Pathol. Oncol. Res. (2010) 16:613–616
DOI 10.1007/s12253-010-9253-y

Minute Myopericytoma of the Neck: A Case Report with Literature Review and Differential Diagnosis

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Received: 29 December 2009 / Accepted: 17 February 2010 / Published online: 20 March 2010

Abstract Reports of cutaneous myopericytoma (MPC) are very rare. The author herein reports a case of minute MPC of the neck. A 56-year-old woman noticed a painful small tumor in the neck, and consulted to our hospital. Dermatologists's diagnosis is a hyperplastic lymph node. Excision of the tumor was performed. Grossly, the tumor was a sold white tumor measuring 3×3×3 mm. Microscopically, it consisted of many vascular channels and perivascular cell proliferation encased by a fibrous capsule. The vascular proliferation showed a hemangiopericytoma (HPC)-like pattren such as staghorn-like vessels. Fibrosis was not present. The HPC-like cells had vesicular nuclei and polygonal cytoplasm. No atypia is recognized. The HPC-like cells focally showed vague nodular proliferation around the vessels. Immunohistocheically, the tumor cells were negative for cytokeratin, and positive for vimentin. The vasculatures were positive for factor VIII-related antigen, CD34, and CD31. The HPC-like tumor cells were positive for α-smooth muscle actin and h-caldesmon, but negative for desmin, S100 protein, melanosome, bcl-2, CD99, and KIT. The Ki-67 labeling was 8% and p53 was negative. The pathologic diagnosis was MPC of the neck skin. The patient is now alive without recurrence 4 years after the excision. A review of the literature revealed 73 cases of MPC from 6 papers. MPC is male predominance, and the patients ages ranges from 13 to 87 years with the median of 47 years. The most common location was lower extremities followed in order by upper extremities, head and neck, and trunk. One MPC occurred within the vasculature, and 3 cases of MPC developed in the scar or trauma lesions. The prognosis after excision is good, but a very minority showed local recurrence. A differential diagnosis was also made.

Keywords Skin · Myopericytoma · Perivascular myoid cells · Histopathology · Immunohistochemistry

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Introduction

Hemangiopericytoma (HPC) is a neoplasm of perivascular pericyte-like cells, and characterized by HPC-like cell proliferation with abundant vascular channels which often show staghorn-like appearances [1, 2]. HPC is a heterogenous tumor and it contained some speicifc neoplasms; the term of HPC is a waste basket diagnosis. By the recent recognition of perivascular myoid cells (PMC), several specific disease entities have appeared recently. Tumors derived from PMC contain myopericytoma (MPC) [3, 4], myofibroma [4], perivascular epithelioid neoplasm (PEComa) [5, 6], glomus tumor [7, 8], angioleiomyoma [9], myofibromatosis [10], and true HPC [11-13]. These neoplasms show a spectrum of tumors that have perivascular myoid cell phenotypes, and there are overlaps among these tumors. Among these, MPC is characterized by HPC-like lesion and immunophenotypes positive for α-smooth muscle actin and h-caldesmon, but negative for desmin and other antigens. Unlike PEComa, MPC does not exhibit melanocytic lineage. To the best of the author's knowledge, reports of cutaneous MPC is relatively scant; 73 cases of MPC have been reported in 6 papers. In this article, the author reports a case of minute MPC, and reviewed the previously-reported cutaneous MPC. A differential diagnosis was also made.

Case Report

A 56-year-old woman noticed a painful small tumor in the neck, and consulted to our hospital. Dermatologists's diagnosis was a hyperplastic lymph node. Excision of the tumor was performed. Grossly, the tumor was a sold white tumor measuring 3×3×3 mm. Microscopically, it consisted of many vascular channels and perivascular cell proliferation encased by a fibrous capsule (Fig. 1). The tumor was located in the deep dermis. The vascular proliferation showed an HPC-like pattern such as staghorn-like vessels (Figs. 2 and 3). Fibrosis was not present. The HPC-like tumor cells focally showed vague nodular proliferation around the vessels (Figs. 3 and 4). The tumor cells showed round and polygonal shape with vesicular nuclei. The cell boundaries were indistinct. Mitotic figures were present in 3 per 50 high power fields. Nuclear hyperchromasia, atypia, and necrosis were absent.

