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An unusual infraclavicular mass in the thoracic compartment.
Literary Review

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An unusual infraclavicular mass in the thoracic compartment. Literary Review

Haemangiopericytoma (HPC) is a rare vascular tumor comprising 1% of all vascular neoplasms and was first described by Stout and Murray in 1942. They are highly vascularized tumours located in any part of the body. Malignant HPCs represent <1% of all vascular tumours and around 5% of all sarcomatous tumours. The majority has a relatively indolent behaviour with presenting symptoms being vague for several months and not specific. Surgical excision is the mainstay of treatment. We present the case of a 65-year old male with HPC of left infraclavicular region with no associated lymphadenopathy. Surgical management included en bloc excision. The patient did not require any adjuvant therapy and showed no signs of recurrence at 1-year follow up.

KEY WORDS: Haemangiopericytoma, Extracranial, Surgical excision, Vascular tumor

Introduction

Hemangiopericytoma (HPC) is a rare vascular tumor comprising 1% of all vascular neoplasms and was first described by Stout and Murray in 1942 as a neoplasm derived from the pericytes of Zimmerman [2]. Since 2002 haemangiopericytoma have been reclassified as solitary fibrous tumour (SFT). One-third of these lesions occur in head-and-neck region [1]. Clinical presentation of conventional HPC is nonspecific. Pain is a late symptom associated with an enlarging mass. Although it is a very evident like brain neoplasia, the site described by us is not reported in the literature. We report in particular after a thorough review of a rare case for the location site a rare case for the location site a case of HPC of the infraclavicular region in a 65-year-old male patient. We performed the bibliographic search for the cases of hemangiopericytoma from 2010 to 2020 the search on the PUBMED database gave results with the key ''hemangiopericytoma'' 1,091articles. More than half of the articles are given by brain localizations, see the great work of metanalysis by Abhimanyu Ghose and other authors who talk about brain localizations (13).

A 65-year old male presented with a 3-month history of a rapidly growing, non-tender lump on his left chest without any systemic symptoms He had fractured his left clavicle during a road traffic accident 20 years before but was otherwise well. On examination there was a firm, fixed 10 x 7 cm lump in the left infraclavicular region, with no associated lymphadenopathy. (Fig. 1). Computerised tomography (CT) of the neck demonstrated a solid formation with necrosis infiltrating trapezius and pectoral muscles with necrotic tissue on core-biopsy (Fig. 2). The patient consequently proceeded to definitive surgical excision (Fig. 1B).

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Histopathologic findings

Histopathology demonstrated densely cellular proliferation with a rich branched vascular network and variable caliber vessels with a 'staghorn' pattern.
Immunohistochemistry was strongly positive for EMA and CD34. A diagnosis of haemangiopericytoma was made. (Fig. 1C, 1D)
In view of complete surgical excision, the patient did not require any adjuvant therapy and showed no signs of recurrence at 1-year follow up.

Discussion

Haemangiopericytoma is a rare mesenchymal tumour [1], arising from capillary pericytes with malignant potential and often occur in adulthood particularly between 50 years to 60 years of age [3]. They are solid and characterized by the proliferation of oval- and spindle-shaped pericytic cells around endothelial-lined vascular channels.[8] Haemangiopericytoma tend to grow slowly and the usual clinical symptom is an asymptomatic solitary mass.[4] On radiological imaging, CT scan shows a well demarcated solid mass [5]. Radiological differential diagnoses include liposarcoma, leiomyosarcoma and malignant fibrous histiocytoma. Percutaneous biopsy is not recommended because of the high risk of bleeding [7]. The distinction between benign or malignant hemangiopericitoma is based on clinical and pathological criteria: big size (>5 cm), high mitotic index, high cellularity and the presence of necrosis or haemorrhage are associated to development of recurrence and metastasis [6]. The incidence of regional metastasis from HPCs is low, and the most patients have local relapse or distant metastasis. The treatment of choice is complete resection with negative surgical margins [9,10]. Lymph node metastases are rare. The definitive diagnosis is anatomopathological after analysing the surgical specimen: CD34 immunoreactivity has been reportedly revealed to be strongly expressed in many cases of HPC; vimentin, keratin, SMA, epithelial membrane antigen (EMA), desmin, CD117, and S-100 protein are sometimes useful for differential diagnosis of HPC from tumors with muscle, epithelial, or neural origin [13]. The prognosis is good if complete resection is achieved, with five- and 10-year survival rates between 70 and 80% [11]. Adjuvant therapy including radiation therapy or chemotherapy has no established role, although it is considered to be a relatively radioresistant tumor [12].

Fig. 1: Left-sided infra-clavicular mass.

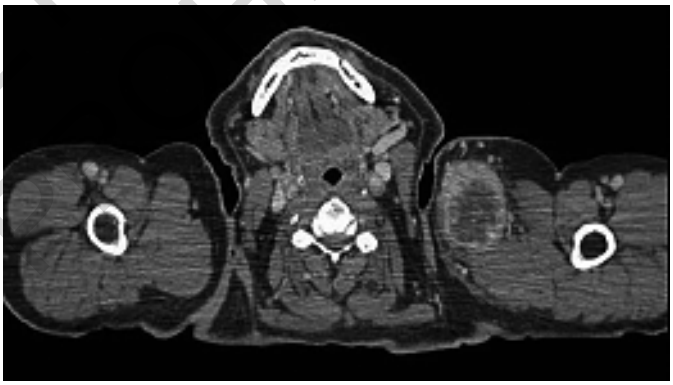


Fig. 2: The solid formation with necrosis infiltrating trapezius and pectoral muscles on CT neck in the left subclavian region.

