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Myopericytoma in right inguinal region: A rare case report from Nepal

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ABSTRACT

INTRODUCTION: Myopericytoma is a rare mesenchymal soft tissue tumor that originates from perivascular myoid cells and mostly benign in nature. With a slight male predisposition, it occurs more commonly in the lower extremities with an insidious and painless presentation. Contrary to the usual, our patient was with a rare presentation in the inguinal region with a dull aching pain.
CASE PRESENTATION: We herein report a case of a 64-year-old female patient with myopericytoma, the first of its kind reported in Nepal, present in the right inguinal region and surgically removed. The mass was single with dull aching pain, non-tender, globular, smooth, mobile, covered with skin and, had been slowly growing for 1 year and had started bleeding for 4 days. On complete surgical excision and subsequent biopsy, a nodule with thin-walled blood vessels and spindle-shaped cells having indistinct cell border, eosinophilic cytoplasm, and oval to spindle-shaped nucleus was observed, features consistent with those of myopericytoma.
DISCUSSION: Myopericytoma can occur in different sites in the body. Its differential diagnoses in the inguinal region include inguinal hernia, lipoma, or an inguinal lymphangioma. Histopathological analysis and immunohistochemistry (IHC) staining are used for diagnosis and confirmation.
CONCLUSION: Myopericytomas are rare and have a substantial propensity to be misdiagnosed as other soft tissue tumors such as sarcomas due to significant overlap in their presentation and histological features. Complete surgical excision of mass is the curative therapy.

1. 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]. 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]. Myopericytes are said to have intermediate features of pericytes and vascular smooth muscle cells. This tumor was first described in 1998 by Granter and colleagues, using the term myopericytoma to comprehensively describe the spectrum of three different tumors: myofibromatosis, typical myopericytoma, and glomangiopericytoma [5]. The tumor is mostly diagnosed based on the histopathologic findings and immunohistochemistry (IHC) staining is done for the confirmations.

Herein, we present the first reported case of myopericytoma from Nepal, which was present in the right groin in a 64-year-old female patient and was surgically excised. The case has been reported in line with SCARE 2020 criteria [6].

2. Case report

A 64-year-old Hindu woman from the remote part of central Nepal presented to the surgical outpatient department of Tribhuvan University Teaching Hospital with a slowly growing, skin-covered gray-brown mass in a right inguinal region for approximately 1 year and bleeding from the mass for 4 days. The swelling was associated with dull aching pain. The swelling later ulcerated, which resulted in bleeding for 4 days. The patient did not complain of fever, weight loss, and presence of other similar lumps. The patient does not have any chronic comorbidities. She did not have any relevant past medical history and relevant genetic history. She is a non-smoker. She was not under any medications. On examination, a single, non-tender, globular, smooth, mobile mass, measuring 7.5 cm × 6.0 cm, with bleeding from the tip was found on the right inguinal region. Mass was non-pulsatile and impulse on coughing was absent.

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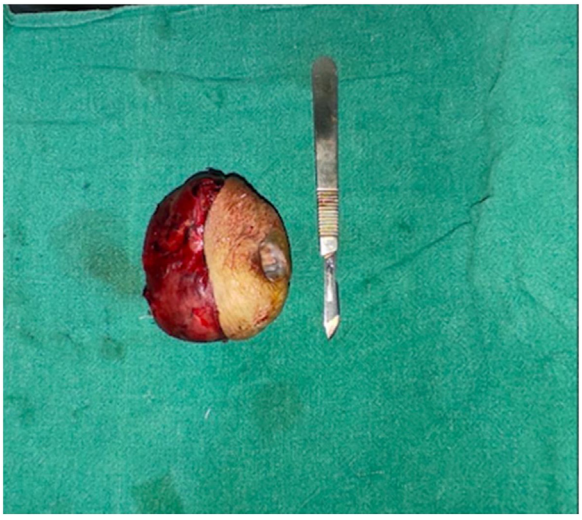

Fig. 1. Gross appearance of surgically excised mass from right inguinal region.

