Title
Myopericytoma in an unusual location

Permalink
https://escholarship.org/uc/item/5z02t0dk

Journal
Dermatology Online Journal, 24(4)

Authors
Ruiz-Arriaga, Leon Felipe
Ramirez Teran, Ana Laura
Ortiz-Hidalgo, Carlos
et al.

Publication Date
2018

License
CC BY-NC-ND 4.0

Peer reviewed
Myopericytoma in an unusual location

León Felipe Ruiz-Arriaga\(^1\) MD, Ana Laura Ramírez Terán\(^2\) MD, Carlos Ortiz-Hidalgo\(^3\) MD, Verónica Fonte-Ávalos\(^2\) MD, Sonia Toussaint-Caire\(^1\) MD, Ma Elisa Vega-Memije\(^1\) MD MSc, Mariana CatalinaDe Anda-Juárez\(^2\) MD

Affiliations: \(^1\)Department of Dermatopathology, ‘Dr. Manuel Gea González’ General Hospital, Mexico City, Mexico, \(^2\)Department of Dermatologic Surgery and Oncology, ‘Dr. Manuel Gea González’ General Hospital, Mexico City, Mexico, \(^3\)Department of Tissue and Cell Biology, School of Medicine, Universidad Panamericana, Mexico City, Mexico

Corresponding Author: Mariana Catalina De Anda-Juárez MD, Department of Dermatologic Surgery and Oncology, ‘Dr. Manuel Gea González’ General Hospital, Calz. de Tlalpan 4800, Tlalpan Centro I, Tlalpan, ZC 14080, Mexico City, Mexico, Tel: 52-55-4000-3000 Ext 1430, Email address: mdeanda73@gmail.com

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 immunohistochemical 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 myofibroma [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 recent WHO classification of soft tissue tumors the MP and the myofibroma are separated; the glomangiopericytoma 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 exophytic sessile dome shaped tumor measuring 1.2cm. The well-circumscribed tumor had a shiny surface and firm consistency (Figure 1A). The dermoscopic image showed a non-melanocytic lesion, with thick and arborizing blood vessels on the surface (Figure 1B). 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.
An excisional biopsy was performed with the differential clinical diagnosis of cystic nodular basal cell carcinoma versus adnexal tumor. The histopathology 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 1 mm asymptomatic tumor in one of its edges (Figure 4A). An excisional biopsy was performed. Histology reported a well-delimited non-encapsulated fusiform cell proliferation with hyperchromatic nuclei and few

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

**Figure 2.** A) Multiple vascular spaces surrounded by myopericytic cell proliferation, 10x. B) 40x. 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), 10x. B) SMA positive, 40x. C) CD34 negative on myopericytes, positive on endothelial cells, 40x. D) Ki67 positive, 40x.

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

References


