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Malignant Myopericytoma of Left Supraclavicular Fossa: A Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. Author SKVG wrote the manuscript and Author IS provided the pictures of pathology slides. Author SMMN reviewed and edited the final draft of the manuscript. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Introduction: Myopericytoma is a rare soft tissue tumour grouped under perivascular tumours an characterized by the circumferential arrangement of perivascular cells around blood vessels. Ma myopericytoma is an extremely rare occurrence.
Case Presentation: A 44 year old lady presented with a supraclavicular lump for 5 months duration.The lesion was excised and the histology revealed an unencapsulated lesion with perivascular arrangement of spindle cells with nuclear pleomorphism and high Ki-67 index suggestive of a malignant myopericytoma.
Discussion: Myopericytomas are benign tumours that occur in skin and subcutaneous layers of the extremities in middle-aged patients. Myopericytomas are often diagnosed following excision as there is no pathognomonic radiological features. These are unencapsulated lesions and consist of concentric oval to spindle shaped cells that characteristically grow around blood vessels in an "onion skin" pattern. Immunohistochemical staining is positive for Alpha smooth muscle actin (ASMA) and H caldesmon. The malignant potential is identified from high cellularity, the number of mitoses, nuclear atypia, Ki 67 value.

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Conclusion: Due to its rarity, myopericytomas impose diagnostic difficulties and it should be considered in the differential diagnosis of hypervascular lumps in the neck. Malignant myopericytomas are aggressive and surgical excision with a wide margin is the treatment of choice.

Keywords: Malignant; Myopericytoma.

1. INTRODUCTION

Myopericytoma is a rare soft tissue tumour grouped under perivascular tumours in World Health Organization (WHO) classification of soft tissue tumours 2002 and is characterized by the circumferential arrangement of perivascular cells around blood vessels [1]. Malignant myopericytoma is an extremely rare occurrence.

2. PRESENTATION OF CASE

A 44-year-old lady presented with a painless gradually enlarging lump over left supra clavicular region for 5 months. It was not associated with fever, night sweats, recent weight loss or neurological symptoms. On physical examination, the lump was deep to the fascia with no evidence of lymphadenopathy. Her respiratory and abdominal examination was unremarkable. Full blood count, C-reactive protein, ESR and, blood picture was within normal limits.

Ultrasound Scan of the neck showed a 5cm * 5cm * 5.5cm vascular lesion. Due to the possibility of a vascular lesion, a computed tomography (CT) scan was performed and showed a lobulated non-enhancing relatively hypodense soft tissue mass measuring 6cm * 6cm * 5.4cm, causing a mass effect on adjacent intercostal muscle with no features of infiltration (Fig. 1 &2). In CT it was suspected that the lesion was arising from a peripheral nerve. An ultrasound-guided core needle biopsy revealed a spindle cell neoplasm. The lesion was completely excised with a supraclavicular incision. The patient had an uneventful post-operative recovery.

The excised mass measured 90x70x50mm with solid whitish cut surfaces. Histology revealed a non-encapsulated spindle cell lesion composed of nodules of spindle cells centered around blood vessels. Some areas of the tumour was composed of solid sheets of tumour cells showing moderate to marked nuclear atypia and focal areas of necrosis. Small tumour cell clusters separated from main tumour were identified in the periphery of the tumour. The spindle cells were arranged around blood vessels which showed thick hyalinised walls and "haemagiopericytoma like" appearance (Fig. 3). Deposition of hyaline material was noted in the periphery of the neoplastic cell nodules. The Nuclei showed mild to moderate pleomorphism. The neoplastic cells were negative for Pancytokeratin, CK7, CK20, EMA, HMB 45, CD3, GFAP and CD34. SMA showed diffuse strong positivity. Desmin was focally positive. Ki 67 index was 60% and positivity was marked in the perivascular area (Fig 4).