An immunohistochemical study was performed with the use of Dako Envision method (Dako, Glostrup, Denmark), as described previously [14, 15]. The antibodies used were as follows: cytokeratin (AE 1/3, Dako), cytokeratin (CAM5.2, Beckton-Dickinson, CA, USA), CD34 (NU-3A1, Dako), S100 protein (polyclonal, Dako), desmin (D33, Dako), α-smooth muscle antigen (1A4, Dako), myoglobin (polyclonal Dako), vimentin (Vim 3B4, Dako), CD31 (JC70A, Dako), p53 protein (DO7, Dako), Ki-67 (MIB-I, Dako), melanosome (HMB 45, DAKO), factor VIII-related antigen (36B11, Novocastra, Newcastle upon type, UK), bcl-2 (124, Dako), KIT (polyclonal, Dako), CD99 (12E7, Dako), and h-caldesmon (h-CD, Dako). The HPC-like tumor cells were negative for cytokeratin, and positive for vimentin. The vasculatures were positive for factor VIII-related antigen, CD34 (Fig. 5), and CD31, but

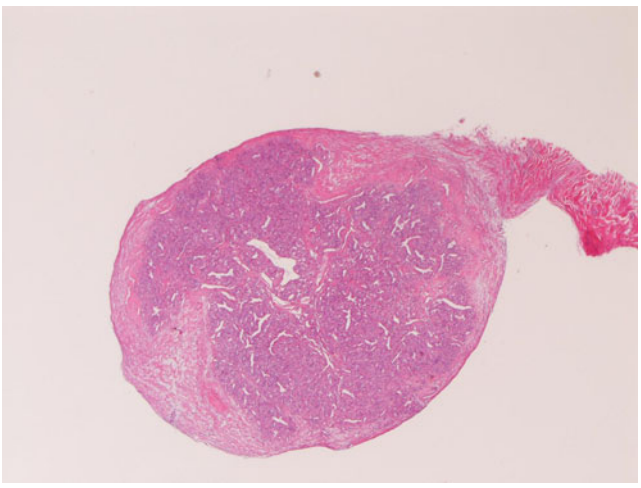

Fig. 1 Low power view of the neck tumor. The tumor is located in the deep dermis. The tumor is small, and measures 4×5×4 mm. The tumor shows hemangiopericytomatous pattern. HE, ×4

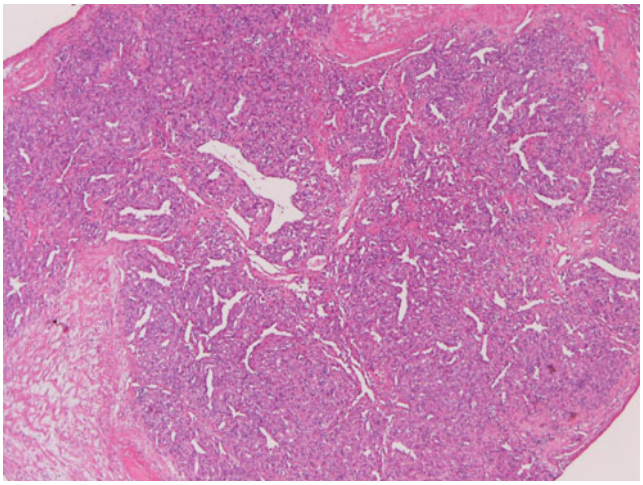

Fig. 2 The tumor shows hemangiopericytomatous pattern. HE, ×10

negative for other antigens examined. The HPC-like tumor cells were positive for α-smooth muscle actin (Fig. 6) and h-caldesmon, but negative for desmin, S100 protein, melanosome, myoglobin, CD31, CD35, factor VIII-related antigen, bcl-2, CD99, and KIT. The Ki-67 labeling was 8% and p53 was negative.

The pathologic diagnosis was MPC of the neck skin. The patient is now alive without recurrence 4 years after the excision.

