Cutaneous Myopericytoma on the Forehead: A Rare Localization

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Indian J Dermatol 2020:65(2):169-71

Sir,
Myopericytoma was first described by Granter et al. in 1998 as benign tumor showing a myoid/pericytic line of differentiation. [1] It is most commonly found in subcutaneous and deep dermis of distal extremities, but in some cases, superficial dermis is also involved. “Cutaneous myopericytoma” usually occurs as a solitary lesion, although multiple lesions may be seen. [2] In most cases, myopericytoma behaves in a nonaggressive manner and the recurrence has rarely been reported after surgical excision. [2,3] Histopathologically, myopericytoma is characterized by a well-circumscribed, nodular proliferation with numerous thin-walled blood vessels and a concentric, perivascular arrangement of ovoid spindle-shaped cells with eosinophilic cytoplasm and vesicular nuclei in the dermis, subcutis, or soft tissues. Mitotic activity and cytological atypia is usually minimal. Immunohistochemically, all cases express smooth muscle actin (SMA) and 90% of cases express caldesmon; other smooth muscle markers are negative. [3] Expanding the clinical spectrum of this uncommon benign tumor, we describe a case of cutaneous myopericytoma of the forehead to review its histopathological and immunohistochemical features.

A 44-year-old female presented to our dermatology department with a slow-growing painful red nodule on her forehead. Except for diabetes mellitus, she did not have any underlying disease or any history of trauma. The dermatologic examination showed a 2 cm × 2 cm lesion with an irregular border, violet atrophic, and nodular plaque located in the frontal region [Figure 1]. Our differential diagnosis included discoid lupus erythematosus, sarcoidosis, and pseudolymphoma, and we performed a punch biopsy. Histopathological examination of the biopsy revealed spindle-shaped myoid-appearing cells in a concentric arrangement, associated with thin-walled vascular channels with no mitotic activity or cytologic atypia [Figure 2a and b]. Immunohistochemically, although the perivascular cells were focally positive for CD34 and CD31 for SMA, desmin was negative [Figure 2c]. After correlating clinical and histopathological findings and the immunohistochemical pattern, we diagnosed this lesion as myopericytoma. Because of complaints of ongoing pain in her scalp, she was referred to plastic surgery for total excision. There was no evidence of recurrence during 8 months of follow-up.

Figure 1: A 2 cm × 2 cm violaceous atrophic plaque with irregular border in the frontal region
Myopericytoma is a very rare benign perivascular neoplasm that arises from the perivascular myoid cells. This kind of neoplasm usually is a slow growing, well-circumscribed, mostly painless firm nodule.\[1\] In contrast, our patient complained of a painful lesion in her forehead. One month after surgery, redness and pain disappeared. Aung et al. evaluated the literature about the “cutaneous myopericytoma” and reported that most patients were male (male:female = 1.5:1), and the patient’s age ranged from 13 to 87 years (median = 47 years). Although some lesions were painful, most were asymptomatic. The lower extremity was most commonly affected (37 cases), followed by the upper extremity (18 cases), the head and neck region (9 cases), and the trunk (2 cases). Most lesions were solitary, although five were multiple, either in single or in multiple anatomic locations. All lesions were mostly nodular, firm gray-white, or hemorrhagic red-brown lesions. In the majority of the cases, the lesions were confined to the dermis and superficial parts of the subcutis.\[4\] Our patient was 44-year-old female. Her lesion was solitary, painful, and mild atrophic and nodular, and it was located on her scalp hairline.

These lesions frequently occur in the lower extremities and can also be found in the upper extremities, the head and neck region, and the trunk. The largest series of “cutaneous myopericytoma” was reported by Mentzel et al. (54 cases, of which 26 were cutaneous myopericytoma).\[3\] Myopericytoma of the scalp is rare; only a few cases have been reported in the literature. We emphasized the localization of the lesion and its painful character. There is only one case of scalp localization of myopericytoma, which was reported by Aung et al. in three cases.\[4\] The distinctive histological feature of myopericytoma is a marked, concentric perivasculare proliferation of myoid tumor cells. These cells are characteristically reactive for SMA and negative for CD34, cytokeratin, and $S100$ protein. Desmin and vimentin are focally positive and generally negative. These features of this neoplasm help distinguish it from other myoid and perivascular tumors.\[5\] The differential diagnosis involves glomus tumor, angioleiomyoma, and nodular hidradenoma.\[4\] These tumors are often painful and generally have a more fascicular pattern than myopericytomas and are distinguished in a straight forward manner on histopathology because of these features.\[7\] The biologic behavior of most cutaneous myopericytomas is benign, though a few malignant and/or recurring cases were described.\[6\] We believe that this case will help other physicians to recognize this rare entity and to improve the understanding of myopericytoma which should be kept in mind in the differential diagnosis of nodular lesions.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

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How to cite this article: Cebeci D, Yaşar Ş, Güneş P, Aytekin S. Cutaneous myopericytoma on the forehead: A rare localization. Indian J Dermatol 2020;65:169-71. Received: July, 2018. Accepted: September, 2018.