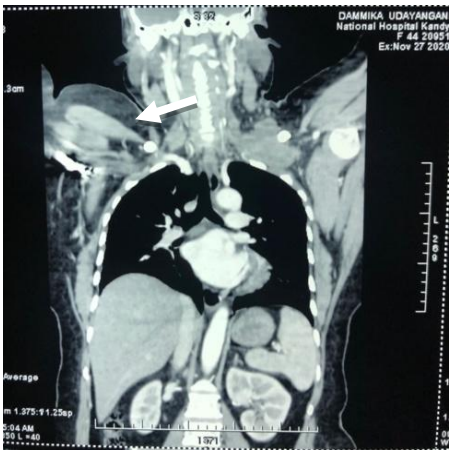

Fig. 1. Coronal image of CT Scan

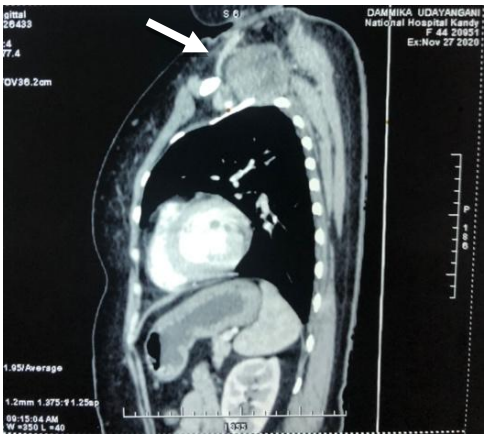

Fig. 2. Sagittal section of CT Scan

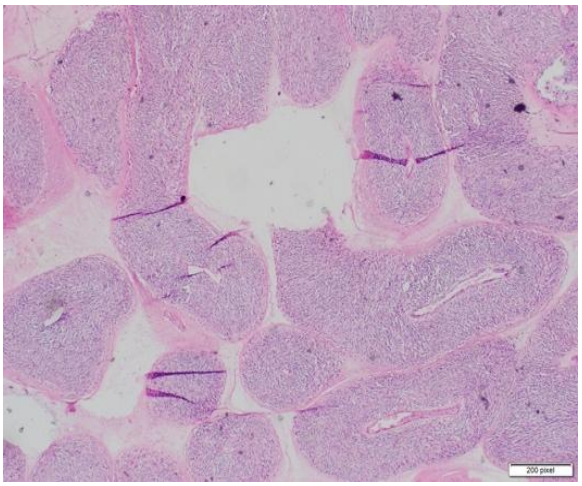

Fig. 3. H&E stain

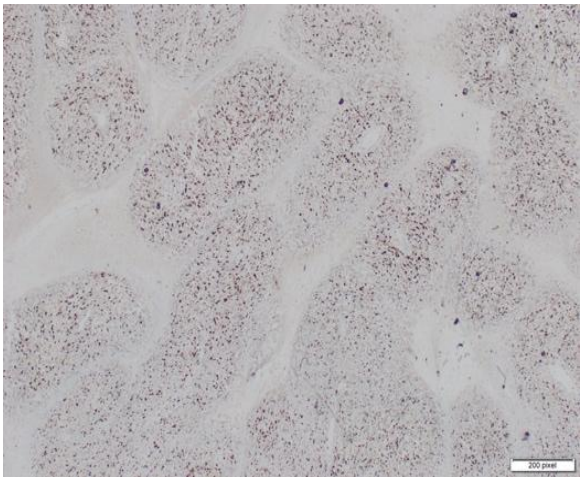

Fig. 4. Ki 67 index

3. DISCUSSION

Pericytes were initially described by Zimmerman as cells adjacent to capillaries and these are pluripotent cells that can differentiate into smooth muscle cells, adipocytes, and osteoblasts [2]. Neoplasms arising from pericytes were first described by Stout and Murray as hemangiopericytomas [2,3].

