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Myopericytoma in an unusual location

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Abstract

Myopericytoma is a soft-tissue tumor of perivascular cells (pericytes). It is slow-growing, usually asymptomatic, and generally benign, although a malignant variant has been described. The etiology is unknown, but it has been associated with local trauma. The most common location is on the distal extremities. Histologically, it is characterized by a well-circumscribed, non-encapsulated proliferation of spindle shaped cells similar to myofibroblasts with oval nuclei and eosinophilic cytoplasm, arranged in perivascular concentric rings. There are few mitoses and no necrosis is reported. The immunohisto-chemical analysis is positive for smooth muscle actin and negative or weakly positive for desmin. A low Ki-67 proliferation index is typical. Treatment is surgical excision with free margins. Recurrences after adequate excision are uncommon. We describe a 48-year-old woman with a myopericytoma in an unusual location (next to the inner corner of her left eye) who was treated with surgical excision; there has been no recurrence after 5 years of follow up.

Keywords: myopericytoma, myopericytic tumor, soft-tissue tumor

Introduction

Myopericytoma (MP) is a perivascular tumor of unknown etiology. In 1996 Requena et al. were the first to use this term for the solitary myofiboma [1], but in 1998 Granter et al. adopted the term to describe benign tumors characterized histologically by a spindle-shaped cell proliferation with myoid perivascular/pericytic differentiation arranged concentrically around blood vessels [2]. This histopathological pattern includes a wide spectrum of tumors: myofibromatosis, glomangioma, infantile hemangiopericytoma, and the malignant myopericytoma described by McMenamin and Fletcher in 2002 [3]. In the most resent WHO classification of soft tissue tumors the MP and the myofibroma are separated; the glomangio-pericytoma is considered a subtype of MP [4].

Case Synopsis

A 48-year-old woman presented to the dermatology department of the 'Manuel Gea González' General Hospital with a subcutaneous tumor next to the inner corner of her left eye. It was an exophitic sessile dome shaped tumor measuring 1.2cm. The well-circumscribed tumor had a shiny surface and firm consistency (**Figure 1**A). The dermoscopic image showed a non-melanocytic lesion, with thick and arborizing blood vessels on the surface (**Figure 1**B). The patient described an evolution of 3 months, with asymptomatic progressive growth. There was no prior history of trauma. The patient was otherwise healthy and had no relevant medical history.

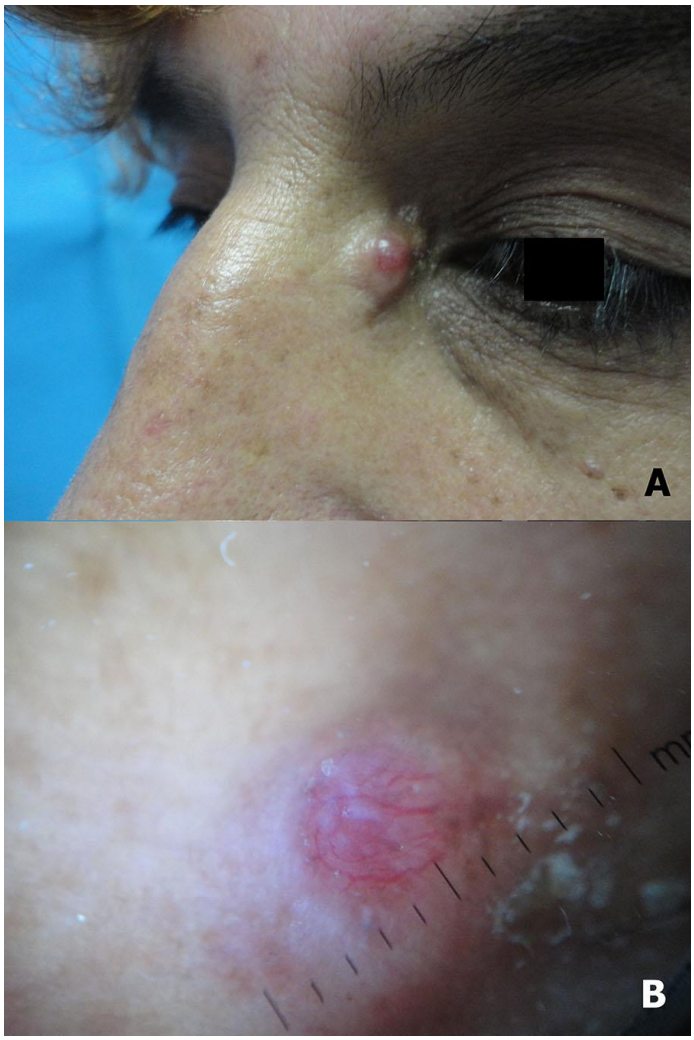

Figure 1. *A) initial clinical image. B) Dermoscopic image.*

An excisional biopsy was performed with the differential clinical diagnosis of cystic nodular basal cell carcinoma versus adnexal tumor. The histo-