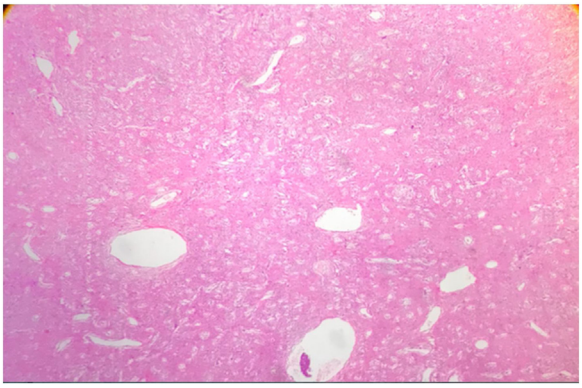

Fig. 2. Microscopic image of biopsy specimen showing variable sized blood vessels and spindle cells at magnification 4.

Ultrasound showed heterogeneous echogenicity mass with irregular margins and slightly increased vascularity.

The mass was surgically excised in a minor operating room by a general surgery resident and the specimen was sent for biopsy as shown in Fig. 1. It was excised under injection of local anesthesia solution of lidocaine and epinephrine. After the excision of the mass, the excision site was sutured using polyglactin and nylon suture. Once the wound was closed, a gauze was rolled over the length of the wound to express any blood if residual bleeding or hematoma were present. The dressing was applied over the wound after the application of topical ointment. There were no complications during the surgical intervention. She was instructed for wound care including cleaning of wound and replacement of dressing.

On histopathologic examination, microscopically, the dermis showed a nodule composed of thin wall variably sized (small, slit-like to cystically dilated) blood vessels and spindle-shaped cells arranged in short fascicular arrangement. Thick-walled blood vessels were also observed. Spindle-shaped cells had an indistinct cell border, eosinophilic cytoplasm and oval to spindle-shaped nucleus. Focal myxoid change was observed. In some areas, cells had myoid appearance. Mitotic figures were not found. The overlying epidermis was acanthotic and focally ulcerated. Cholesterol clefts were also seen. All these histological findings are shown in the histological picture in Figs. 2–4. The features were consistent with that of myopericytoma.

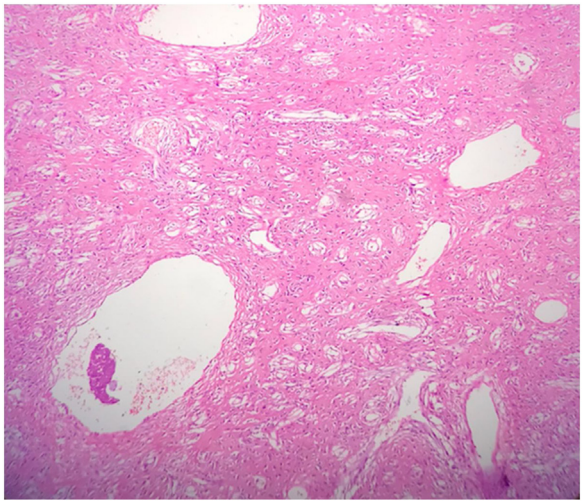

Fig. 3. Microscopic image at magnification 10.

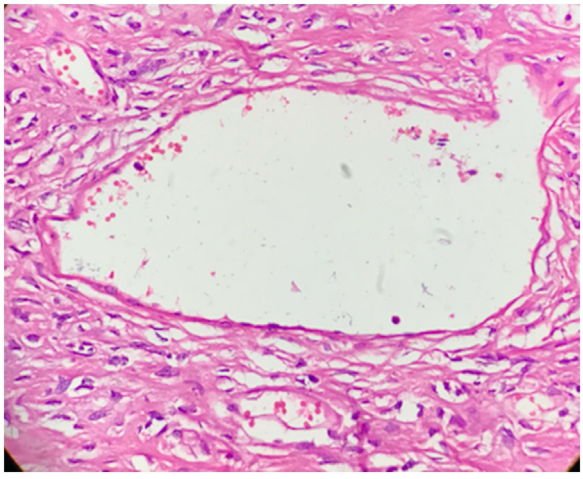

Fig. 4. Microscopic image at magnification 40.

