Myopericytoma presenting as a painful dark subungual discoloration

Sofia Antunes-Duarte[1], Marta Aguado Lobo[1], Pablo Espinosa Lara[1], Ana Fraga[1], Luís Soares-de- Almeida[1,2]

Affiliations: [1]Dermatology Department, Hospital de Santa Maria, Centro Hospitalar Universitário Lisboa Norte, E.P.E., Lisbon, Portugal, [2]Unit of Research in Dermatology, Institute of Molecular Medicine, Faculty of Medicine, University of Lisbon, Portugal

Corresponding Author: Luís Soares de Almeida, Hospital de Santa Maria–Serviço de Dermatologia Piso 5, Avenida Prof. Egas Moniz, 1649-035 Lisboa, Portugal, Tel: 351-217805000, Email: soaresdealmeida@chln.min-saude.pt

Abstract

Myopericytoma is an uncommon benign neoplasm that arises from the perivascular myoid cells. It typically presents as a painless well-circumscribed cutaneous or soft-tissue nodule, most commonly on the extremities of adults. Histologically, it is characterized by spindle-shaped myoid-appearing cells with a concentric arrangement in vessel walls, that are immunoreactive to alpha-smooth muscle actin and often for h-caldesmon, but negative for other smooth muscle markers. Herein, we present an unusual case of a painful subungual myopericytoma presenting as a dark subungual discoloration.

Keywords: myopericytic, myopericytoma, tumor

Introduction

Myopericytoma (MP) was first described by Granter et al. in 1998 as a benign tumor showing a myoid/pericytic line of differentiation [1]. It arises most commonly in the dermis or subcutaneous tissue of the extremities in adults and typically presents as a well-circumscribed, slow-growing, painless firm mass [2]. In most cases, MP behaves in a nonaggressive manner, with rare recurrence following excision [3]. Expanding the clinical spectrum of this rare tumor, we report an unusual case of a painful subungual MP presenting as a dark macule and review its histopathological and immunohistochemical features.

Case Synopsis

An apparently healthy 59-year-old man, presented to our outpatient department with a 5-year history of a dark subungual macule on the third finger of the left hand. He denied a prior history of trauma. The lesion was initially asymptomatic, but he described local pain during the last months.

Upon physical examination, a 9×4mm sized dark macule was observed on the ulnar aspect of the subungual region of the third finger of the left hand (**Figure 1**).

A clinical diagnosis of glomus tumor was suspected and an exploratory surgical procedure was

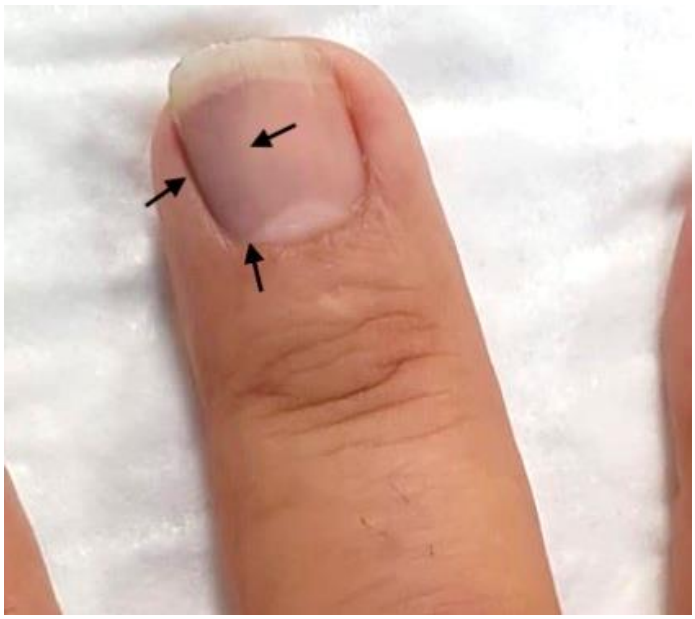

Figure 1. *Subungual myopericytoma, clinical photographs: on the third finger of the left hand a dark-blue macule over the ulnar aspect of the subungual region is observed.*

