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(CASE REPORT)

Fast-growing forearm Myopericytoma, prince Osman Digna Referral Hospital, Port Sudan City, Red Sea State, Sudan; 2025: Case report and review of literature

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Abstract

Background: Myopericytoma is a rare, benign, slow-growing soft tissue tumor that developed from perivascular smooth muscle-like myoid cells (myopericytes) and shares features of both glomus and smooth muscle cells. It usually involves the skin and soft tissues and frequently affects the extremities. It is characterized by the concentric organization of these oval-to spindle-shaped myoid cells surrounding delicate vascular channels, displaying a characteristic concentric perivascular cell proliferation.

Methods: A thirty-four-year-old male teacher from Port Sudan City presented with right posterior proximal forearm swelling for one week. The condition started one week prior to admission with posterior proximal forearm swelling, which rapidly increased in size, was not associated with pain, did not interfere with daily activity, and was not associated with fever or any other lumps in other sites of the body. On examination, there is right posterior proximal forearm swelling about 10×6 cm oval in shape, well demarcated, normal skin over it, no visible dilated veins, there is no hotness or tenderness, with a positive fluctuation and transillumination test, not attached to skin or underlying structure, and there is neither palpable thrill nor bruit. The lump was excised and was sent for histopathology.

Result: The histopathology finding revealed features of a perivascular tumor consistent with myopericytoma.

Conclusion and recommendations: Myopericytoma has a slow-growing course, unlike our case, which is rapidly growing over one week; this gives another manifestation of this tumor which may help to understand its etiology.

Keywords: Myopericytoma; Myopericytes; Fast-Growing; Histopathological Examination; Sudan

1. Introduction

Myopericytoma (MPC) is a rare, benign, slow-growing soft tissue tumor that developed from perivascular smooth muscle-like myoid cells (myopericytes) and shares features of both glomus and smooth muscle cells. It usually involves the skin and soft tissues and frequently affects the extremities. It is characterized by the concentric organization of these oval to spindle shaped myoid cells surrounding delicate vascular channels, displaying a characteristic concentric perivascular cell proliferation [1-13].

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It was first mentioned by Granter and his colleagues in 1998 as a distinct disease entity. They proposed the term "myopericytoma" to describe a group of tumors that show perivascular myoid cell development that mimic both neoplastic and vascular pathologies. In 1992, Dictor et al. defined myopericytes as transitional cells between smooth muscle cells and pericytes in the blood vessels; however, the etiology of MPC is not clear yet [4, 7, 8, 12, 14].

In 2013, the World Health Organization classified myopericytomas as a discrete entity in its classification of soft tissue and bone tumors. They are classified into pericytic tumors showing differentiation towards perivascular myoid cells or myopericytes [3, 4, 5, 10, 11, 14].

MPC has limited information in the literature due to its low incidence. It usually affected males approximately two times more than females. It can present at any age group, but it usually occurs in the middle-aged population, commonly around the fifth decade. It has no specific clinical manifestations but can present as a solitary, distinct, non-painful, slow-growing, well-circumscribed nodule smaller than 2 cm or a soft tissue mass for several years; however, it can also present as multiple nodules. It usually affects the skin and superficial soft tissues of the lower extremities. It is exceedingly rare in visceral organs; where only 11 cases previously reported in the kidney, which have been benign. It is rare to be found in the fingers or within the blood vessels. Intracranial myopericytoma is rare, and up to date there are only nine cases reported in the literature recently. Limited cases of head and neck MPCs were described [1-13, 15].

Myopericytoma (MPC) tumors are usually benign, although limited cases of MPC up to 20% may be malignant with recurrence and distant metastasis [5, 7, 11, 15].

Currently there are no standard management guidelines available for myopericytoma (MPC); however, the diagnosis can be achieved by histological examination. Due to shared morphological characteristics with other neoplasms such as myofibroma, leiomyosarcoma, angioleiomyoma, endometrial stromal sarcoma, infantile hemangiopericytoma, and glomus tumor, MPC can be differentiated by immunohistochemistry, i.e., positive staining for smooth muscle actin, smooth muscle myosin heavy chain, h-caldesmon, and calponin and negative staining for desmin. MPC can also be diagnosed by MRI, where it appears as superficial, well-defined, displaying soft tissues that are well vascularized, often with areas of internal hemorrhage [1, 2, 7, 10, 13, 15].

The gold standard method for treatment is complete surgical excision [1, 7, 12].

2. Case Presentation

A thirty-four-year-old male teacher from Port Sudan City admitted on 3/8/2022, complaining of right posterior proximal forearm swelling for one week. The condition started one week prior to admission with posterior proximal forearm swelling, which rapidly increased in size, associated with no pain, not interfering with daily activity, and not associated with fever or any other lumps in other sites of the body. The patient had no chronic illness, no history of surgical operations, no history of similar conditions, no history of blood transfusion, and no family history of similar conditions. On examination, there was right posterior proximal forearm swelling about 10×6 cm oval in shape, well demarcated, normal skin over it, no visible dilated veins, there was no hotness or tenderness, a positive fluctuation and transillumination test, it was not attached to skin or underlying structure, and there was neither a palpable thrill nor a bruit. Investigations: CBC: (Hb: 12 g/dl, WBCS: $5.4*10^3$, PLT: $234*10^3$); viral screening for HBV, HCV, and HIV was negative. The patient underwent surgical excision of the lump and was sent for histopathology, which revealed features of a perivascular tumor consistent with myopericytoma (figures 1, 2).

