Case Report

Establishing Rare Diagnosis of Myopericytoma of The Foot

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Abstract

Myopericytoma is a benign tumor that consists of oval to spindle-shaped myoid-like cells with a strong proclivity for concentric perivascular growth. Due to the mild nature of the disease, myopericytoma is often difficult to diagnose and might be overlooked. We reported a 59-year-old female who presented severe and persisting pain in the fourth toe in the last 4 years. The diagnosis is not considered initially due to clinical symptoms of pain without any significant growth of the toes hence the patient was initially diagnosed with ganglion cyst. However, further, a combination of clinical, radiological, histopathological, and immunohistochemical examination concludes the diagnosis of myopericytoma.
Keywords: *myopericytoma, histopathological, immunohistochemical.*

Menegakkan Diagnosis Langka *Myopericytoma* di Kaki

Abstrak

Myopericytoma adalah tumor jinak yang terdiri dari sel-sel seperti myoid berbentuk oval hingga gelendong dengan kecenderungan kuat untuk tumbuh secara perivaskular konsentris. Karena sifatnya yang ringan, myopericytoma seringkali sulit didiagnosis dan mungkin diabaikan. Kami melaporkan seorang wanita berusia 59 tahun yang mengalami nyeri parah dan terus-menerus pada jari kaki keempat dalam 4 tahun terakhir. Myopericytoma pada awalnya tidak dipertimbangkan pada kasus ini melihat manifestasi klinis nyeri tanpa diikuti perubahan ukuran jari kaki sehingga pasien didiagnosis kista ganglion. Namun melalui pemeriksaan lebih lanjut, yakni kombinasi pemeriksaan klinis, radiologi, histopatologi, dan imunohistokimia disimpulkan diagnosis mioperisitoma.
Kata kunci*: myopericytoma, histopatologis, imunohistokimia.*

Introduction

Myopericytoma (MP) is a noncancerous neoplasm composed of myoid-like cells that exhibit a pronounced inclination toward concentric perivascular expansion and range in shape from elliptical to spindle. In 1998, Granter et al. were the first to identify it; they examined seven specimens of the soft tissue mass which located in the superficial and subcutaneous soft tissues of the distal extremities.[1] A well-defined, unencapsulated nodular proliferation characterized by numerous thin layered wall capillaries and a concentric, perivascular configuration of spindle-shaped ovoid myopericytes is frequently observed in soft tissues, subcutis, or the dermis. This condition is prevalent in adults. While benign myopericytomas predominate, instances of malignant myopericytomas have been documented.[1–3] The natural history of malignant MP lesions has been described in only one case series and one report, which is an exceedingly low incidence rate. The largest series comprised five cases, of which one was situated in the mediastinum, three in the neck, and one in the lower extremities.[4,5]

Regarding differential diagnosis, MP tumors pose a distinct challenge for surgeons and may be easily missed. This case report describes a 33-year-old male who is predominantly right-handed and visited our outpatient clinic with a painless swelling and redness of the pulp of his left index finger that had been present for two months. The patient exhibited no other signs of illness or medical conditions. Subsequently, a conventional X-ray showed that the distal phalanx had suffered extensive damage, resulting in near-total destruction of the bone tissue. A non-malignant form of MP was identified through the process of histopathological examination of an incisional biopsy. After a span of 81 days following the operation, there was no discernible evidence of the lesion recurring. The treatment involved the surgical removal of the tumor, followed by the careful separation of the tumor from surrounding tissues. A comprehensive literature review is suggested to be carried out on the management and outcomes of all documented cases of hand and wrist MP treatment. This document provides a reference for a case involving myopericytoma located in the neck. A 23-year-old woman detected a non-painful small mass in the supraclavicular fossa on her left side. Following a duration of five months, the patient's mass exhibited persistent growth, prompting them to seek medical attention. No impairments in motor or sensory functions were detected. Imaging indicated the existence of a mass that seemed to originate from the middle scalene muscle. A core needle biopsy, guided by computed tomography, detected the existence of a spindle cell neoplasm with smooth muscle differentiation. The surgical procedure involved a total removal. Myopericytoma was suspected based on the findings from the histopathological and immunohistochemical examinations of the tissue sample. A myopericytoma is a rare tumor that primarily occurs in the head and neck region. As far as we know, this is the first recorded case of a myopericytoma originating from a scalene muscle.