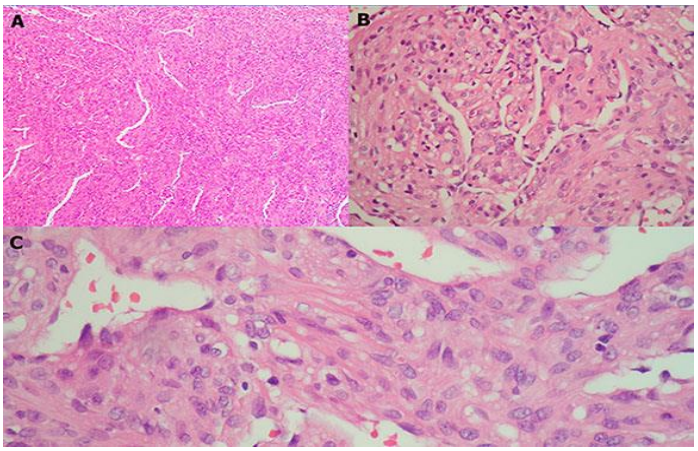

Figure 2. ***A)****, Multiple vascular spaces surrounded by myopericytic cell proliferation, 10×.* ***B)*** *40×.* ***C)*** *Vascular lumen with erythrocytes inside, myopericytic cells with spindle shaped nucleus, some hyperchromatic. H&E.*

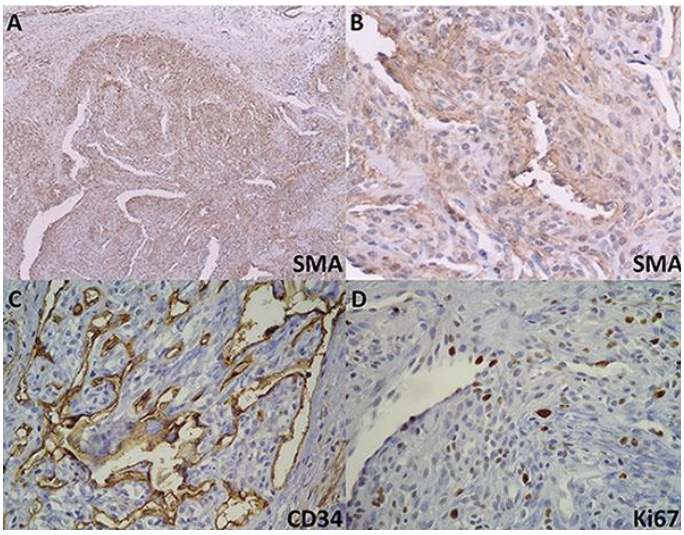

Figure 3. *Immunohistochemical analysis. A) Tumor cells positive for smooth muscle actin (SMA), 10×. B) SMA positive, 40×. C) CD34 negative on myopericytes, positive on endothelial cells, 40×. D) Ki67 positive, 40×.*

pathology study revealed a proliferation of fusiform cells with oval nuclei, eosinophilic cytoplasm, some mitosis, and low-grade atypia. These cells were arranged in fascicles with a swirling pattern mixed with collagen fibers and numerous thin-walled blood vessels (**Figure 2**). The immunohistochemical analysis was positive for smooth muscle actin and showed a Ki-67 proliferation index of 30% (**Figure 3**). The patient did not return for follow up.

One year after the surgery, the patient came back with a linear scar with a 1mm asymptomatic tumor in one of its edges (**Figure 4A**). An excisional biopsy was performed. Histology reported a well-delimitated non-encapsulated fusiform cell proliferation with hyperchromatic nuclei and few

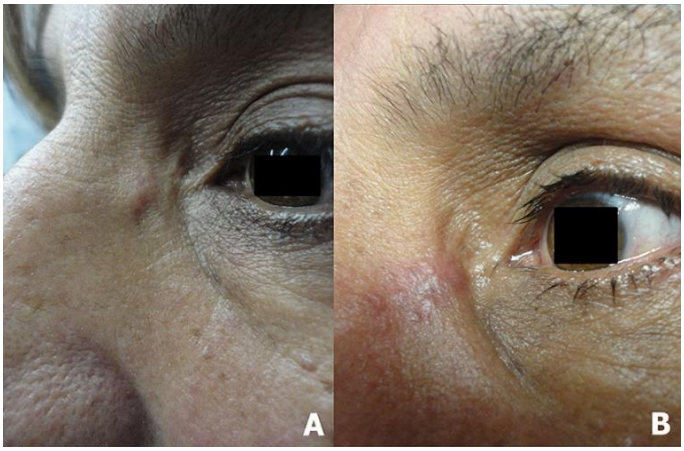

Figure 4. *A) Persistent tumor one year after the first surgery. B) Clinical result after 5 years.*

mitoses in a hyaline stroma, with blood vessels. The diagnosis was an MP with positive margins. Two months later, we performed a re-excision that reported free margins with positive tumor very close to the tumor bed, so we decided to widen the deep surgical margin. The final histological report showed clear margins.