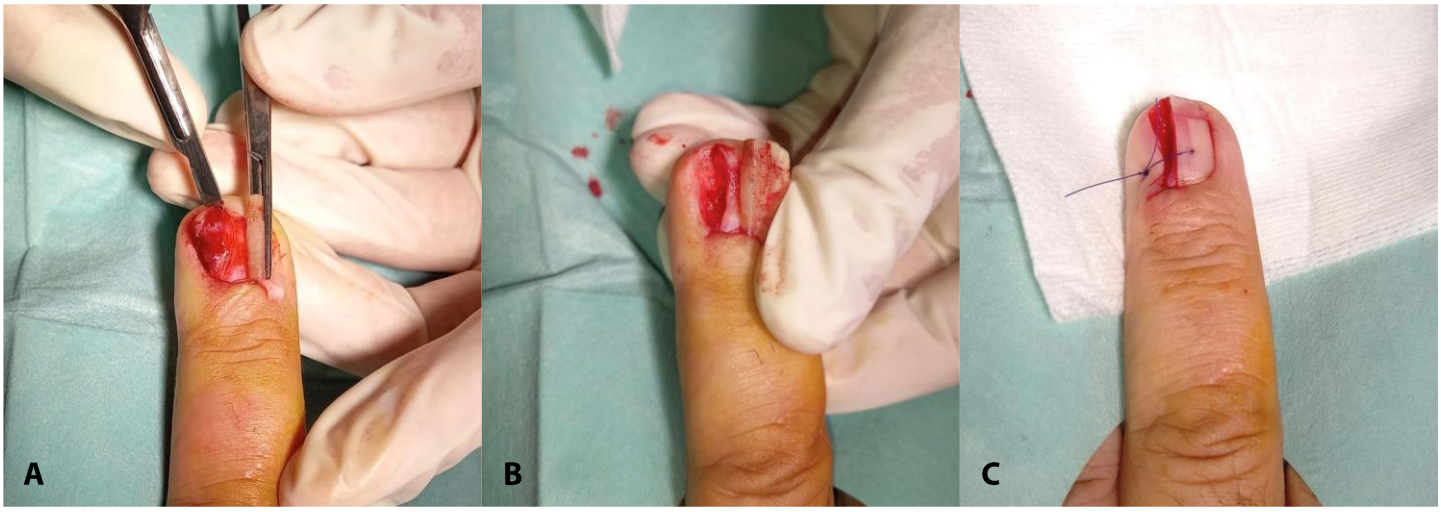

Figure 2. *Intraoperative photographs of subungual myopericytoma excision.* ***A)*** *Partial removal of the nail plate revealing a polypoid tumor emerging from the nail matrix.* ***B)*** *After complete resection of the tumor.* ***C)*** *Immediate post-operative photograph.*

performed. A careful partial avulsion of the nail plate revealed a polypoid tumor emerging from the nail matrix that was completely excised (**Figure 2**).

Histopathologic examination showed an unencapsulated, well-circumscribed dermal proliferation of plump, spindled myoid cells arranged concentrically around vessels (**Figure 3A-C**). Cytologic atypia and mitoses were absent. Immunohistochemical stains revealed diffuse and strong staining of the lesional cells with α-smooth muscle actin. The tumor was focally positive with h-caldesmon and CD34 for tumor cells around the lesional vessels (**Figure 3D-F**). These findings were consistent with the diagnosis of MP.

There was no evidence of recurrence during five months of follow-up.

Case Discussion

Myopericytoma is a very rare benign tumor with fewer than 200 cases described in the literature [4]. It is believed to represent a distinct clinicopathological entity within the spectrum of perivascular neoplasms traditionally comprising glomus tumors and hemangiopericytomas. They are composed of oval to spindle-shaped myoid-appearing cells with a striking tendency for concentric perivascular growth [2,5]. Although the etiopathogenesis of MP remains unclear, cases have been reported after local trauma or associated with acquired immunodeficiency syndrome and Epstein Barr virus infection [4].

