Diagnostic dilemma in soft tissue swelling over finger

Ruchi Nagpal¹, Arvind Ahuja²

Abstract
Myopericytoma is a benign tumor that shows a pericytic line of differentiation. The lesion is generally located in the dermis and subcutaneous tissue and has a predilection for distal extremities, but may be found in any part of the body. We present a case of 48-year old male who came with painful swelling in right middle finger for the past 2 years. Histopathology showed proliferation of spindle-shaped cells exhibiting a hemangio-pericytomatous pattern with various sized vessels lined by flattened endothelium and surrounded by a concentric arrangement of spindle-shaped cells. Tumor cells showed membranous positivity for smooth muscle actin but negative for desmin and CD 34. Hence diagnosis of intravenous myopericytoma was established.

Keywords: myopericytoma, intravenous, actin, CD34

Case report
Myopericytoma (MP) is a benign tumor that shows differentiation towards pericytes [1]. The lesion is generally located in the dermis and subcutaneous tissue and has a predilection for distal extremities, but it may be found in any part of the body [2]. It is composed of oval to spindle-shaped myoid-appearing cells with a strong tendency towards concentric perivascular growth [2,3]. An intravascular myopericytoma (IVMP) is a distinct, histological variant [1] and has been rarely documented.

A 48-year-old male presented with painful swelling in left middle finger for the past 2 years. There was no history of trauma or previous injury. On examination, there was palpable subcutaneous lump measuring 0.5 cm in diameter on the volar aspect of left middle finger and was clinically diagnosed as ganglion cyst. Radiograph showed soft tissue swelling over the middle finger (Figure 1). The resected specimen (0.8 X 0.5 cm) sent for histological examination revealed a solid tumor that had replaced part of the vessel wall and completely filled the lumen (Figure 2a). The tumor showed proliferation of spindle-shaped cells exhibiting a “hemangio-pericytomatous” pattern (Figure 2b). The presence of various sized vessels with flattened endothelium surrounded by a concentric arrangement of rounded or slightly spindle-shaped cells was evident (Figure 2c). The lesional spindle cells were arranged in sheets in the intervascular areas. Few thick walled vessels with plump endothelial cells were also noted. No atypia, mitosis or necrosis were observed. Immunohistochemistry: tumor cells showed membranous staining for smooth muscle actin (SMA) (Figure 3a) but negative for desmin (Figure 3b) and CD 34 (Figure 3c). The muscular wall of the vessel within which tumor was located showed cytoplasmic positivity for desmin (Figure 3b). Based on the above findings diagnosis of intravascular myopericytoma was established.
Figure 1. Dorsovolar radiograph of the left hand showing swelling in the middle finger.

Figure 2. Characteristic of intravascular myopericytoma:
(a) Intravascular proliferation of the tumor cells (H&E, 10X).
(b) Spindle-shaped cells exhibiting a “hemangio-pericytomatosus” pattern (H&E, 20X).
(c) Vessels with flattened endothelial cells surrounded by a concentric arrangement of rounded or slightly spindle-shaped cells (H&E, 40X).

Figure 3. Immunohistochemistry:
(a) Diffuse and intense membranous positivity for smooth muscle actin (SMA).
(b) Muscular wall of the vessel shows cytoplasmic positivity for desmin whereas tumor cells are negative.
(c) Negativity for CD34 with endothelial internal control.
The first completely intravascular variant was described by McMenamin and Calonje (2002), which was located within a subcutaneous vein of the thigh [4]. While most MPs are painless, the intravascular subtype tends to have a painful course which can be attributed to the thrombus associated with the lesion [4,5]. The tumor shares the morphological characteristics with many entities that can be intravascular - angioleiomyoma, glomus tumor, perivascular myoma, hemangiopericytoma, myofibroma, myofibromatosis, papillary endothelial hyperplasia and pyogenic granuloma [3,4,6]. The distinctive feature of intravascular myopericytoma from other perivascular myoid tumors is that it shows a biphasic pattern: whorled nodules with bundles of mature eosinophilic cells, and immature small mesenchymal cells associated with numerous thin-walled hemangio-pericytoma like vessels [4,7,8]. Angioleiomyomas generally have a more fascicular pattern than myopericytomas and are composed of fascicles of smooth muscle cells with cigar-shaped nuclei with abundant brightly eosinophilic cytoplasm that express desmin in addition to SMA [4,8]. Moreover, it does not have the concentric pattern that accentuates the vessel walls which is characteristic of myopericytoma [4]. In our case, tumor cells were negative for desmin. MPs are usually benign lesion but they may recur or even show malignant transformation, therefore histopathological findings should always be confirmed by immunohistochemistry and patient has to be kept on follow-up [7,9]. Eight months post-operatively our patient had no symptoms and there was no evidence of recurrence. To conclude, IVMP is a rare entity and few cases, of which only three in the upper extremity, have been reported in the literature so far [5,9].

References