



ORIGINAL RESEARCH PAPER

Pathology

MYOPERICYTOMA OF THE BASE OF THE MIDDLE FINGER – A RARE DIAGNOSIS AT A RARE SITE

KEY WORDS:

Myopericytoma, base of finger, Immunohistochemistry

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ABSTRACT

Myopericytoma is a soft tissue tumor found in the subcutaneous and superficial soft tissues in the extremities. Myopericytoma represents a benign perivascular myoid tumor, but few rare cases have been found to show malignant transformation and forms a spectrum with myofibroma. It is classified under Perivascular tumors in WHO soft tissue and Bones edition 5th The present study reports a unique case of myopericytomas found at the base of middle finger of the patient. The masses were surgically excised, and on pathologic and immunohistochemical examination, the diagnosis of myopericytoma was made. Myopericytoma is a rare disease entity; however, it is important because it can mimic more ominous conditions

INTRODUCTION

Myopericytoma is a soft tissue tumor found in the subcutaneous and superficial soft tissues in the extremities(1). In the past, cases of myopericytoma may have been diagnosed as hemangiopericytoma or solitary myofibroma. Later in 1998, Granter et al 1st described as a separate subcutaneous and superficial soft tissue lesion of perivascular origin, and only in 2002 it was described as an independent entity under perivascular neoplasms in WHO of soft tissue and Bones(2) . Myopericytoma is a rare disease entity; however, it is important because it can mimic more ominous conditions. The present study reports a unique case of myopericytomas found at the base of middle finger of the patient. The masses were surgically excised, and on pathologic and immunohistochemical examination, the diagnosis of myopericytoma was made.

CASE REPORT

A 64yr old male visited surgery OPD for swelling over his right middle finger since five years. The swelling was situated at the base of the middle finger. The swelling was painless, not associated with any discharge. Patient didn't give any history of trauma or any other such similar swelling anywhere else o the body.

On examination, swelling was 2x1 cm2 in size, non tender, mobile, firm. Overlying skin appeared to be normal.

Ultrasonography of the local part was performed as the initial part of the assessment. Ultrasonography revealed 21x10 mm2 hypoechoic lesion with few foci of calcification with a possibility of a neoplastic etiology .

The patient then underwent surgical excision, and the excised specimen was then sent to the histopathology section.

Gross- The received specimen was 1.5x1x1 cm3 in size, brownish in color, firm in consistency. Its cut surface was whitish and homogenous.

Multiple sections were taken and processed and stained with Hematoxilline & Eosin stain.

Microscopic findings- Multiple sections examined. The sections show well circumscribed, nodular lesion composed of cytologically bland, oval to spindle shaped, myoid tumor cells with multilayered, concentric growth around numerous

small vessels. At places, fascicular arrangement is also seen. At places hypercellular areas and hypocellular areas and collagenous area are seen. Vascular proliferation with dilated, congested and branching vessels are seen. No evidence of Atypia/mitosis/necrosis were seen in sections examined.

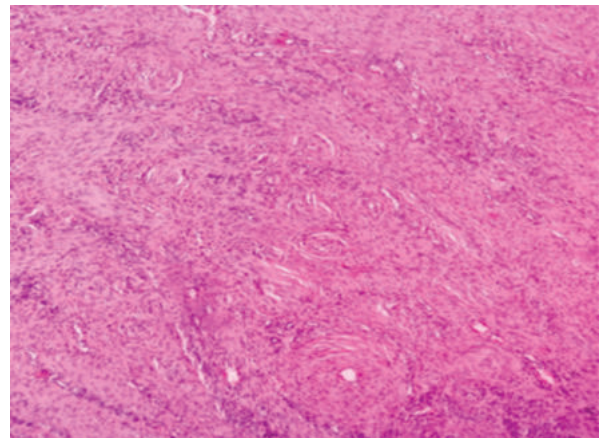


Fig. 1 H&E 10x, myopericytoma contain numerous blood vessels surrounded concentrically by uniform myoid tumor cells