Most myopericytomas present as indolent, painless tumors in either the deep or superficial soft tissues. Myopericytoma commonly presents as a solitary tumor, predominantly in the soft tissue of the lower limb. Myopericytoma, despite its mild nature, can present diagnostic challenges and may be overlooked or dismissed.Four Typically, a thorough examination of the pathology and immunohistochemical features is required to differentiate the mass from other masses that exhibit similar characteristics. Based on clinical, radiological, and histopathological evaluations, we have identified that the case described in this report is a myopericytoma affecting the fourth toe.

Case Presentation

A 59-year-old female presented severe and persisting pain in the fourth toe in the last 4 years. The pain firstly presented on the left foot with intermittent nature. The pain was aggravated with walking and relieved with rest. The pain intensity and duration increased over time. The patient had tried physiotherapy but showed no significant improvement. The patient did not observe any lump in the area. Past medical history is not significant with no signs of infection. The patient had no history of another tumor. The patient had no prior treatment such as alternative medicine, herbal nor massage manipulation.

On physical examination of the plantar, it showed a mass on the 4th toe with no signs of venectation, wound, deformity, erythema, edema nor exudation (Fig.1). On palpation, it was felt on the plantar side with circumference length of 7cm compared to 6.5cm on the contralateral side. Tenderness was felt with visual analogue score (VAS) of 3-4. Distal sensory and capillary refill time was normal. Range of motion showed some limitation due to pain. A plain radiological examination showed no fracture or any bone abnormalities (Fig. 2). Ultrasonography was then conducted with a solid mass with a cystic component with a benign characteristic that suggested a benign tumor (Fig. 3). Laboratory examination showed no significant abnormalities.

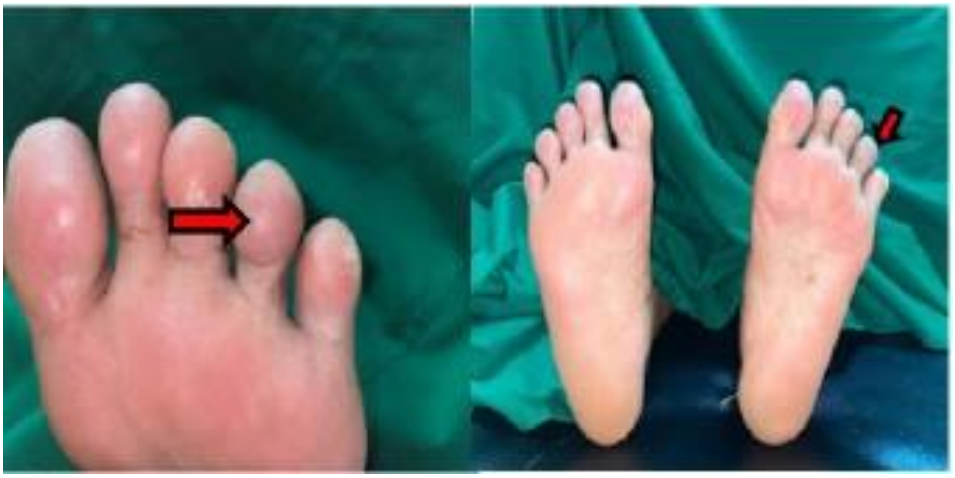

Figure 1. Clinical Picture of the 4th Toe with The Tumor on The Plantar Side (Arrow)