Our patient was discharged on the same day of the surgical procedure. She is on close follow-up. There is no recurrence at the excision site on physical examination. No diagnostic methods were used for the assessment of recurrence. The patient is satisfied with the treatment she received.

3. Discussion

Myopericytoma is a relatively rare mesenchymal soft tissue tumor that primarily originates from subcutaneous tissues [5]. It is a part of the spectrum of diseases that constitute tumors arising from the perivascular myoid cells, also known as pericytic or perivascular tumors [5]. The recent WHO Classification of Soft Tissue tumors classifies perivascular tumors as benign or malignant, of which myopericytoma belongs to the benign group [7]. While earlier classifications listed MPC and MF as different entities, the latest WHO classification has included myofibroma as a part of myopericytoma [7]. The more diffuse variant of myopericytoma is a condition that presents with multiple microscopic nodules constituted of cells of perivascular origin. This condition has been termed as myopericytosis [8]. Myopericytomas are rarely malignant [9].

Myopericytoma presents over a course of several months to decades in the dermis or subcutis, more commonly in the lower extremities. However, these tumors have also been reported to arise from different visceral organs including the nervous system,

lung, liver, kidneys, urinary bladder, and stomach [2]. Although myopericytomas have been thought of having a greater male preponderance; myopericytomatosis has a slightly higher female predominance [10]. Clinically, most myopericytomas have been thought to be painless. Unlike the majority of cases, the aforementioned patient presented with dull aching pain of the swelling in the right inguinal region. Patients with painful myopericytomas have been reported, more commonly in Japan [11]. The primary differential diagnoses of a mass in the inguinal region would include a hernia or other more common swellings like lipoma; however, ulceration and bleeding from the surface of the skin overlying the swelling is highly unlikely of these conditions.

Diagnostic imaging of this patient did not corroborate with the findings that are anticipated in cases of myopericytoma as these tumors are typically well-defined, homogenous, and highly vascular [9]. The imaging findings of this patient are more consistent with that of myopericytomatosis, in which diagnostic imaging reveals a more heterogenous mass with ill-defined swelling [9]. Although immunohistochemical tests were not performed, one would normally expect a positive stain for MSA (muscle specific actin), SMA (smooth muscle actin), and h-caldesmon in myopericytoma cells. Myopericytomas are sometimes immunoreactive to desmin [5].

Surgical resection is the treatment of choice in these tumors because of excellent prognosis and also because the definitive diagnosis requires histopathological affirmation. Although this patient had features that favoured a diagnosis of myopericytomatosis, the presence of a single nodule on microscopy confirmed the diagnosis of myopericytoma. Furthermore, the absence of mitotic figures and cellular atypia made the diagnosis of malignant myopericytoma unlikely.

4. Conclusion

Owing to a diagnostic dilemma due to its morphological overlap with certain other soft tissue tumors such as sarcomas and because of its clinical presentation, myopericytomas are frequently misdiagnosed. It is one of the differential diagnoses of the lump in the inguinal region. These are mostly benign and can be cured by complete surgical excision of the lesion.

Declaration of Competing Interest

The authors report no declarations of interest.

Sources of funding

There are no any sources of funding.

Ethical approval

Case reports are exempt from ethical approval in our institution, Tribhuvan University Institute of Medicine, Maharajgunj.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Authors contribution

All the authors contributed equally for the preparation of this case report. Author Naveen C. Bhatta was the consulting Doctor for our patient in outpatient department, and he was the surgeon who surgically resected the mass. He provided the photograph of surgically resected mass and he was also involved in preparation of manuscript. Authors Kshitiz Acharya and Navin Poudel collected all the informations from the patient, asked for written consent and also collected the histopathological report and microscopic slides photographs from pathology department of Tribhuvan University, Institute of Medicine. Authors Aramva Bikram Adhikari, Subarna Giri and Kanchana Bali were involved in the review of previous literatures and preparation of manuscript. All the authors individually did final proofreading of the manuscript before submission.

Registration of research studies

Not applicable.

Guarantor

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