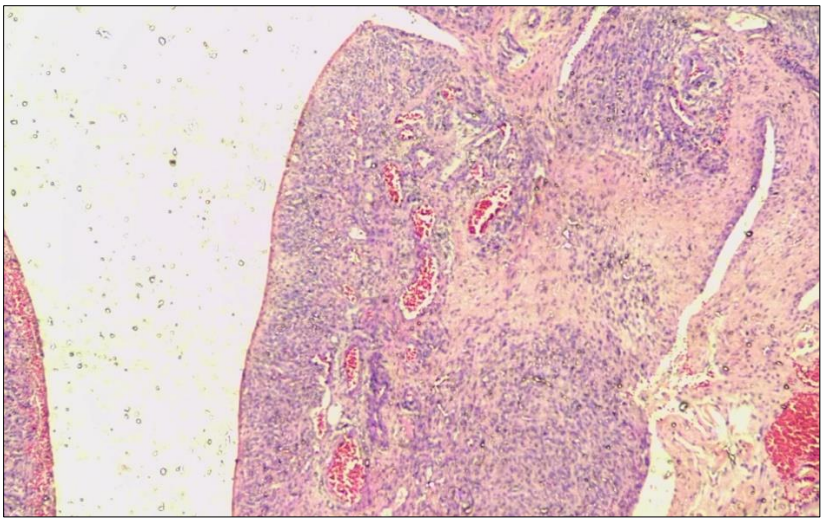

Figure 1 H&E stain: nodular pattern of the tumor, Fast-Growing Forearm Myopericytoma, Prince Osman Digna Referral Hospital, Port Sudan City, Red Sea State, Sudan; 2025

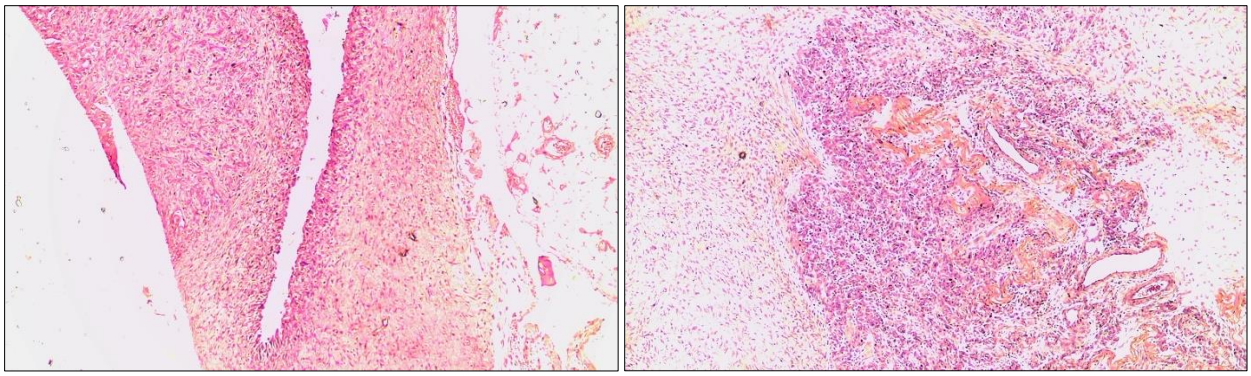

Figure 2 H&E stain: slides showing blood vessels surrounded by proliferating pericystic cells with myoid features, Fast-Growing Forearm Myopericytoma, Prince Osman Digna Referral Hospital, Port Sudan City, Red Sea State, Sudan; 2025

3. Discussion

Myopericytoma (MPC) usually affects middle-aged individuals and is more common in males than females, like our case [4, 7, 9]. Unlike the international studies, our case represents a rapid growth myopericytoma (MPC) [1, 4, 13]; instead, it was a painless, solitary, well-demarcated nodule within the subcutaneous tissue of the forearm, like many studies [1, 5, 7, 8, 9, 13]. Although there are no standard management guidelines available for MPC, the gold standard method for treatment is complete surgical excision, like in our case [1, 7, 12]. This case is diagnosed by histopathological examination that revealed benign myopericytoma, which is consistent with the global reports [1, 2, 4, 6, 11, 13, 15].

4. Conclusion

Myopericytoma is a rare soft tissue tumor usually benign with a slow-growing course, unlike our case, which is present with rapid and fast-growing over one week; this gives another manifestation of this tumor that may help to understand its etiology.

Compliance with ethical standards

Disclosure of conflict of interest

There is no conflict of interest

Statement of ethical approval

The article describes a case report. Therefore, no permission from ethical committee was required. Informed consent was obtained from the patient for publication of this case report.

Statement of informed consent

Informed consent was obtained from the patient for publication of this case report and accompanying images.

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