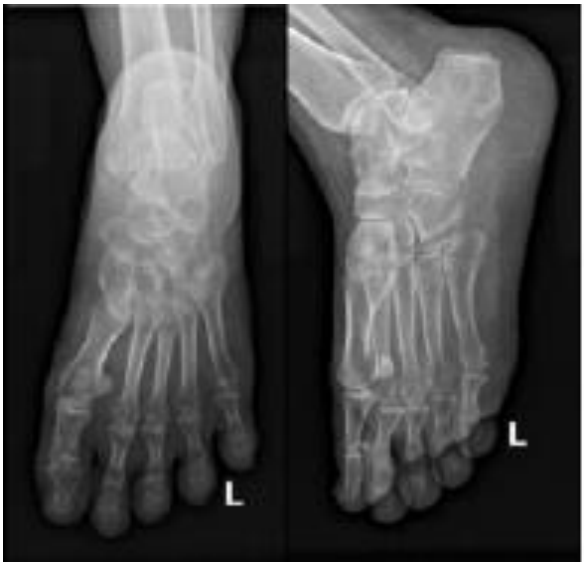

Figure 2. X-Ray of the Left Foot

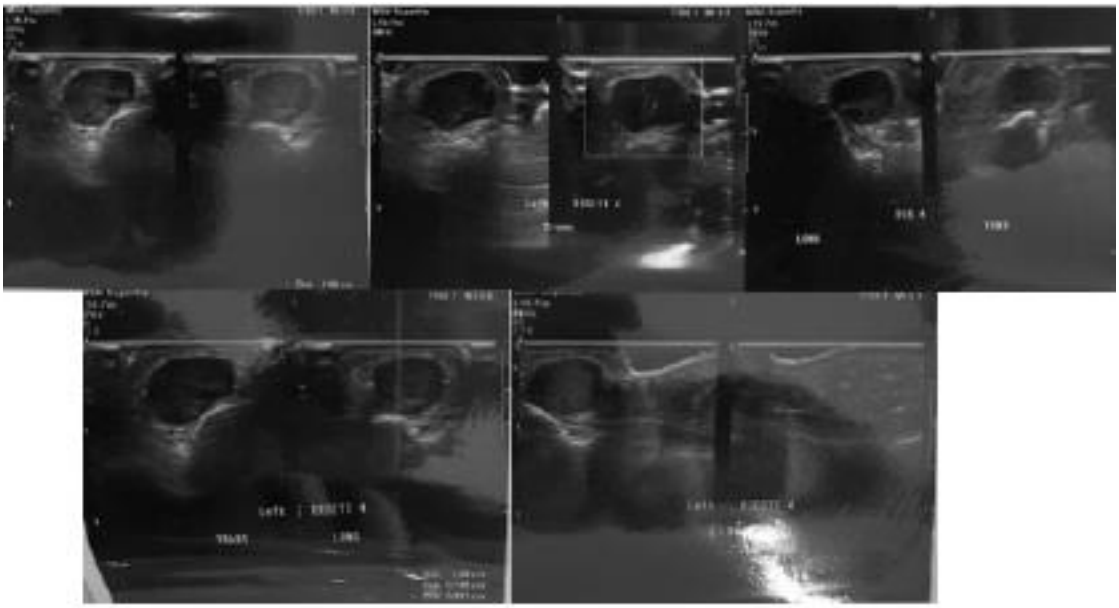

Figure 3. USG Examination

Following this, the patient was transferred to the operating room due to severe and persistent pain in the lesion. We performed a wide excision of the tumor (Fig. 4). Longitudinal incision was made on the overlying tissue on the plantar aspect of the 4th toe. Soft tissue dissection was conducted, and the tumor was exposed. Excision of the tumor was then conducted with pathological examination following the excision (Fig. 5). Pathologic examination suggests the diagnosis of myopericytoma. Immunohistochemical analysis was then conducted that further supported the diagnosis of myopericytoma.