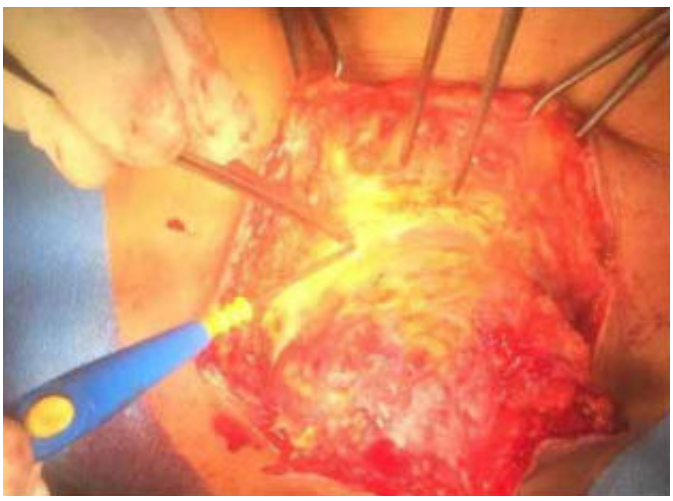
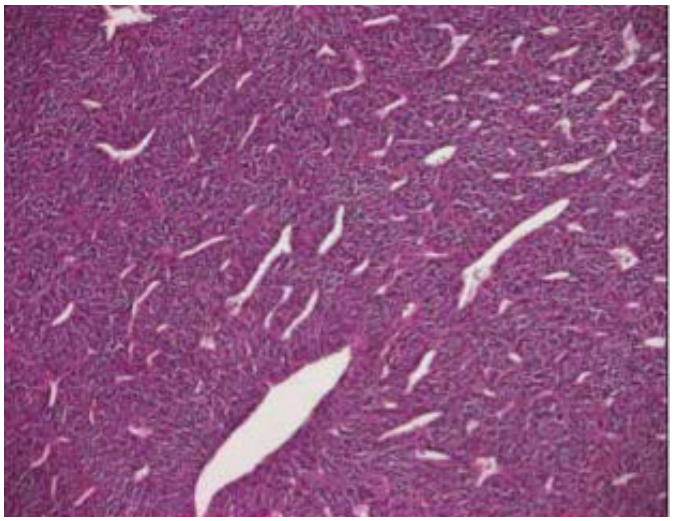
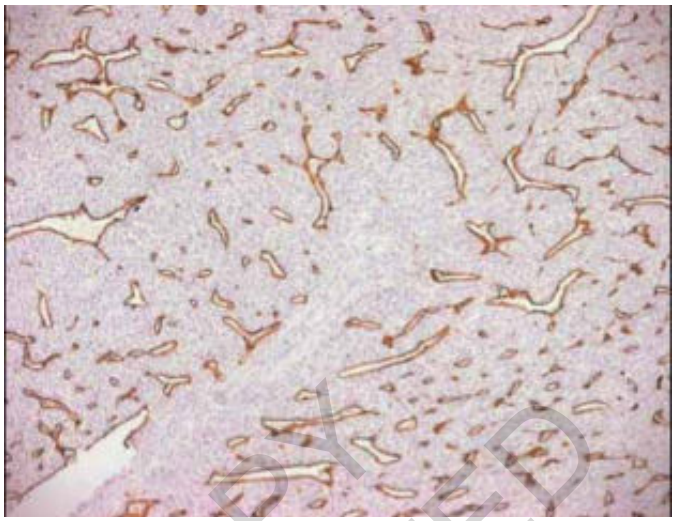


Fig. 3: Surgical excision of the mass.



A)



A)

Fig. 4: A) istopathological specimen (X10 magnification) demonstrating a rich vascular network with vessels demonstrating a 'staghorn' pattern; B) immunohistochemistry demonstrating positive endothelial staining (brown discoloration) for CD34 (X5 magnification).

Riassunto

Riportiamo un caso di tumore composto unicamente da cellule di Leydig derivate da cellule stromali ovariche e non da cellule di Leydig dell'ilo preesistenti, causa di disordine sessuale e irsutismo in menopausa.
Il tumore stromale ovarico composto esclusivamente di cellule stromali di Leydig è raro, < 0,4% tra tutti i tumori ovarici. Può verificarsi anche in donne giovani, ma quasi sempre, si verifica in menopausa o in postmenopausa. Riportiamo un caso di tumore ovarico delle cellule di Leydig nel midollo lontano dall'ilo, derivato da cellule stromali ovariche piuttosto che da cellule di Leydig dell'ilo preesistenti, con sintomi clinici complessi e non comuni. In questa relazione, sottolineiamo a) l'importanza dell'anamnesi, dell'osservazione e della considerazione di tutti i sintomi clinici, b) il valore dell'ecografia transvaginale per la diagnosi, e c) il ruolo del psicologo nel sostenere la paziente interessata nel suo disagio esistenziale e sociale.

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