Discussion

The negative reaction for cytokeratins and positive reaction for vimentin indicate that the present tumor is a definite

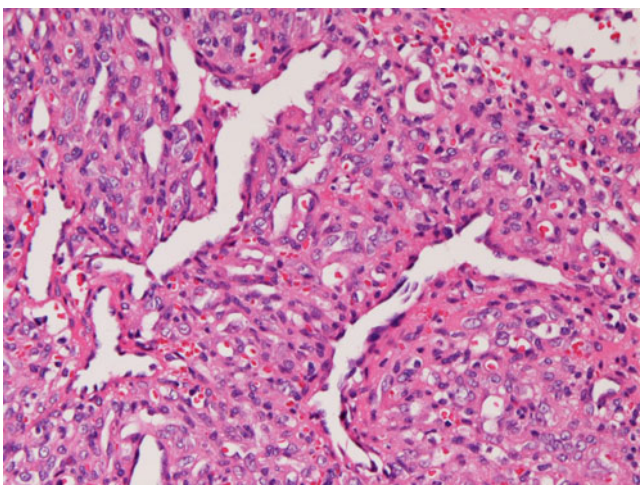

Fig. 3 The tumor is composed of vascular endothelial cells and perivascular stromal cells. The vasculatures show staghorn appearances characteristic of hemangiopericytoma. The tumor cells are free of atypia. Vague nodular pattern is seen around the vasculatures. HE, ×200

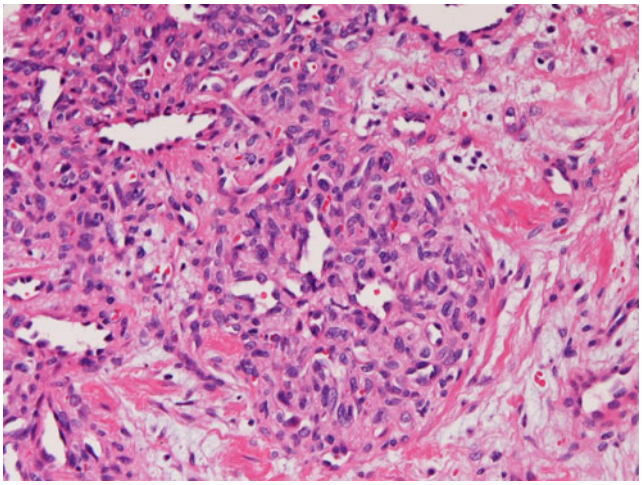

Fig. 4 A nodular pattern around the vasculatures is seen. HE, ×200

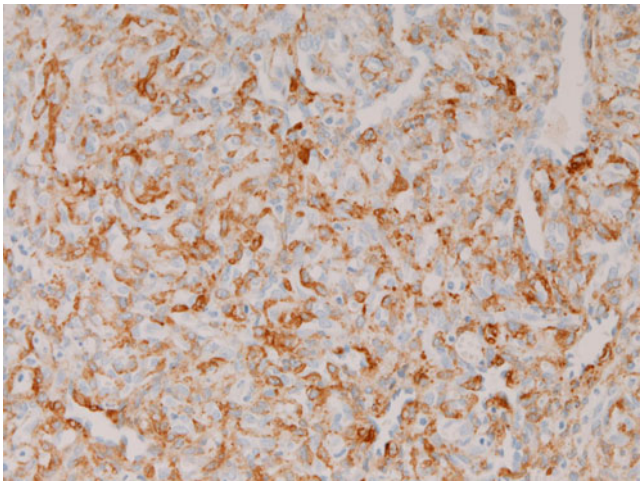

Fig. 6 The tumor cells are strongly positive for α-smooth muscle actin. ×200

mesenchymal tumor. The present case showed HPC-like features, and no atypical features were seen. The tumor cells formed nodular proliferation around the vessels, and immunohistochemically positive for α-smooth muscle actin and h-caldesmon and negative for desmin. These histological and immunohistochemical features of the present cases are typical for MPC [3, 4]. Conventional HPC is unlikely, because tumor cells expressed smooth muscle markers, indicating that the present case is derived from perivascular myoid cells. The absence of atypia and mitotic figures, the low Ki-67 labeling, the absence of p53 protein, and the clinical course indicate complete benign nature of the present case.

