PLEOMORPHIC HYALINIZING ANGIECTATIC TUMOR OF THE ABDOMINAL WALL

A case report and review of the literature

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Abstract

Pleomorphic Hyalinizing Angiectatic Tumor (PHAT) is one of rare soft tissue tumors, which is recently described. It is considered as a tumor of uncertain lineage and intermediate malignancy. This tumor occurs in adults as a slow growing subcutaneous mass and has a predilection for lower extremity, other locations are rarely reported. Here we report a case of PHAT arising primarily in the abdominal wall and review the literature. A 50-year-old male was referred to our hospital for evaluation of a slow growing mass since 6 years. A physical examination revealed a 10 x 7 cm large subcutaneous mass in the left region of the anterior abdominal wall.

Radiologic explorations revealed oval mass, well-defined, predominantly cystic with a tissue component. Wide surgical excision of the mass was done and sent for histopathological evaluation, which confirmed the diagnosis of PHAT of the soft parts.

Microscopically, the tumor was characterized by clusters of ectatic, fibrin-lined, thin-walled vessels. The cells were spindled to pleomorphic, mitotically inert and contained focally intracytoplasmic hemosiderin. They were surrounded by a neoplastic stroma that was fibrous and contained a variable inflammatory component.

Immunohistochemical study showed that the tumor cells were positive for CD34, and negative for S-100 protein, smooth muscle actin and desmin.

We present an extremely rare case of PHAT arising primarily in the abdominal wall, which is to our knowledge, the first case described. This tumour is usually diagnosed using histopathological and immuohistochemistry analysis, thereby allowing the differentiation with other soft tissue tumors.

Keywords: Pleomorphic hyalinizing angiectatic tumors, Soft tissue tumors, PHAT

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Introduction

Pleomorphic Hyalinizing Angiectatic Tumor (PHAT) was first described by Smith *et al* in 1996 [1] and since then, less than 100 cases have been documented [2]. It is considered as a rare mesenchymal tumor of uncertain lineage and intermediate malignancy [1,3]. This tumor usually occurs in adults as a slow growing subcutaneous mass and has a predilection for lower extremity [4]. Other rarer sites involved were forearm, shoulder, axilla, buttock and buccal mucosa [5]. It is histologically characterized by clusters of thin‑walled ectatic vessels surrounded by hyalinized, fibrin and collagen material [3]. Here we report a case of PHAT arising primarily in the abdominal wall which is an extremely rare location and review the literature. To our knowledge, this was the first reported case of PHAT described in that site.

Materials and methods

Clinical data

A 50-year-old male was referred to our hospital for evaluation of a slow growing mass since 6 years. A physical examination revealed a large subcutaneous mass in the left region of the anterior abdominal and no particular complain was given (Fig. 1). No lymphadenopathy was found.

Radiologic findings

Contrast Enhanced Computed Tomography (CECT) of the abdomen (Fig. 2) showed an oval mass of the anterior abdominal wall, well defined, predominantly cystic with a tissue component and septa endings heterogeneously enhanced after injection of contrast medium. This mass developed at the expense of the left muscle of the abdomen and measured 12 x 11 x 10 cm. It pushed the muscular fascia and exerted a mass effect on the left liver and the stomach with the disappearance of the safety margin in contact with the gastric anterior wall. Chest Computed Tomography (CT) showed no distant metastasis.

Wide surgical excision of the mass with a tumor‑free margin were done and sent for histopathological analysis.

Results

Gross findings

Gross examination, showed a mass of soft consistency, maroon color, which measured 15 x 8 x 7 cm. It was surmounted by a cutaneous and subcutaneous fragment that measured 10 x 4 cm. At the cut, the mass had important cystic changes and several buds, of which the largest measured 2.5 cm in diameter (Fig. 3).