Myopericytoma affects mainly adolescents and young adults and a male predilection has been reported consistently [5]. Classically, it presents as a single subcutaneous well-circumscribed nodular tumor mainly located on the extremities, with only rare cases of multicentricity [5]. Myopericytoma is usually asymptomatic or only painful when manipulated and grows slowly but progressively [5].

We found a single case of subungual MP reported by Boix-Vilanova et al. in 2020, which presented as an asymptomatic hyperkeratotic lesion on the pulp of the finger (**Table 1**), [4]. To the best of our knowledge, our case is the first reported case of a painful subungual MP, presenting as a dark subungual macule, leading to the main clinical suspicion of a glomus tumor. Differential diagnosis of black discolorations of the nail bed is broad, including benign lesions, like subungual hemorrhage and benign melanocytic pigmented lesions. Malignant tumors include subungual squamous cell carcinoma and melanoma [6]. Therefore, when clinically indicated, a nail bed biopsy should be performed to ensure the correct diagnosis and allow prompt treatment or reassurance [6]. In this case, an exploratory surgical procedure revealed a polypoid tumor that was completely excised.

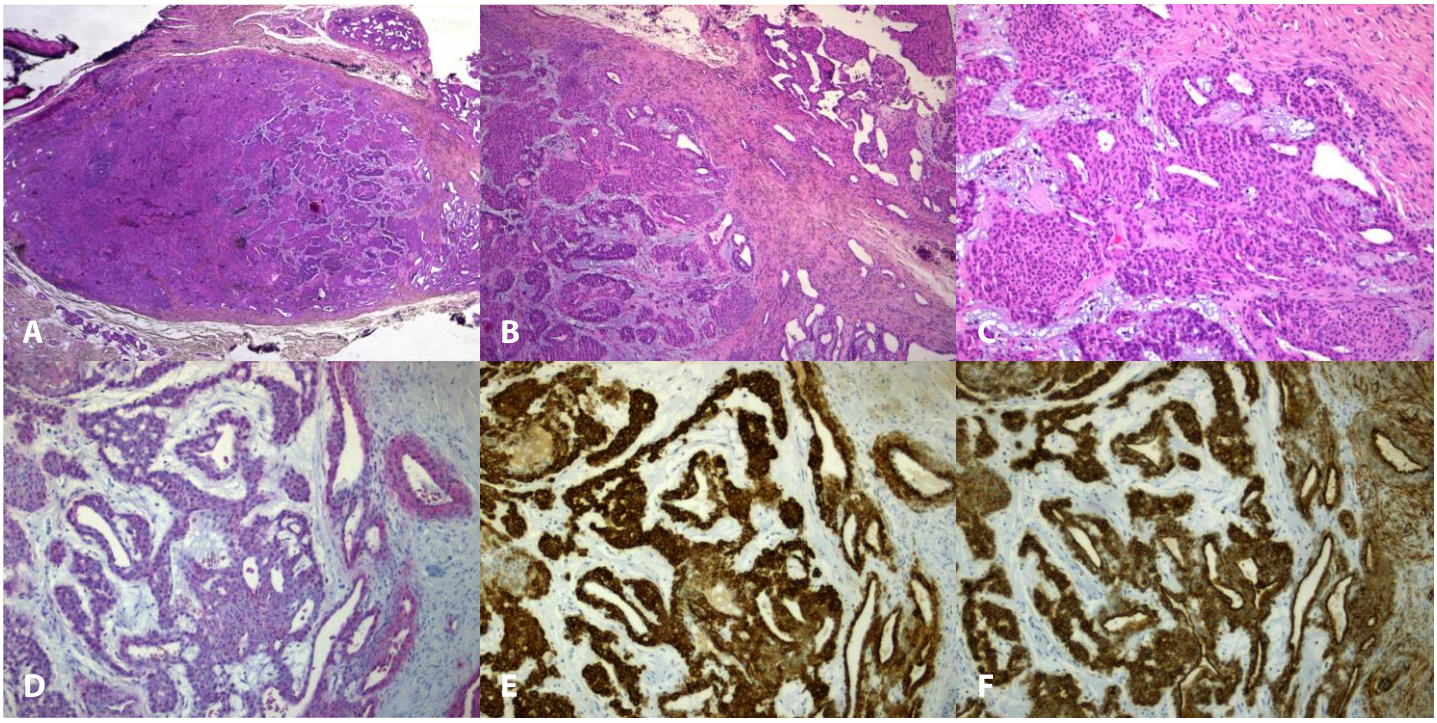

Figure 3. *Subungual myopericytoma, histopathology.* ***A-C****) A well-circumscribed, superficial dermal proliferation, without capsule, composed of plump, spindled myoid cells with eosinophilic cytoplasma, arranged concentrically around vessels. H&E, 16×, 40×, 100×, respectively.* ***D)*** *Immunohistochemical study showed intense and diffuse positivity for smooth muscle actin. 100×, and* ***E)*** *focal positivity for h-caldesmon****,*** *100×.* ***F)*** *CD34 positivity around the lesional vessels, 100×.*

Histopathologically, MP is characterized by a well-circumscribed, unencapsulated nodular proliferation with numerous thin-walled vessels and a concentric, perivascular arrangement of ovoid spindle-shaped myopericytes in the dermis, subcutis, or soft tissues [7]. Mitotic activity and cytologic atypia are usually minimal [7]. However, a rare atypical/malignant variant histologically characterized by high cellularity, significant mitotic activity, pleomorphism, necrosis, and metastasis