

Case Report

A subcutaneous pleomorphic hyalinizing angiectatic tumor of soft parts of the right chest wall: report of a rare case

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Abstract: Pleomorphic hyalinizing angiectatic tumor of the soft parts is an extremely rare mesenchymal tumor consisting of spindled and pleomorphic tumor cells and clusters of ectatic, fibrin-lined vessels. It typically occurs in the subcutaneous tissues of the distal extremities, usually the ankles and feet. Here we present a case of pleomorphic hyalinizing angiectatic tumor of the soft parts of the right chest wall in a 51-year old female. The tumor was subcutaneous, nonencapsulated, and about 2.0 cm×1.0 cm. Microscopically, the tumor was composed of numerous ectatic, fibrin-filled, thin-walled blood vessels, surrounded by spindled or pleomorphic tumor cells arranged in sheet-like or fascicular architecture, or randomly. Mitotic activity of the tumor cells was low. Immunohistochemical analysis shows that the tumor cells were positive for CD34 and vimentin, but negative for CD31, CK, desmin, EMA, HMB45, Myo D1, P63 and S-100. Ki67 index was about 1%.

Keywords: Pleomorphic hyalinizing angiectatic tumor, pleomorphic hyalinizing angiectatic tumor of the soft parts, subcutaneous, chest wall

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]. PHATs typically present as slow-growing masses, which are occasionally mistaken for hematomas or Kaposi sarcoma [5-10]. 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 [5-13]. Here we describe a case of subcutaneous PHAT in a 51-year old female in her right chest wall. 5 years ago she had a mass in the same position and the mass was resected at that time. However, biopsy and pathological examination after resection were not done. The current tumor may be a recurrent tumor of the one 5 years ago. Microscopically, the tumor was composed of spindled or pleomorphic tumor cells. Numerous ectatic, fibrin-filled, thin-walled blood vessels present in the tumor tissues.

Immunohistochemical analysis shows mainly positive staining of CD34 and vimentin. Diagnosis of PHAT was made according to the clinical and pathological findings.

Case presentation

Clinical history

The patient was a 51-year old female. She had a subcutaneous mass in her right chest wall for 1 year. The mass was circumscribed but nonencapsulated, and about 2.0 cm×1.0 cm. No pain, skin ulceration or other abnormality was complained. 5 years ago the patient had a mass in the same position and the mass was resected at that time. However, diagnosis was not clear for pathological examination was not done at that time.

Materials and methods

Specimens resected were fixed with 10% neutralbuffered formalin and embedded in paraffin blocks. Tissue blocks were cut into 4 μm-thick

