Clinicopathologic features in a case of intermuscular myopericitoma of thigh

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Myopericytoma is a benign tumor with the most common presentation as a well-circumscribed, slow-growing mass. It is frequently misdiagnosed as a sarcoma. We presented a 23-year-old patient with a history of a sciatic pain of the right leg. A careful physical examination discovered tumor-like mass in the posterior part of the thigh. Neurological finding showed a reduction of myotatic reflexes on the right leg with a weaker muscle strength on the right leg. The right leg musculature was slightly hypotrophic in the range of 2-3 cm comparing to left leg. Initially electrophysiological and radiological diagnostic with magnetic resonance imaging (MRI) of the lumbar spine, pelvis and thighs were normal. Magnetic resonance imaging of the right thigh discovered a slow growing 2.1 × 3.8 cm sized mass that was initially described by radiologist as a neurinoma. Patient was admitted to department of neurosurgery and operated on for a tumor removal. Tumor was located intimately to femur and sciatic nerve and after careful dissection completely removed. Patient was doing well after surgery and discharge after three days from the hospital. In the postoperative period the symptoms disappeared. Histopathology showed a myopericytoma. Postoperative MRI after three months of follow up showed no tumor residues, and after 6 and 12 months there was no tumor recurrence. Myopericytoma behave in a benign fashion, but, because local recurrences and rarely metastases may occur in atypical and malignant neoplasms, a careful follow-up after radical resection is recommended.

Key words: myopericytoma, thigh, surgery

Introduction

Myopericytoma (MP) is a benign tumor that originates from perivascular myoid cells. It is composed of cells that show a myoid/pericytic line of differentiation towards perivascular myoid cells called myopericytes. The most common presentation is a well-circumscribed, slow-growing painless firm mass, composed of oval to spindle-shaped myoid-appearing cells with a striking tendency to a concentric perivascular growth (1). An intravascular variant has been reported only rarely (2, 3). MP is frequently misdiagnosed as a sarcoma.

Case report

A 23-year-old woman presented with a 6-month history of a painful, slow growing 2.1 × 3.8 cm sized mass in the deep intermuscular tissue of her right lateral thigh. He had no history of illness and / or trauma. The patient could not walk along the hill or climb stairs. Initially, she suffered from an occasional intensive pain and had a feeling that the musculature of the right thigh becomes harder following by the muscle spasm that involves the whole leg. The leg did not swell or change colour. She was taking general painkillers and topical medications that given short-term benefits. The problems become later more pronounced but still responsive to analgesics drugs. She reported waking up one day with a stiff leg and a bent knee, which she was able to stretch only afterwards a massage. The leg pain lasted all day, like muscle cramps. She performed a surgical-orthopedic consultation and a standard X-ray, then a consultation by a vascular surgeon that excluded a vascular injury. The administration of painkillers by injection in the emergency room had no effect. Upon physical examination, there was a reduction of myotatic reflexes on the right leg. Muscle strength was weaker on the right leg in general (4, 5). Needless short contraction of the right leg muscles was painful. There was no lack of sensibility. Patient controls the sphincters. The right leg musculature was slightly hypotrophic in the range of 2-3 cm comparing to left leg. There was no oedema on the extremities. Neurological finding of cranial nerves...
and upper extremities was normal. She was moving with the help of two crutches, while saving the right leg that was paretic.

**Magnetic resonance imaging**

Magnetic resonance imaging (MRI) scan of the pelvis and thighs showed a heterogeneous mass in the right thigh of oval form, hyper signalled, clearly limited, and located in the middle third of the posterolateral thigh, between the long head of the biceps femoral muscle, semitendinosus and adductor magnus muscles. The mass was very close to the thigh bone and sciatic nerve and has a diameter of 3.8 x 2.1 x 2.7 cm (L, W, AP) and an intensive contrast enhancement. A hypotrophy of the right thigh muscles in comparison to the left side, associated with hypersignals of oedema around the lesion, was noticed (Fig. 1).