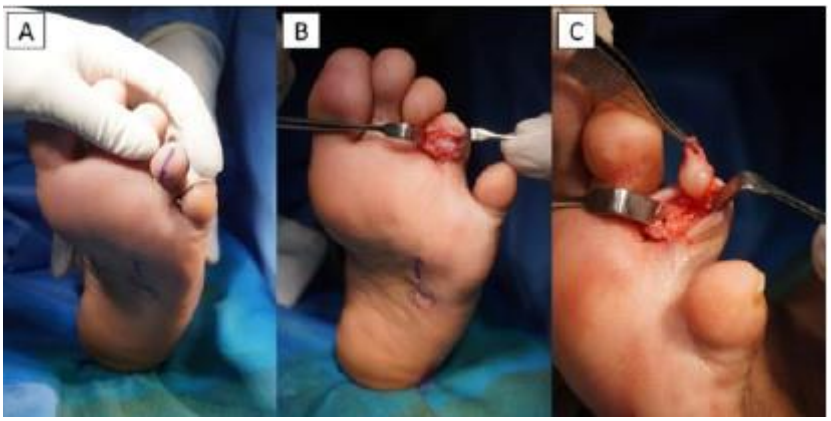

Figure 4. A. Drapping and Design; B. Tumor Exposed; C. Tumor Removal

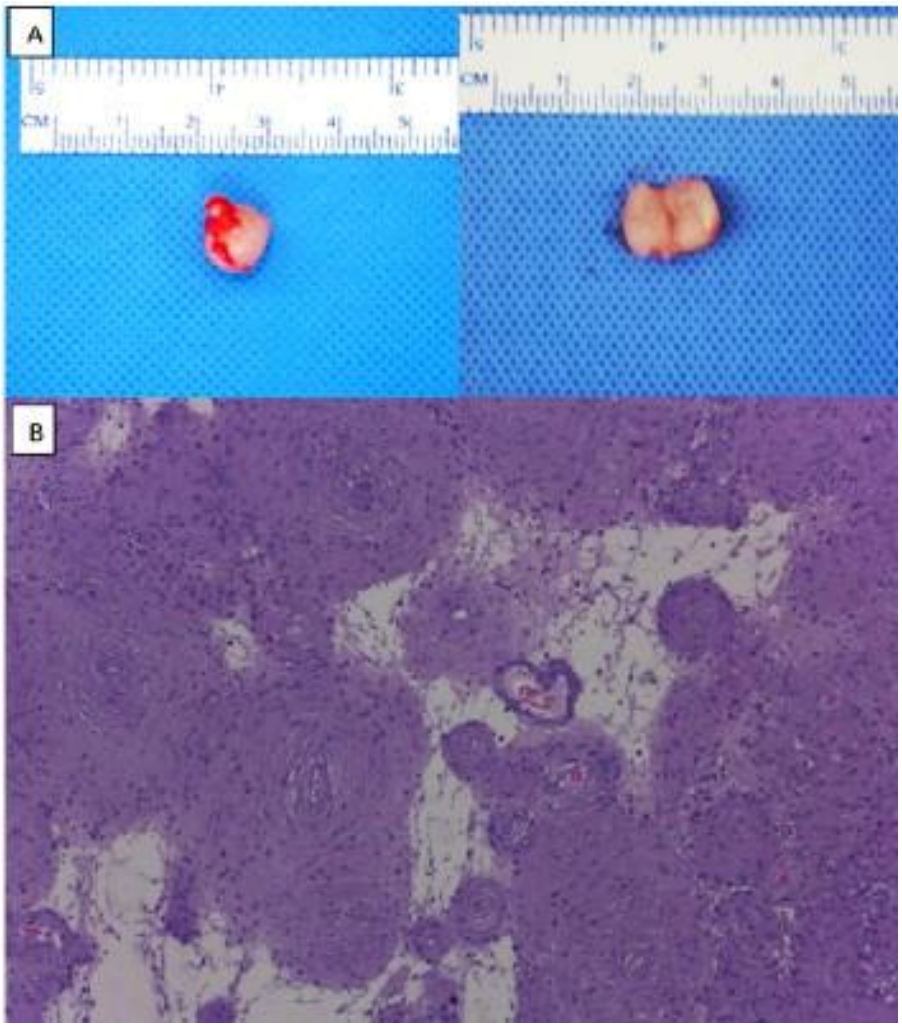

Figure 5. A. Macroscopic Excised Tissue1.2x1x0.5 cm; B. The Histologic Features are Most Consistent with Myopericytoma

Discussion

Myopericytomas are aggregates of soft tissue that originate from myopericytes and exhibit pericyte and vascular smooth muscle cell characteristics. As of 2002, the World Health Organization classified myopericytomas as distinct perivascular tumors of bone and soft tissue. A prevalent and conspicuous characteristic is the concentric arrangement of tumor cells encircling vascular channels, which imparts an onion-ring aesthetic. Lubricated and capable of invaginating into the lumina of intralesional blood vessels, small foci comprised of spindle cells containing copious amounts of eosinophilic cytoplasm were encased in a myxoid matrix arranged in fascicles or whorls.[6] Typically, they manifest as benign nodules within the subcutaneous tissues located in the distal extremities. The mechanism of myopericytomas are still unknown. However, some studies associated myopericytomas with trauma and chronic scars.[7] As mentioned in this case, they affect people of all ages, with a peak occurrence at the age of 50.

