur with incomplete resection [8]. There is currently no recommended role for the administration of chemotherapy or radiotherapy. Although very rare, malignant myopericytoma has been reported. These are typically deepseated and infiltrative tumors with nuclear pleomorphism, brisk mitotic activity, and internal necrosis [4,7]. They are associated with an aggressive clinical course and often distant metastatic disease [12,13].

Pathologically, the tumor cells of myopericytomas are composed of spindle-shaped eosinophilic cells with a striking multi-layered, concentric growth around blood vessels [8]. Immunohistochemically, the tumor cells in myopericytomas are generally positive for smooth actin as a myogenic marker and negative for desmin. cluster of differentiation 34 as a endothelial vascular marker is negative and sometimes positive [4,7,8]. Although this tumor rarely recurs or becomes malignant, Mentzel et al reported local recurrence in 2 of 54 cases [4]. There are some reports of malignant changes [4,11], therefore, longterm follow-up is necessary.

CONCLUSION

Myopericytoma is a soft-tissue tumor that has recently been adopted by WHO as a distinct entity, including a spectrum of myoid tumors of perivascular differentiation. Although often benign, complete surgical resection with clear margin is required to prevent recurrence [4].

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