

Myopericytoma of lip: A rare lesion in an unusual location

Shaista M. Vassenwala, Nishat Afroz, Hena A. Ansari, Arshad Hafeez Khan¹, Rabeya Basari, Suhailur Rehman

Departments of Pathology and ¹Plastic and Reconstructive Surgery, Jawaharlal Nehru Medical College, AMU, Aligarh, Uttar Pradesh, India

Address for correspondence:

Dr. Hena A. Ansari, Department of Pathology, Jawaharlal Nehru Medical College, AMU, Aligarh, Uttar Pradesh - 202 002, India.
E-mail: hena.jnmc@gmail.com

ABSTRACT

Myopericytoma is a rare benign tumor with perivascular myoid differentiation. It shares histomorphologic features with the so-called hemangiopericytoma, myofibroma and glomus tumor. We hereby report the case of a 14-year-old boy who presented with a slowly growing, painless, firm mass on upper lip, diagnosed as myopericytoma on the basis of histopathology and immunohistochemistry. To the best of our knowledge, this is only the second such reported case.

KEY WORDS: Benign neoplasm, immunohistochemistry, lip, myopericytoma

INTRODUCTION

Myopericytoma is a relatively new entity which is listed as a perivascular tumor in the World Health Organization classification of soft tissue tumors.^[1] This benign neoplasm shares a very interesting histogenetic and morphologic relationship to other similar tumors, namely, glomus tumors or glomangiopericytomas, myofibromas and the some what isolated hemangiopericytoma.^[2,3]

Myopericytomas usually arise in the skin and subcutis of lower extremities but other reported sites of involvement are the proximal extremities, head and neck region and trunk. Multiple lesions have also been documented.^[1]

Only three cases of myopericytoma are reported involving the soft tissue of the oral cavity, and there is only a single report of involvement of the lip.^[4-7] To the best of our knowledge, this is the second such case, in which a 14-year-old male patient was diagnosed with a myopericytoma of the upper lip, with the help of immunohistochemistry (IHC).

CASE REPORT

A 14-year-old boy presented to the plastic and reconstructive surgery out-patient section with a slowly enlarging, painless mass on the lip that had been present for 1-year. There was no history of trauma, tuberculosis, or any other infection. On examination, a freely movable, round, well-circumscribed, firm, nontender mass, 2 cm in size, was seen on the upper lip. The mouth, teeth and periodontum were normal. There was no other intraoral soft tissue lesion or infection. Regional lymphnodes were not palpable. A clinical diagnosis of pyogenic granuloma was made, and the lesion was excised and submitted for histopathological evaluation.

The excised lesion consisted of skin covered firm nodule, 2 cm in diameter. The cut surface was whitish and solid in appearance. The hematoxylin and eosin — stained

Access this article online
Website: www.ijpmonline.org
DOI: 10.4103/0377-4929.155317
Quick Response Code:


sections [Figure 1] showed proliferating nodules of spindle cells arranged around blood vessels and capillaries of varying caliber. The nodules were separated by fibrocollagenous bundles. The nuclei were spindle as well as ovoid to round and vesicular, and the cytoplasm was eosinophilic. There was no necrosis or atypia. The mitotic rate was low (1/10 high power fields). The vascular stroma showed scattered skeletal muscle bundles. The tumor was noncapsulated, and there was 2 mm free margin from line of excision.

Immunohistochemistry revealed strong positivity for smooth muscle actin and weaker stain for CD34 [Figure 2] suggesting that the tumor cells were myopericytic in origin. The final diagnosis of myopericytoma was thus established. The patient was followed-up for 6 months and showed no recurrence.

DISCUSSION

Pericytes are perivascular stem cells which considered to be pluripotent and capable of differentiating into smooth muscle

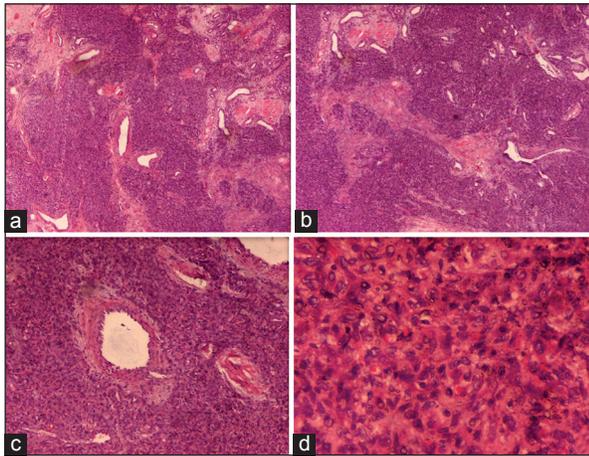


Figure 1: (a and b) Nodular sheets of tumor cells around blood vessels of varying caliber (H and E, $\times 4$) (c) mesenchymal tumor cells arranged around a blood vessel, with a perivascular collagenous zone free from tumor (H and E, $\times 10$) (d) tumor cells display plump ovoid to spindle nuclei, with bland appearance and low mitotic activity (H and E, $\times 40$)

cells and glomus cells, among others.^[8] The myopericyte thus represents a transitional form between pericytes and the smooth muscle cells of vessels.

