

A Subcutaneous Pleomorphic Hyalinizing Angiectatic Tumor of Soft Parts of the Right Ankle: A Case Report

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Abstract- Pleomorphic hyalinizing angiectatic tumor (PHAT) is a rare, slow-growing mass, which is locally aggressive and classified as tumors of borderline malignancy. In this study, we report a case of subcutaneous ankle mass in 43-year-old female since 2 years ago with a pathological diagnosis of PHAT, characterized by clusters of ectatic, fibrin-lined, and thin-walled vessels which are surrounded by a spindled or pleomorphic neoplastic stroma.

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Introduction

Pleomorphic hyalinizing angiectatic tumor (PHAT) is a rare, locally aggressive tumor that typically occurs in the subcutaneous tissues of the distal extremities (1-5) and is first reported by Smith *et al.*, in 1996 (6).

PHATs typically present as slow-growing masses, which are occasionally mistaken for hematomas or Kaposi sarcoma. These tumors are slightly more common in women than men. As PHATs frequently recur locally (50% local recurrence rate), they are classified as tumors of intermediate (borderline) malignancy (6-14).

It is characterized by clusters of ectatic, fibrin-lined, thin-walled vessels which are surrounded by a spindled or pleomorphic neoplastic stroma that contains a variable inflammatory component. The tumor cells are similar to those of malignant fibrous histiocytoma but differ from them by having a low mitotic figure, intranuclear cytoplasmic inclusion, and CD34 expression. The peculiar vascular pattern, low mitotic figure, intranuclear cytoplasmic inclusion and mast cell infiltration are also reminiscent of neurilemmoma. However, PHAT usually shows an infiltrative border and no S-100 protein expression (15-17).

In this study, we report a case of subcutaneous ankle mass in 43-year-old female 2 years ago with a pathological diagnosis of PHAT.

Case Report

Clinical history

The patient was a 43-year-old female. She had a subcutaneous mass in her right ankle for 2 years. The mass had progressively increased in size and was circumscribed but none capsulated, and about 2.5 cm×2 cm. No pain, skin ulceration or other abnormality was noted. She had no past medical history or family history. Physical examination showed a firm and mobile mass in the right ankle without tenderness. The overlying skin was intact.

Laboratory workup was normal. Radiographic findings showed a solid heterogeneous enhancing subcutaneous mass in the right ankle (Figure 1). Excisional biopsy was performed to treat the tumor.



Figure 1. Radiographic picture

Pathologic findings

The excised lesion measured 2.2 cm*2 cm in size with variegated appearance and tan to yellowish colored on the

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cut surface with the hemorrhagic area (Figure 2). Microscopic findings showed moderately cellular spindle cell proliferation which was infiltrated fatty by tissue having pleomorphism and some intranuclear vacuolization associated with hyalinized dilated blood vessels some containing thrombosis and fibrin set within an edematous focally myxoid stroma. Mitotic activity was low and focal hemosiderin deposition was seen (Figure 3).



Figure 2. Gross picture

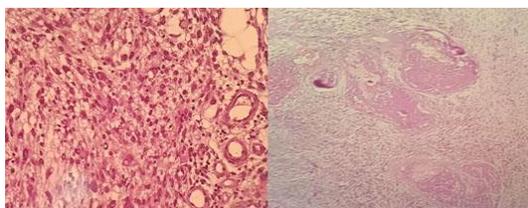


Figure 3. Microscopic picture

Immunohistochemical study showed that the tumor cells were positive for vimentin and CD 34 and negative for cytokeratin S100 desmin and SMA.

The final pathologic diagnosis was a pleomorphic hyalinizing angiectatic tumor (PHAT).

Discussion

Pleomorphic hyalinizing angiectatic tumor (PHAT) is a rare, locally aggressive tumor that typically occurs in the subcutaneous tissues of the distal extremities (1-5) and is first reported by Smith *et al.*, in 1996 (6).

PHATs typically present as slow-growing masses, which are occasionally mistaken for hematomas or Kaposi sarcoma. These tumors are slightly more common in women than in men. As PHATs frequently recur locally (50% local recurrence rate), they are classified as tumors of intermediate (borderline) malignancy (6-14).

It is characterized by clusters of ectatic, fibrin-lined, thin-walled vessels which are surrounded by a spindled or pleomorphic neoplastic stroma that contains a variable inflammatory component. In our case, Microscopic findings showed moderate cellular spindle cell proliferation which was infiltrated by fatty tissue having

pleomorphism and some intranuclear vacuolization associated with hyalinized dilated blood vessels some containing thrombosis and fibrin set within an edematous focally myxoid stroma. Mitotic activity was low and focal hemosiderin deposition was seen.

In view of its histologic features, PHAT bears similarities with other soft tissue tumors and could easily be misdiagnosed as any of them. Examples of such tumors include neurolemma, low grade malignant fibrous histiocytoma, ancient schwannoma, cellular angiofibroma, solitary fibrous tumors, and undifferentiated pleomorphic sarcoma. In order to differentiate between these tumors, a combination of pathologic examination and immunohistochemical stains are used. Most PHAT tumors that have been subjected to immunohistochemical staining have been found to stain positively for CD34 but negative for S-100 and desmin. In the index patient, the tumor was characteristically strongly positive for CD34 and negative for desmin and S-100. It also showed positive staining for vimentin which has been observed to be positive in most cases (3,15,20,21).

In our case, immunohistochemical findings showed that the tumor cells were positive for vimentin and CD 34 and negative for cytokeratin S100 desmin and SMA.

The legs and feet/ankles are the most frequently involved site. Other locations such as the upper extremities, buttocks, inguinal region, knee/patella, chest wall, shoulder, axilla, back, hands, waist, breasts, and perineum were also reported. Curative resection of the tumor usually entails wide excision with negative margins. This reduces the risk of recurrence. The published literature documented recurrence rates in the range of 33-50%. Although the majority of the cases recur as PHAT, rare recurrences with sarcomatous components have been reported. Due to the biology of these tumors, some experts recommend that they are viewed as locally aggressive, low-grade tumors. However, according to the WHO classification of soft tissue tumors, pleomorphic hyalinizing angiectatic tumors are benign tumors of uncertain differentiation (9,13,18,22).

PHAT is a rare locally aggressive tumor that typically occurs in the subcutaneous tissue of the distal extremities. We report a case of PHAT which presented as an ankle mass, and we reviewed the clinicopathologic features of this tumor. Regarding the rarity of this tumor review of all diagnosed cases will help to simplify the accurate diagnosis.

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