Pleomorphic hyalinizing angiectatic tumor of chest wall

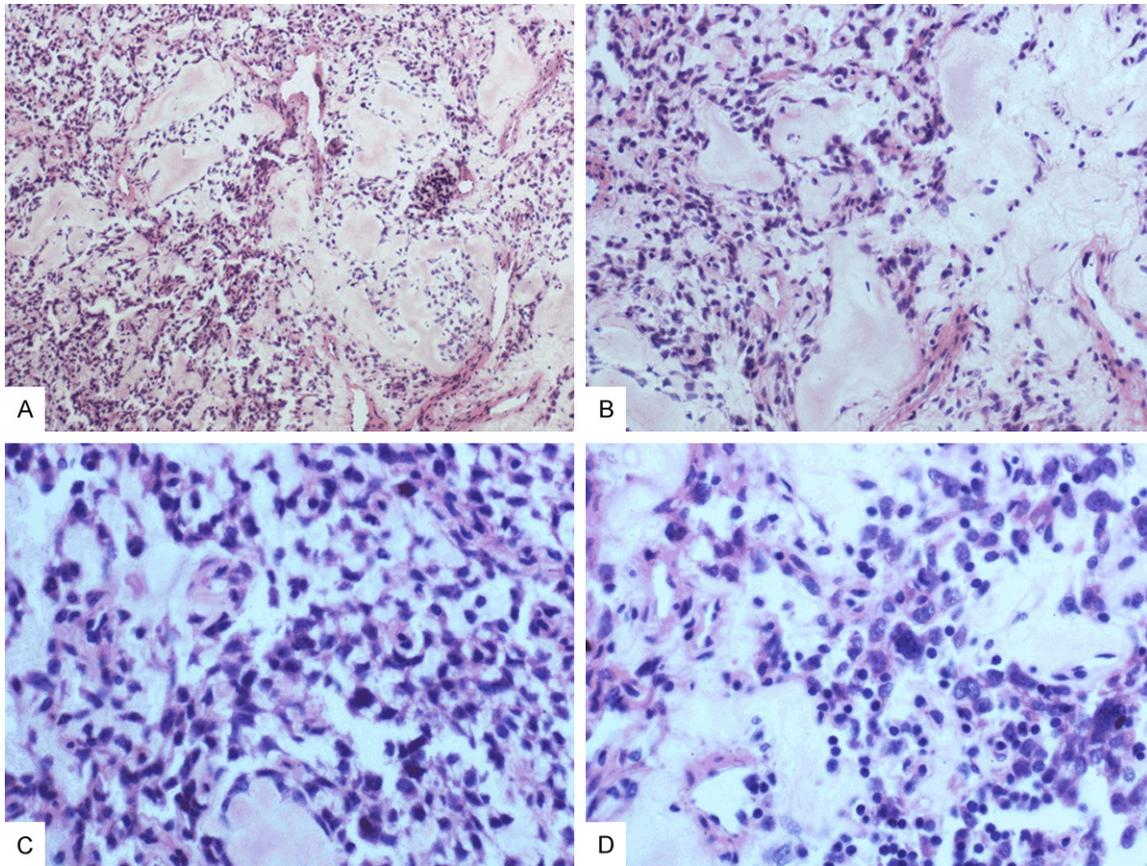


Figure 1. Microscopic findings of the tumor. Tumor tissues were cellular with spindled or pleomorphic tumor cells arranged in sheet-like or fascicular architecture, or randomly. The tumor tissues consist numerous ectatic, fibrin-filled, thin-walled blood vessels (A. $\times 100$, B. $\times 200$). Most of the tumor cells were large with plump nuclei (C, D. $\times 400$). Some tumor cells have bizarre-appearing hyperchromatic nuclei (C, D). Chronic inflammatory cell infiltrate was present in some areas the tumor tissues (D).

sections and were dewaxed in xylene and rehydrated stepwise in descending ethanol series. Then the sections were boiled in citrate buffer (pH 6.0). Endogenous peroxidase activity and non-specific binding were blocked with 3% H_2O_2 and non-immune sera, respectively. The sections were incubated with the following primary antibodies: actin-sm (1:50, DAKO), AE1/AE3 (1:50, DAKO), CD31 (1:50, DAKO), CD34 (1:100, DAKO), desmin (1:50, DAKO), EMA (1:100, DAKO), HMB45 (1:50, Abcam), Ki67 (1:200, DAKO), myoD1 (1:50, DAKO), P63 (1:100, DAKO), S-100 (1:50, DAKO), and vimentin (1:200, DAKO) overnight at 4°C. The catalyzed signal amplification system (Maixin Biotechnology, Fuzhou, Fujian, China) was used for staining of these proteins according to the manufacturer's instructions. The antibodies were detected by a standard avidin-biotin complex method with biotinylated secondary antibodies

(Maixin) and an avidin-biotin complex (Maixin), and developed with diaminobenzidine. Counterstaining was done lightly with hematoxylin, and the sections were dehydrated in alcohol before mounting.

Results

Gross features

The tumor was subcutaneous in the right chest wall and about 2 cm \times 1 cm. The mass was soft and circumscribed but nonencapsulated.

Microscopic features

Tumor tissues were cellular with spindled or pleomorphic tumor cells arranged in sheet-like or fascicular architecture, or randomly (**Figure 1**). The tumor tissues also consist of numerous ectatic, fibrin-filled, thin-walled blood vessels

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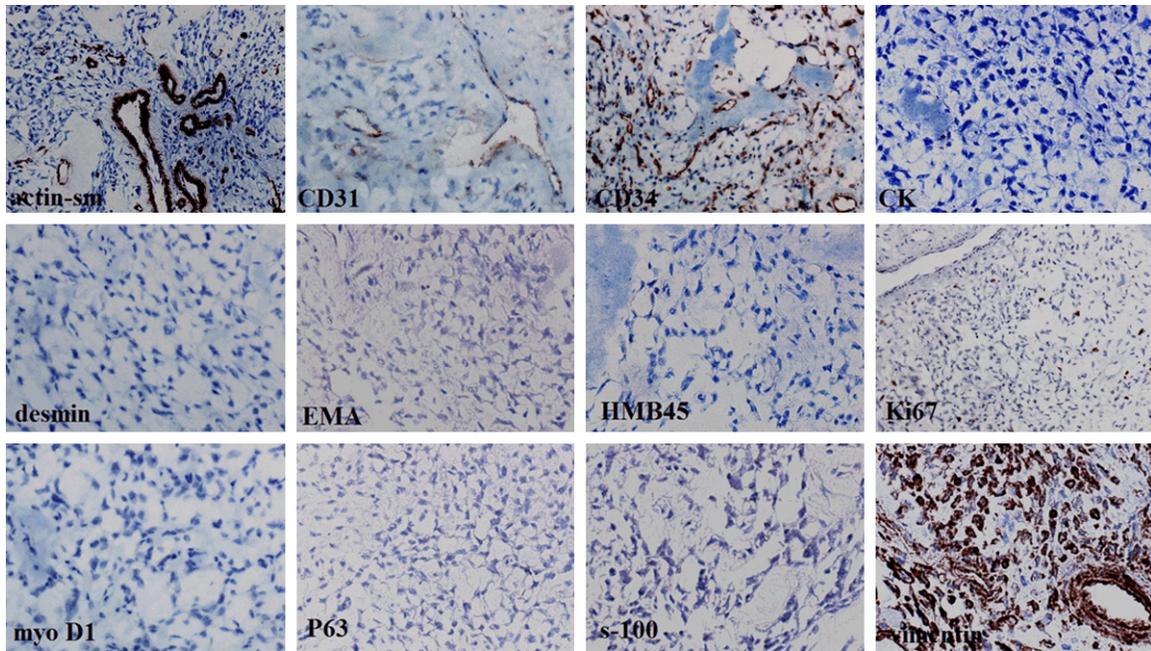