This tumor has few mitoses and an absence of necrosis [5, 6]. The immunohistochemical analysis is positive for smooth muscle actin (SMA). It is negative or weakly positive for desmin and negative for S100 protein, HMB45, CD34, and cytokeratin. A low Ki-67 proliferation index is typical [6]. Immunohistochemical analysis performed on our patient's MP was SMA positive and CD34 negative with a Ki-67 proliferation index of 30%. The differential diagnosis for this subcutaneous tumor includes myopericytoma, glomus tumor, myofibroma, and angioleiomyoma (**Table 1**), [7].

Myopericytoma affects mainly adolescents and young adults, and it is more prevalent in males (ratio 44:29), [8]. It seems to be idiopathic, but there are cases reported after local trauma or associated with acquired immunodeficiency syndrome [9]. The clinical appearance of this tumor is a solid nodule with or without a pupuric or white halo. The tumors are sometimes fixed to deep tissues. Myopericytoma typically predominates on distal regions of the limbs, although some cases appear on the head and neck [10, 11]. It is usually asymptomatic or only painful when manipulated and grows slowly but progressively [9, 12]. The dermoscopic findings show a papular structure with scale, a blue-grey mosaic pattern, separated by white filaments, and multiple telangiectasias. Definite diagnosis is made by histopathology examination.

Treatment is surgical excision. It has a low recurrence rate (10-20%) and there are some reports of spontaneous regression after an incisional biopsy [6, 8]. Our patient has been followed up for 5 years with no evidence of recurrence (**Figure 4**).

We performed a literature search for cutaneous myopericytoma from 1996 to 2017 (**Table 2**); we found 21 cases, 10 women and 11 men. Ages ranged from 9 to 87 years (mean of 46.71 years). Most cases were found on the extremities (n = 18). Only 3 cases were reported on the face, two on the nose and one on the lip. These reports describe MP as a papular yellowish-to-skin colored neoplasm from 3 to 30mm in size, generally asymptomatic.

The case we report had residual tumor after the first surgery and presented with clinical re-growth after 1 year; the tumor was then fully excised.

Conclusion

MP is a benign proliferation of myoid pericytic origin and unknown etiology. MP can also present around vessels in the liver and lung.

The clinical appearance of cutaneous MP is usually a subcutaneous small slowly growing, asymptomatic neoplasm. The diagnosis is made with histopathology examination and immunohistochemistry.

Surgical excision is the treatment and it rarely recurs after complete excision.

Table 1. *Cases of MP in dermis and subcutis.*

Autor and year of publication	Sex	Age (years)	Topography	Morphology	Evolution (months)	Recurrence/ residual tumor	Depth	Symptoms	Treatment
Jeffries 2009[12]	Female	9	Left knee	7-mm pink to violaceous flat-topped papule with a pink halo	6	No recurrence	D/SC	None	Surgery
Morzycki 2017[13]	Male	33	Left index finger	Swelling and erythema	2	No recurrence	SC	None	Surgery
Dray 2006[6]	Male	30	Right hand	12mm nodule	UK	UK	D/SC	UK	Surgery
	Female	13	Right ankle	25mm nodule	24	Residual disease	D/SC	UK	Surgery
	Female	13	Left ankle	30mm nodule	5	UK	D/SC	UK	Surgery
	Female	71	Left ankle	10mm nodule	UK	No recurrence	D/SC	UK	Surgery
	Male	39	Sole of foot	11mm nodule	24	No recurrence	D/SC	UK	Surgery
	Male	60	Left hand, dorsum	8mm nodule	UK	UK	D/SC	UK	Surgery
	Male	48	Knee	9mm nodule	144	UK	D/SC	UK	Surgery
Mentzel 2006[14]	Female	87	Forearm	UK	UK	No recurrence	D	UK	Surgery
	Female	84	Forearm	UK	UK	No recurrence	D	UK	Surgery
	Female	49	Upper arm	UK	UK	No recurrence	D	UK	Surgery
	Male	19	Calf	UK	UK	No recurrence	D	UK	Surgery
	Male	63	Thigh	UK	UK	No recurrence	D/SC	UK	Surgery
	Male	47	Lower leg	UK	UK	No recurrence	D/SC	UK	Surgery
	Female	38	Knee	UK	UK	No recurrence	D	UK	Surgery
Laga 2008[15]	Male	64	Nose	4 discrete Nodules (left nasal tip 1.5 cm, left nasal ala 0.8 cm and right nostril at the level of the soft triangle 0.3 and 1.0 cm)	4	No recurrence	UK	None	Surgery
Sapelli 2009[9]	Male	28	Vermilion of the lower lip	freely movable 1.5-cm ovoid mass with a central round ulceration circumscribed by a round purplish hue	12	No recurrence (3 years)	D	None	Surgery
Numata 2009[10]	Male	59	Nose	soft, elastic, skin-coloured tumour with telangiectasia, 45 × 40 × 36 mm	60	Untreated	D/SC	None	Punch
Aung 2015[16]	Female	45	Wrist (dorsum)	Scaly, keratotic papule	UK	No recurrence	D	UK	Surgery
Ruiz-Arriaga 2017	Female	48	Inner corner left eye	Exophitic sesile dome shaped tumor, 10 x 10 x 5 mm	12	No recurrence	D	None	Surgery

D: dermis. D/SC: dermis/subcutis. UK: unknown.

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