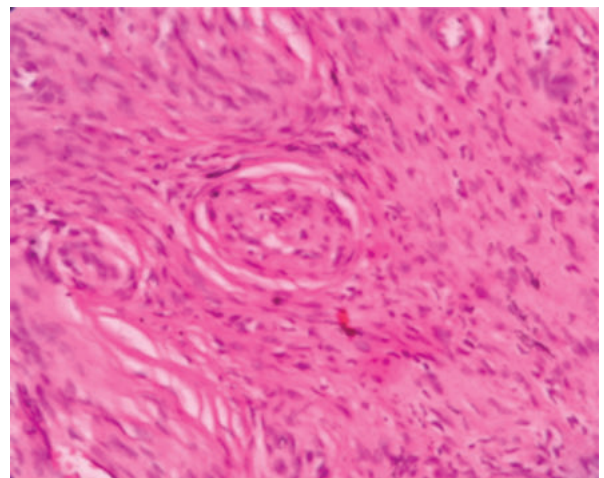


Fig. 2 H&E 40x, blood vessels surrounded by plump spindle myoid tumor cells

We performed Immunohistochemistry analysis, which showed that the tumor cells were immunoreactive for Smooth Muscle Actin and Vimentin, focal positive for desmin and were immunonegative for S-100 and CD34. The final histopathological diagnosis was Myopericytoma a rare and benign smooth muscle tumor.

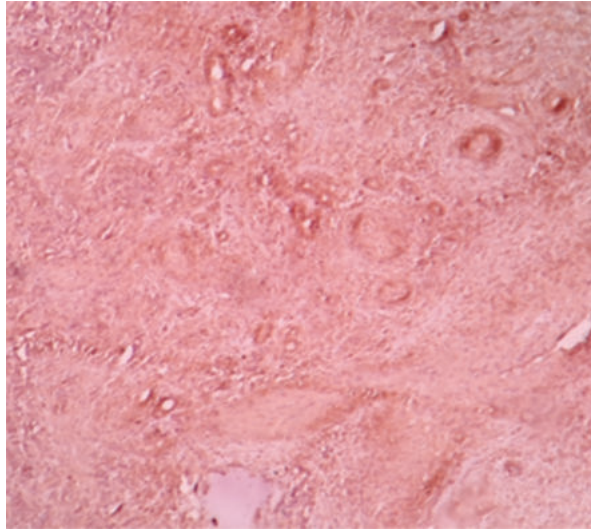


Fig. 3 Immunohistochemical stain for smooth muscle actin (SMA) highlights the concentric perivascular growth of the neoplastic pericytes

Postoperatively, patient had normal sensory and motor functions.

DISCUSSION

Myopericytoma is a rare and a new entity classified under perivascular myoid tumors, traditionally comprising of glomus tumor and its variants, myopericytoma and Hemangiopericytoma like tumor of the nasal passage(3). Histologically, perivascular neoplasms are modified myoid cells that support/ invest blood vessels, i.e. from glomus cells and pericytes.

In the past, myopericytoma have been diagnosed as 'hemangiopericytoma' or 'solitary myofibroma', until it was first described by Granter et al in 1998, with 7 cases as a soft tissue mass located in the subcutaneous and superficial soft tissue of lower extremities(4). In 2002 World Health Organisation classified myopericytomas as an independent perivascular neoplasm of soft tissue(2).

Myopericytoma is mainly a benign perivascular myoid tumor, but few rare cases have been found to be malignant. It develops as a solitary, rarely multiple lesions may involve same anatomical region or different, painless, slowly growing mass , most commonly located in superficially over lower extremities followed by upper extremities. Other sites involve the neck, trunk and oral cavity(3). There is a strong association of the clinical course and the depth of the neoplasms, the metastasizing neoplasms represent deep-seated lesions; in contrast, superficial malignant myopericytoma do not show local recurrence or metastasis(2). Myopericytoma may occur at any age but it is most commonly encountered in adults, median age being 60 years(3).

Etiology of origin of myopericytoma is unclear. An association with EBV in patients with AIDS has been reported(1). Few articles show an association between myopericytoma and trauma(5). Mutations in the PDGFRB gene seem to represent a common pathogenesis for myopericytoma(6)(7).

In our case, histopathological examination was consistent with the diagnosis of myopericytoma being a well circumscribed concentrically arranged myoid cell tumor

cells around small dilated vessels. It has been emphasized that myopericytoma represents a perivascular myoid neoplasm that shares morphologic features with myofibroma, angioleiomyoma, and glomus tumor, but the characteristic concentric perivascular growth of neoplastic cells is typically seen in myopericytoma only (2). Immunohistochemistry further strengthens the diagnosis. The cells were reactive for SMA and Vimentin, which proves its origin of myoid cells while they are immunonegative for CD34 and S- 100, what is of help in the distinction from other perivascular myoid neoplasms as, for instance, myofibroma and angioleiomyoma(2).

Myopericytoma is a rare condition, of which very little is known. A multidisciplinary approach and on going research will be helpful for better understanding of this lesion.

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