Hemangiopericytoma has numerous branching blood vessels, which is a morphologic pattern shared by other mesenchymal tumors as well. As a result, in the past mesenchymal neoplasms of different lines of differentiation have been included under the term “Hemangiopericytoma”, including myopericytomas. Myopericytomas were grouped under pericytic/perivascular tumours in the World Health Organization classification of Soft tissue tumours 2002 [1]. These tumours show differentiation towards contractile perivascular cells and grow in a characteristic circumferential pattern around blood vessels. Unlike hemangiopericytoma, myopericytoma is a true pericytic lesion and it has a close relationship to myofibromatosis, glomus tumour, and infantile haemangiopericytoma, and these tumours form a single morphological spectrum of tumours that show pericytic differentiation [1,4].

Myopericytomas are benign tumours that tend to occur in middle-aged patients. It commonly occurs in the skin and subcutaneous layers of extremities, followed by the head, neck, and trunk [2,5]. However, it can arise in unusual sites like kidneys, thoracic spine, lungs, and brain [6,7]. Most myopericytomas are single lesions but there are reports of multiple lesions occurring especially after trauma [8]. Malignant myopericytomas are an extremely rare entity and so far, there is one reported case of a malignant myopericytoma arising from the neck in a female who had a lentigo maligna excised at the same location [4]. Most lesions usually present as painless gradually enlarging subcutaneous nodules or masses, but they can be painful and present with neurological deficits when found close to a nerve [1,6,9].

Myopericytomas are often diagnosed following excision as there is no pathognomonic radiological features. An ultrasound scan may reveal a well-circumscribed hypoechoic lesion with marked internal vascularity demonstrated on colour doppler [10]. These lesions appear as hypo attenuated areas in non-contrast CT scans and they become enhanced in contrast-enhanced CT (CECT). However, in this case, the CECT did not show enhancement of the lesion. An image-guided core biopsy can be helpful in preoperative diagnosis but the high vascularity of the lesion may limit its applications. As in the index case, the core biopsy may reveal only spindle cells without demonstrating the characteristic perivascular arrangement of cells. Nevertheless, there are reports of ultrasound-guided core biopsy being safely performed in myopericytomas to obtain preoperative diagnosis [10].

Morphologically these are well-circumscribed lesions and are not capsulated. Myopericytomas consist of concentric oval to spindle-shaped cells that characteristically grow around blood vessels in an “onion skin” pattern. The cells have an eosinophilic or amorphous cytoplasm [1,3].

Immunohistochemical staining is positive for Alpha smooth muscle actin (ASMA) and H caldesmon. Occasionally tumours may show focal desmin positivity. This tumour is negative for S100, CD 31, and CD 34 [1,11]. Malignant myopericytomas resemble some morphological and immunophenotypic characters of benign myopericytoma. The malignant potential is identified from high cellularity, the number of mitoses, nuclear atypia, Ki 67 value, and some tumours may exhibit necrosis, lymphovascular invasion, and perineural invasion [7,9,12]. The index lesion had a perivascular arrangement of spindle cells with nuclear pleomorphism and a high Ki-67 index suggestive of a malignant myopericytoma.

Myopericytomas have a recurrence rate of about 10%-20% after excision which can be due to inadequate wide excision of tumour [9,13]). The tumour has cords of extension beyond the main lesion that can remain and proliferate later and some speculate that myopericytomas are a multifocal disease and recurrences are merely new primary lesions rather than true recurrences [9].

Malignant myopericytomas are aggressive and can cause extensive metastases [4,12]. Surgical excision with a wide margin is the treatment of choice which can be followed by chemotherapy or radiotherapy [6,7]. These patients need stringent follow-up for local recurrences and metastasis [4,12].

Due to its rarity, myopericytomas impose diagnostic difficulties and it should be considered in the differential diagnosis of hypervascular lumps in the neck like metastasis of papillary thyroid cancer, ectopic thyroid tissue, sarcoma, and Castleman disease of hyaline vascular type [10,13].

4. CONCLUSION

Malignant myopericytomas are aggressive and can cause extensive metastases. Surgical excision with a wide margin is the treatment of choice. Due to its rarity, myopericytomas impose diagnostic difficulties and it should be considered in the differential diagnosis of hypervascular lumps of neck.

CONSENT

All authors declare that ‘written informed consent was obtained from the patient for publication of this case report and accompanying images.

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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