Differential diagnosis includes tumor showing HPC-like features, i.e. solitary fibrous tumor (SFT), myofibroma, myofibromatosis, angioleiomyoma, glomus tumor, perivascular epithelioid cell neoplasm (PEComa), extragastrointestinal stromal tumor (eGIST), synovial sarcoma, and malignant MPC.

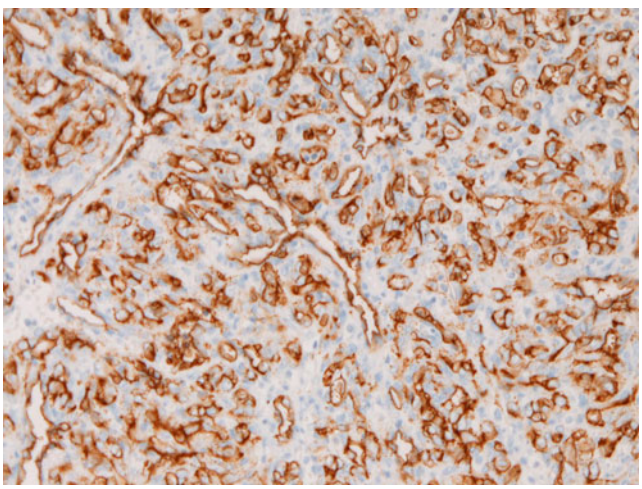

Fig. 5 CD34 stain highlights the vascular endothelial cells. The tumor cells were negative for CD34. ×200

The present tumor is different from SFT. In general, SFT is collagenized tumor with patternless pattern and pericytomatous pattern [16]. SFT is almost always positive for CD34. SFT may express bcl-2 and CD99 (MIC1) [16]. In the present study, such histological and immunohistochemical features as SFT were not recognized. Therefore, the present tumor is different from SFT.

The present case is apparently different from myofibroma [4], myofibromatosis [9], and angioleiomyoma [10], a disease spectrum showing perivascular myoid cell phenotype, in that the present case lacked fibrous areas and leiomyomatous areas.

Glomus tumor shows perivascular cuboidal epitheloid cells positive for α smooth muscle actin, and have an organoid patten of the glomus organ. The present case lacked such glomangioma features.

PEComa usually shows epitheliod cells expressing smooth muscle actin and melanocytic markers [5, 6]. The present case did not show epithelioid morphologies, and was negative for melanosome and S100 protein. Therefore, the present case is not PEComa.

eGIST is almost always positive for KIT and CD34, and frequently showed mutations of KIT and/or platelet-derived growth factor receptor-α [17-21]. The present tumor was negative for CD34 and KIT, indicating that the present tumor is not eGIST.

Minute synovial sarcoma is usually biphasic tumor [22]. Minute monophasic mesenchymal synovial sarcoma shows malignant spindle cells, and positive for cytokeratin and/or epithelial membrane antigen [22]. The present case lacked evidence for malignancy and positive cytokeratin immunoreactivities. Therefore, the present tumor is different from synovial sarcoma.

A few cases of malignant MPC have been reported: McMenamin and Fletcher reported 5 cases of malignant MPC [23]. In their report, malignant MPC is composed of hyperchromatic myoid-appearing ovoid to spindle cells. The tumor cells show highly mitotic figures. The nodular proliferations of tumor cells are focal. Their cases frequently showed infiltrative growth and metastasis. The present case is small, and well defined. In addition, the present case lacked nuclear atypia and significant mitotic figures. Therefore, the present case is not malignant MPC.

Finally, a review of the literature revealed 73 cases of MPC from 6 papers [3, 4, 24-27]. MPC is male predominance; the male to female ratio was 44: 29. The patients ages ranges from 13 to 87 years with the median of 47 years. The most common location was lower extremities followed in order by upper extremities, head and neck, and trunk. One MPC occurred within the vasculature, and 3 cases of MPC developed in the scar or trauma lesions. Immunohistochemical staining of α-smooth muscle actin was 100%, and h-caldesmon 91%. Desmin was always negative. The prognosis after excision is good in the 71 patients, but a very minority (2 patients) showed local recurrence. No remote metastasis was found. There were no patients that died of this neoplasm.

Conflict of interest None

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