The myopericytoma was recently characterized as a tumor with a hemangiopericytoma-like vascular pattern.^[1,3] The term “myopericytoma” was initially proposed by Granter *et al.*^[2] The most common location for this tumor is skin and subcutis in adult patients.^[1]

Myopericytomas are benign lesions, although a few recurring and/or malignant cases have been described.^[3,6,8] They have been reported to arise in immunodeficient patients, in which case they are associated with the Epstein–Barr virus (EBV).^[9]

Myopericytomas show overlapping histological features with other perivascular tumors, namely the hemangiopericytoma, glomangiopericytoma and myofibromas. Some tumors may show myofibroma-like areas while others may show features of glomus tumors. It is thus evident that these differing lesions are actually part of a continuum or spectrum of perivascular neoplasms.^[1,2]

In the differentiation from myofibromas, the most helpful features are the characteristic perivascular proliferation and hemangiopericytoma — like vascularity in myopericytoma, and more well-defined presence of paucicellular areas, and zonation in myofibromas.^[10]

Lobular capillary hemangioma, more commonly known as the pyogenic granuloma, was provisional the clinical impression in this case. The oral cavity is a common location. However, on low power, the lobular architecture and small capillaries, with a larger feeder vessel offers a clue to the diagnosis.

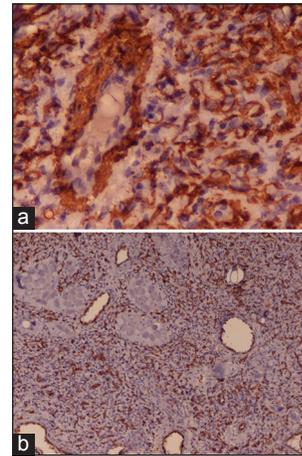


Figure 2: (a) Tumor areas show strong positivity for smooth muscle actin ($\times 40$) (b) CD34 positivity is limited to endothelial cells in stromal vascular channels

Superficial/dermal epithelioid hemangioma should also be considered in the differential diagnosis of myopericytoma. These lesions demonstrate a proliferation of small blood vessels and lack a prominent lymphoid collection. IHC shows positive reactivity of cells for CD31, CD34 and factor VIIIrAg.^[1]

Myopericytoma arising in association with a deficient immune system has to be differentiated from Kaposi’s sarcoma. The former may be associated with EBV, whereas the latter is associated with human herpes virus-8 infection. On histopathology, Kaposi’s sarcoma shows dermal proliferation of irregular slit-like vascular channels with extravasated erythrocytes, hemosiderin and plasma cells. Intracytoplasmic periodic acid-Schiff positive eosinophilic hyaline granules can be detected in the tumor cells.^[1]

The occurrence of myopericytoma in association with EBV in patients with acquired immunodeficiency syndrome is a distinctive phenomenon. EBV-associated smooth muscle tumors are a separate class of neoplasms,^[11] to which myopericytoma can now be added.

Wide local excision is the recommended treatment, with careful follow-up. Recurrences have been described in cases of myopericytomas, as well as malignant features.^[6,8] Multinodular tumors or deep-seated tumors behave more aggressively when compared to superficial nodules.^[1] Pleomorphism and high mitotic rate are the determining factors for malignant myopericytoma.^[8]

It thus appears that myopericytomas are great mimickers and can arise in varied locations. Their diagnosis more often than not, requires the judicious use of immunomarkers. Multiple lesions or lesions in unusual locations or with a history of rapid growth should be with assessed with high clinical suspicion of immunodeficiency or possible malignant behavior.

REFERENCES

1. Fletcher CD, Unni KK, Mertens F, editors. World Health Organisation Classification of Tumours. Pathology and Genetics of Tumours of Soft Tissue and Bone. Lyon, France: IARC Press; 2002. p. 138-9.
2. Granter SR, Badizadegan K, Fletcher CD. Myofibromatosis in adults, glomangiopericytoma, and myopericytoma: A spectrum of tumors showing perivascular myoid differentiation. *Am J Surg Pathol* 1998;22:513-25.
3. Mentzel T, Dei Tos AP, Sapi Z, Kutzner H. Myopericytoma of skin and soft tissues: Clinicopathologic and immunohistochemical study of 54 cases. *Am J Surg Pathol* 2006;30:104-13.
4. Datta V, Rawal YB, Mincer HH, Anderson MK. Myopericytoma of the oral cavity. *Head Neck* 2007;29:605-8.
5. Ide F, Obara K, Yamada H, Mishima K, Saito I. Intravascular myopericytoma of the oral mucosa: A rare histologic variant in an uncommon location. *Virchows Arch* 2007;450:475-7.
6. Terada T. Myopericytoma of low grade malignancy in the oral cavity. *Rare Tumors* 2012;4:e9.
7. Sapelli S, Ribas M, Martins WD, de Noronha L, Gomes AP. Myopericytoma of the lip: Report of case. *Head Neck* 2009;31:561-4.
8. McMenamin ME, Fletcher CD. Malignant myopericytoma: Expanding the spectrum of tumours with myopericytic differentiation. *Histopathology* 2002;41:450-60.
9. Lau PP, Wong OK, Lui PC, Cheung OY, Ho LC, Wong WC, *et al.* Myopericytoma in patients with AIDS: A new class of Epstein-Barr virus-associated tumor. *Am J Surg Pathol* 2009;33:1666-72.
10. Dray MS, McCarthy SW, Palmer AA, Bonar SF, Stalley PD, Marjoniemi V, *et al.* Myopericytoma: A unifying term for a spectrum of tumours that show overlapping features with myofibroma. A review of 14 cases. *J Clin Pathol* 2006;59:67-73.
11. Deyrup AT, Lee VK, Hill CE, Cheuk W, Toh HC, Kesavan S, *et al.* Epstein-Barr virus-associated smooth muscle tumors are distinctive mesenchymal tumors reflecting multiple infection events: A clinicopathologic and molecular analysis of 29 tumors from 19 patients. *Am J Surg Pathol* 2006;30:75-82.

How to cite this article: Vasenwala SM, Afroz N, Ansari HA, Khan AH, Basari R, Rehman S. Myopericytoma of lip: A rare lesion in an unusual location. *Indian J Pathol Microbiol* 2015;58:211-3.

Source of Support: Nil, **Conflict of Interest:** None declared.

