Pleomorphic hyalinizing angiectatic tumor of soft parts involving the upper limb

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Received: 4 September 2009 / Accepted: 26 October 2009
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Abstract Pleomorphic hyalinizing angiectatic tumor (PHAT) of soft parts is a low grade, rare, soft tissue tumor which commonly occurs on the lower extremities of adults of either sex, though lesions in other anatomic locations have been described. We present a case of a 65-year-old female patient who presented with a left forearm swelling and underwent a wide local excision. Histopathology revealed it to be a PHAT.

Keywords Pleomorphic hyalinizing angiectatic tumor · Soft tissue neoplasms · Extremities · Excision

Introduction

PHAT of soft parts is a rare entity described first in 1996 by Smith et al. [1]. To date, only a few cases have been reported in the literature, with lesions typically occurring on the lower extremities of adults of either sex. Other rarer sites involved are arm, shoulder, axilla, buttock and buccal mucosa [2]. As this tumor has a potential for local recurrence, a wide local excision is usually indicated.

Case report

A 65-year-old female patient reported with complaints of swelling in the left forearm of 13 years duration. It was insidious and small to start with and remained so for approximately 10 years. She noticed a gradual increase in size over 3 years associated with dull ache on and off. There were no other symptoms. General examination was unremarkable. On local examination, a solitary, non-tender, mobile, non-pulsatile, firm, 5 × 4 cm globular swelling was seen on the lateral aspect of left forearm, 3 cm proximal to the wrist joint (Fig. 1). The radial pulse was normally felt. A clinical diagnosis of a nerve sheath/soft tissue tumor was considered. Preoperatively, an arterial doppler was done and was found to be normal. The patient was taken up for a wide local excision and frozen section. Intra-operatively the lesion was found to be encapsulated, highly vascular and was free from the surrounding structures and could be excised with ease. Cut section showed a fleshy tumor with areas of hemorrhage and necrosis. A frozen section could not give a conclusive diagnosis. Microscopy (paraffin section) revealed ectatic vascular channels lined by endothelial cells, occasional thrombosis and perivascular eosinophilic fibrin material. The surrounding fibrous

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Fig. 1 Swelling over the left forearm above wrist joint

stroma showed dense hyalinization with lymphocytes, foam cells and hemosiderin laden macrophages. Focal cellular areas composed of spindle cells, few bizarre cells with pleomorphic nuclei and scant mitosis were also seen (Fig. 2). Differential diagnoses included neurilemoma

Fig. 2 Photomicrograph showing ectatic vascular channels surrounded by fibrous hyalinized stroma with lymphocytes and foam cells. (H & E, $\times 100$)

and PHAT. Immunohistochemically the tumor was S-100 negative and hence a diagnosis of PHAT was considered. Patient had an uneventful recovery and was discharged. She has been kept under close follow-up and there has been no recurrence in the last 5 years.

Discussion

PHAT is a non-metastasizing soft tissue tumor, occurring within the superficial subcutaneous tissues and muscles. It occurs in adults between the ages of 10 and 83 years with a median of 51 years and is seen more commonly in females [3]. Most of the patients present with slowly growing painless masses. The commonest site to be involved is the lower extremity. Rarer sites include arm, chest wall, axilla, popliteal fossa, buttocks inguinal region, perineum, buccal mucosa and breast [2]. PHAT might be misdiagnosed as other soft tissue lesions because of its rarity. Malignant fibrous histiocytoma and neurilemoma should be considered as differential diagnosis, as cellular pleomorphism and ectatic, hyalinized blood vessels with infiltration of variable chronic inflammatory cells could be present in above tumors [2–5]. Characteristic features of neurilemoma like palisading of nuclei with formation of Verroca bodies are absent in PHAT. Immunohistochemically, PHAT is strongly positive for CD34, VEGF, CD99 and negative for S-100 protein [1, 5]. Surgical excision with tumor-free margin is the treatment of choice for this lesion as it has a considerable potential for local recurrence [2]. Low-dose radiotherapy could be useful in cases of incomplete resection to avoid recurrence and to-date, there has not been any documentation of metastasis [1]. Hence wide local excision and close postoperative follow-up are the best therapeutic approaches.

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