**Intraoperative finding**

Tumor was approached through a midline longitudinal linear incision of the right thigh. Dissection was done between biceps femoral muscle, semitendinosus and adductor magnus muscle. Tumor was intimaly to the thigh bone and sciatic nerve. After dissected from the muscles, sciatic nerve and bone, tumor was completely removed. Tumor was well-circumscribed and encapsulated (Fig. 2).

**Histopathological findings**

The soft tissue mass was microscopically composed of spindle-shaped myoid-appearing cells in a concentric arrangement around blood vessel wall. Tumor cells were diffusely positive for vimentin and smooth muscle actin (sma), focally positive for desmin, and negative for EMA, GFAP, S100 protein, CD31 and myoglobin. There were 16 mitosis in 10 visual fields with partly noticed muscle fibres focally infiltrated with the tumor cells. The cells were round to spindled with eosinophilic cytoplasm and indistinct borders. Among the tumor cells, there were small fields of necrosis. From these findings, a diagnosis of myopericitoma was made. As the tumor shows a myoctic infiltration and mitosis, according to the nowadays criteria, it belongs to the malignant tumors of this histological category (Fig. 3).
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Discussion

The term myopericytoma (MP) was adopted by Granter et al. in 1998 (4) to describe a tumor that was closely related to myofibroma, with a distinctive perivascular arrangement of lesional oval to spindle cells in a concentric multi-layered pattern (5). MP has also been proposed as the term to encompass the entities myofibromatosis, adult myofibroma, glomangiopericytoma and infantile haemangiopericytoma (1, 4, 6, 7). These tumors often show overlapping histological features and are believed to be part of a spectrum of lesions that show apparent differentiation towards myopericytes (6-8).

The novel concept of the existence of myopericytes was originally proposed by Dictor et al. (10) in a report describing a tumor that involved the thyroid gland of a 5-year-old boy. This tumor showed histological features reminiscent of myofibromatosis and hemangiopericytoma. Based on immunohistochemical analysis and electron microscopy, Dictor et al. proposed that the lesional cells included a population of cells that they termed ‘myopericytes’ (10). The authors also suggested that myopericytes were the constituent cells in infantile myofibromatosis.

The most common anatomic setting for this tumor is the skin and the superficial soft tissues in adult patients (11, 12). The distal extremities are frequently involved, but with increased recognition, a wider distribution has been described (1, 8). In a comprehensive study of 54 cases, Mentzel et al. (13) found that the lower extremities were most commonly affected, followed by the upper extremities, the head and neck region, and the trunk. Mainville et al., in 2012 (8), reported a tumor in the left atrium, and Song et al. (14) as multiple pulmonary nodules. Other localizations are presented as well (15-18). The most common presentation is a well-circumscribed and slow-growing painless nodule, although occasional cases are painful (6, 8).

Figure 3. A-D Immunohistochemical staining of pathological sections. The tumor cells in myopericytoma are diffusely positive for vimentin and SMA, focally positive for desmin; G-I The presence of round to spindled cells with eosinophilic cytoplasm and indistinct borders around the numerous vessels.
Most cases of MP are benign lesions, although a few recurring and/or malignant cases have been described (8, 19-21). Maiville et al. described metastasis of primary atrial tumor (13).

The clinical outcome of rare malignant myopericytoma seems to be strongly associated with the depth of the neoplasm. However, the study of more cases with expanded follow-up are necessary to substantiate this hypothesis (6).

In our case, we present a deep located mass with some malignant histopathological features of necrosis and infiltration, so a carefully MRI follow-up was needed even after radical surgical resection. The first control MRIs were performed after 3 months, and then after 6 months; subsequently once a year. During the 6 years of clinical and radiological follow-ups, the patient had no recurrences nor pain.

The differential diagnosis of lesions includes a number of tumors that can have a perivascular arrangement such as glomus tumors and angioleiomyomas (6-8, 21). MP is a recently delineated benign neoplasm, but the presence of infiltration and necrosis may suggest a malignant feature. Most cases of myopericytoma behave in a benign fashion. However, as local recurrences and rarely metastases may occur in atypical and malignant neoplasms, a careful follow-up after radical resection is recommended.

Conflict of interest

The Authors declare to have no conflict of interest.

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