Figure 2. Immunohistochemical examination. Actin-sm and CD31 staining was seen in blood vessels, but not in tumor cells. CD34 was locally positive in tumor cells. CK, desmin, EMA, HMB45, Myo D1, P63 and S-100 were negative in tumor cells. Vimentin was diffusely positive in tumor cells.

(**Figure 1A, 1B**). Most of the tumor cells were large with plump nuclei (**Figure 1C, 1D**). Some tumor cells have bizarre-appearing hyperchromatic nuclei (**Figure 1C, 1D**). However, mitotic activity of the tumor cells was low (<1/50HPF). Chronic inflammatory cell infiltrate was present in some areas of the tumor tissues (**Figure 1D**). Nuclear pseudoinclusions were not prominent in this case.

Immunophenotype

Immunohistochemical analysis shows that the tumor cells were diffusely positive for vimentin, locally positive for CD34, and negative for actin-sm, CK (AE1/AE3), CD31, desmin, EMA, HMB-45, Myo D1, P63, and S-100 (**Figure 2**). CD31 and CD34 immunostaining also indicates abundant capillaries in tumor tissues. Ki67 index was about 1% (**Figure 2**).

Discussion

PHAT of the soft tissue is a rare mesenchymal tumor listed as a locally aggressive, nonmetastazing neoplasm in the new World Health Organization classification [5]. It was described first in 1996 by Smith et al [14]. PHATs typically occur in the subcutaneous tissues of distal

extremities. Other rarer sites involved include shoulder, axilla, buttock, renal hilum, and so on [1-5, 9]. Here we present a case of PHAT in a 51-year old female in her right chest wall. The mass was subcutaneous and circumscribed, though nonencapsulated. Microscopically, this kind of tumor is characterized by abundant angiectatic vessels with hyaline deposition in and around the vessels. Numerous ectatic, fibrin-filled, thin-walled blood vessels also present in the current case. The tumor cells of the current case were spindle or pleomorphic. Though the nuclei of the tumor cells were plump and some with bizarre shapes, mitotic activity was very low (<1/50HPF), which was different from undifferentiated high grade pleomorphic sarcoma. Prominent nuclear pseudoinclusions occasionally present in PHATs, but not in this case.

Immunohistochemical studies indicate that the tumor cells of PHATs mainly express CD34, vimentin, CD99, and VEGF [9]. According to Smith et al, CD34 expression in PHATs occasionally presents [14]. In this case, only vimentin was diffusely positive. CD34 was locally positive, but not in all tumor cells. However CD34 is not a specific marker for blood vessels. Moreover, CD31, another marker for blood ves-

sels, is negative in this case. So far, the differentiation of PHATs is not determined yet. The Ki67 index of PHATs is commonly low and was about 1% in this case. The low Ki67 index in PHAT also indicate it a non-malignancy. Main differential diagnoses include undifferentiated high grade pleomorphic sarcoma and neurilemoma as tumor cell pleomorphism and hyalinized blood vessels may present in these tumors [13, 14]. The tumor cells lack of mitotic figures, and Ki67 index was low, which was different from undifferentiated high grade pleomorphic sarcoma. CD34 was positive and S-100 was negative in this case, which was consistent with the immunophenotype of PHAT but not neurilemoma.

PHATs frequently recur locally (50% local recurrence rate) and are considered as borderline neoplasms because of their aggressive behavior [4, 5]. The patient we describe here had a mass in the same position as the current tumor 5 years ago. The mass was resected at that time. However, pathological examinations were not done so we cannot tell if they were the same. However as the masses located at the same site, the current tumor is likely a recurrent tumor of the one 5 years ago. Most of the recurrent tumors retained the typical morphological appearance of original PHAT. There are also reports of rare PHATs recurred with the appearance of sarcomas [15]. For these reasons, wide local resection with tumor-free margin for the treatment of the disease is suggested [4].

Conclusion

PHAT is a rare, locally aggressive tumor that typically occurs in the subcutaneous tissues of the distal extremities. PHATs typically present as slow-growing masses but frequently recur locally and may occasionally mistaken for hematomas or Kaposi sarcoma, which should be differentiated according to both clinical and pathological findings.

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Disclosure of conflict of interest

None.

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