Lower extremities were the most commonly affected, followed by upper extremities including digits, hands, and arms. However, myopericytomas of the foot and ankle are uncommon.[6] While the epidermis and subcutaneous soft tissues comprise the majority of lesions, prior studies have documented instances of myopericytomas in various anatomical sites, including the oral cavity, thoracic spine, lung, and kidney.[7] Although malignant myopericytomas are uncommon, a previous study has reported three cases of extremity malignant myopericytomas, two of which presented as large, painful deep masses with infiltrative growth and metastasis. The clinical prognosis of rare malignant myopericytomas appears to be significantly correlated with the depth of the neoplasm, according to a previous study.[6]

In this case, we presented a case with a painful nodule in the plantar aspect of the toes with progressively increasing pain. Painful nodules are the most common symptom of unusual intravascular myopericytomas in the extremities.[8,9] However, the plantar position of the lesion could lead to frequent mechanical trauma that leads to pain. A previous study reported similar symptoms with myopericytoma in the plantar area with symptoms of intermittent pain and discomfort.[1,8] In all cases, the symptoms of myoperocytoma are so general that they were not unique to be considered in the initial differential diagnosis.[7] Thus, supportive measures should be conducted to further conclude the diagnosis and plan the treatment.

Both X-ray and ultrasonography (USG) were conducted, which identified the possible existence of a soft tissue tumor. An ultrasound examination will identify a distinct hypoechoic abnormality that can be distinguished from the surrounding tissues. The ultrasound can occasionally detect the hypervascular features of the lesion. It is crucial to obtain a traditional x-ray because the lack of skeletal alterations suggests that the mass is benign rather than malignant.10 to 12 The X-ray analysis revealed no anomalies upon inspection. Noting the absence of phleboliths is crucial for distinguishing highly vascular lesions. Although MRI is utilized, distinguishing this neoplasm from malignant lesions such as sarcoma remains a difficult task. Myopericytomas generally exhibit similar signal intensity to muscle on T1-weighted images and higher signal intensity than fat-saturated and T2-weighted images. Administering gadolinium results in a significant enhancement of contrast, indicating the highly vascular characteristic of this abnormality. The numerical values 7 and 10. Digital subtraction angiography can be utilized to enhance the visualization of the vascular characteristics of the lesion, evaluate its vascular status prior to surgery, and aid the surgeon in preoperative planning. Usually, lesions are removed through surgical excision, and the chances of them coming back are minimal.

A histopathological examination should be able to conclude the diagnosis of myopericytoma. Nevertheless, the pathological differential diagnosis for myopericytoma comprises myofibroma, angioleiomyoma, and glomus tumor, as a result of histologic overlap.[7] The presence of elongated, dilated blood arteries that are surrounded by spindle-shaped tumor cells with abundant eosinophilic cytoplasm in a concentric manner are the character of Myopericytomas. In contrast to myopericytoma, the prevalence of zonation or a biphasic appearance is indicative of myofibroma. Angioleiomyomas predominately consist of thick-walled vessels and exhibit a desmin-positive immunohistochemical profile. The majority of myopericytomas, on the other hand, are desmin-negative and have thin-walled vessels. Glomangioma cells are perivascularly organized but do not contain spindle cells.[7,13] However, while a concentric arrangement of cells accentuates blood vessel walls in MP, it is not seen in glomus tumors. Furthermore, glomus tumors lack areas with spindle cells and abundant eosinophilic cytoplasm, which mimic myofibroma.[2,6]

The immunohistochemical examination is essential for making a definitive diagnosis of myopericytoma. The spindle cells in the myopericytomas displayed extensive expression

of SMA, but did not exhibit any reactivity to CD34. Furthermore, the absence of desmin labeling on the tumor cells suggests that their smooth muscle phenotype was not fully developed. Furthermore, it was noted that cytokeratin and S100 protein, typically present in nodular hidradenomas, exhibited negative findings.[6] Myopericytomas commonly display vimentin, smooth muscle actin, muscle-specific actin, and sometimes h-caldesmon immunohistochemical markers. However, these proteins, namely S-100 protein, P-53 protein, and CD99, are typically not detected.[7] We used immunohistochemical analysis to conclusively diagnose myopericytoma in this specific case.

The majority of myopericytomas are benig n tumors that are treated with complete excision with a clear margin. Incomplete tumor excision causes recurrences despite the low rate of recurrence in complete excision.[14,15] We believe that surgical excision is adequate treatment for benign myopericytoma, which was diagnosed pathologically, despite the fact that we cannot make a conclusive statement due to the short 5-month follow-up period for our patient.6 From the case, we learnt to be more aware of other differentials of chronic plantar pain, moreover the insidious nature of myopericytoma makes it challenging to diagnose it early over the disease course.

Conclusion

The diagnosis of myopericytoma should be cosidered through clinical and radiological examination and should be confirmed by the immunohistochemical examination.

Conflicts of Interest

There is no conflict of interest in this case.

Acknowledgments

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