A Rare Tumor in a Young Woman: Myopericytoma

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ABSTRACT: Myopericytoma is a soft tissue tumor arising from perivascular cells called pericytes. They mostly localized in lower extremities with a middle-aged male predominance. Here, 22 year-old woman was presented with a mass lesion on the medial part of the distal left arm. The mass was painful and had grown progressively over two years. Magnetic resonance imaging demonstrated a lobulated, contrast enhanced mass. The lesion was treated surgically and final diagnose of the specimen upon pathology was a myopericytoma. Myopericytomas are uncommon, benign perivascular neoplasms. But local recurrences and rarely metastases including visceral organs can result in more unfavourable conditions. Thus, early diagnose and marginal excision is important in those patients.

Key words: Excision, myopericytoma, surgery, tumor

I. INTRODUCTION

Myopericytoma is an uncommon tumor that arises from perivascular cells adjacent to capillaries and vascular smooth muscle cells called myopericytes. It is usually asymptomatic, slow-growing and benign natured, although malignant variant has been described (1). It usually localizes in lower extremities in a middle-aged male patients (2). The etiology is unknown, but its association with trauma was shown by Laga et al (3). In this study, it was aimed to present a young female patient with myopericytoma occured in upper extremity.
II. CASE

Previously healthy, a 22 year-old female patient admitted to hospital with a mass lesion on the left arm. In detailed history it is learned that, initially it was a spot lesion but had grown progressively over two years and became painful. Lesion was 2x4 cm-sized and located on the medial part of the distal left arm (Fig 1).

Figure 1: Intraoperative image of myopericytoma

There was no abnormality in laboratory findings. Doppler ultrasound revealed a hypoechoic, solid lesion consist of arterial and venous structures. Magnetic resonance imaging depicted a hypervascular and well-circumscribed soft tissue tumor with imaging characteristics of a sarcoma. After informed consent the lesion was treated surgically and final diagnose of the specimen according to pathologic examination was myopericytoma, a benign smooth-muscle cell neoplasm. Patient was discharged two days after operation without any problem. In six month follow up period there was no recurrence.

III. DISCUSSION

Myopericytomas are rare, usually benign-natured neoplasms that show a vascular pattern like hemangiopericytoma. It commonly arises from skin or superficial soft tissues in adults but may occur at any age (4). Metzel et al. revealed that lower extremities were mostly affected, however the upper extremities, the head and neck region and the trunk can also be affected (5). Typical clinical features of myopericytoma are; single, painless, well-circumscribed and slow-growing solid mass, but unusual intravascular myopericytomases are usually painful (6).

This case showed a painful mass on the upper extremity although not have been invased to vascular structures. Etiology of the myopericytoma is not clear. Laga et al. reported an association between trauma and myopericytoma (3).
In recent studies it is showed that BRAF gene mutations are present in 15% of benign myopericytomas and 33% of which were infiltrative or recurrent (1). Despite the benign character of the myopericytoma, a few recurring and/or malignant cases have been described within the unusual locations like thorax, lungs, heart, gastrointestinal tract, brain and urinary tract and these clinical situations may lead to aggressive operations like marginal excision, radical nephrectomy and so on (1).

According to literature, magnetic resonance imagings (MRI) and ultrasonographic signs are found to be insufficient for the diagnose. Definitive diagnose is required excisional biopsy and histological examination (7). In this case, ultrasonography report revealed a lymphatic nodule and MRI revealed a sarcoma as a preliminary diagnose, but accurate diagnose was myopericytoma according to pathology result. Definitive treatment is the surgical excision of the mass. However it must be kept on mind that inadequate and inappropriate surgical interventions may lead to recurrences (8).

IV. CONCLUSION

In conclusion, in spite of the benign behaviour of the myopericytoma, malignant forms were identified in the literature with dramatic course. So early diagnose and complete excision is important in those patients.

REFERENCES