has been reported [7]. Immunohistochemically, all cases express α-smooth muscle actin, and most also express h-caldesmon (90%), [7]. For CD34, the tumor cells around lesional vessels and in the intervascular areas are sometimes focally positive [8]. In contrast, these cells are usually negative for desmin, vimentin, cytokeratin, and S100 protein [3,7]. These features of this neoplasm help distinguish it from other myoid and perivascular tumors, such as glomus tumor, angioleiomyoma, and nodular hidradenoma [3,7].

In our case, the perivascular arrangement of cells could be consistent with a glomus tumor, as clinically suspected. However, the striking concentric arrangement of cells characteristic of MP is not seen in a glomus tumor [2,8]. Moreover, areas with spindle cells and abundant eosinophilic cytoplasm that resemble myofibroma are also not found in a glomus tumor [2,8].

Table 1. *Published cases of subungual myopericytomas.*

References	Age (years)	Sex	Finger	Clinical features	Path	Special Stains	Treatment	Follow-up (period)
Boix-Vilanova et al [4]	48	M	Third finger of right hand	Asymptomatic hyperkeratotic lesion on the pulp	9 months	+ SMA, h-caldesmon, CD31, CD34, Ki-67 (10%)	Surgical excision	No recurrence (1 year)
Current report	59	M	Third finger of left hand	Painful subungual dark macule	5 years	+ SMA, h-caldesmon (focal), CD34 (focal)	Surgical excision	No recurrence (5 months)

M,male; SMA, smooth muscle actin.

Treatment of MP is surgical excision [5]. It has a low recurrence rate (less than 4%), even when excision is incomplete and there are some reports of spontaneous regression [4]. Malignancy in MP appears to be extremely rare; only eight such cases have been reported in the literature to date [9].

Conclusion

Myopericytoma is a rare benign neoplasm of myoid perycitic origin, typically presenting as an asymptomatic slow-growing cutaneous or soft-tissue nodule on the extremities. The current case well highlights its characteristic histopathological and immunohistochemical features and is of particular interest due to its painful character, unusual appearance, and clinical appearance as a subungual black discoloration.

Potential conflicts of interest

The authors declare no conflicts of interest.

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