Histopathological findings

Microscopically, the lesion was often delimited by a fibrous capsule and had vascular and cellular areas. Cellularity was variable and composed of hyper and hypocellular areas. The vascular areas were comprised of many clusters of ectatic, fibrin-lined and thin-walled vessels. The cells were spindled to pleomorphic, mitotically inert and contained focally intracytoplasmic hemosiderin (Fig. 4). They were surrounded by a neoplastic stroma that was fibrous or focally myxoid and contained a variable inflammatory component which was composed of lymphocytes, plasma cells, macrophages and mast cells. The tumour cells infiltrated the adjacent adipose tissue focally.

Immunohistochemical study showed that the tumor cells showed strong membranous positivity for CD34 and were negative for S-100 (Fig.5), smooth muscle actin and desmin. Based on the above features, diagnosis of PHAT was given.

Discussion:

Pleomorphic Hyalinizing Angiectatic Tumor (PHAT) is a rare soft tissue neoplasm that was firstly described by Smith *et al* in 1996 [1]. Since then, less than 100 cases have been documented and were either single case reports or small case series [2]. WHO classification of soft tissue tumors, categorized this tumor under tumor of uncertain differentiation [3]. It usually occurs in adults, with equal male and female distribution, as a slow growing subcutaneous mass and has a predilection for lower extremity, particularly in foot and ankle [4,6]. Other rarer sites involved were forearm, shoulder, axilla, buttock and buccal mucosa [5]. To the best of our knowledge, the involvement of the abdominal wall has never been reported according to literature data [1]. Thus, this is the first reported case of PHAT described in that site. Clinically, PHAT presents as a slow growing, painless mass with a size

varying from less than 1cm to more than 20 cm [7]. It may mimic several benign or malignant tumors including hematoma, lipoma, desmoid tumor, and Kaposi's sarcoma requiring thereby tissue biopsy for diagnosis [8]. Pathological study shows in the macroscopic examination that PHAT is unencapsulated, poorly circumscribed, firm mass having a lobulated appearance and a variegated cut surface with tan to maroon in color [3,5,9]. Histologically, it is composed of clusters of thin‐walled ectatic vessels, embedded in a spindled stroma and surrounded by hyalinized, fibrin and collagen material. The tumor cells are spindled to pleomorphic and contain intranuclear inclusions and fine haemosiderin granules. Mitotic activity is low or absent. The stroma may include variable amount of inflammatory infiltrate, usually mast cells are present [3]. In the series reported by Folpe and Weiss [10], the authors emphasized the identification of a pattern at the periphery of some tumors, which they termed early PHAT [11]. It represents the precursor for classic PHAT [3]. These areas are characterized by low to moderate cellularity composed of bland, hemosiderin-laden spindled cells with wavy nuclei arranged in fascicles. The cells infiltrate the surrounding adipocytes in a manner reminiscent of dermatofibrosarcoma protuberans [11]. Early PHAT may have features overlapping thereby with haemosiderotic fibrolipomatous tumor [3]. Immunohistochemically, the tumor cells express CD34 and lack S100 [3].

The two main differential diagnoses of PHAT are ancient schwannoma and undifferentiated pleomorphic sarcoma [6, 11, 12]. In fact, areas of variable cellularity with atypical spindle cells, low mitotic activity, and hyalinized blood vessels may mislead to the diagnosis of ancient schwannoma. However, PHAT is uncapsulated, lack Antoni A and B zones and formation of verrocay body. Immunohistochemistry showed negative staining of S-100 protein and strong positive staining for CD34 [6].

Although the cellular pleomorphism may be suggestive of undifferentiated pleomorphic sarcoma, it can be excluded by low mitotic activity, CD34 expression and the presence of intranuclear cytoplasmic pseudoinclusion [7].

For the treatment, it is recommended to make a wide surgical excision of the mass with a tumor‐free margin. Concerning the prognosis, according to the literature, between 30% and 50% of PHATS recur locally which can usually be controlled by re-excision [3]. The very rare cases reported showed recurrence of PHAT as a myxoid pleomorphic sarcoma [3, 11].

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Conclusion:

In conclusion, to the best of our knowledge, this is the first reported case of PHAT arising primarily in the abdominal wall. PHAT is a rare tumor that may mimic other benign and malignant tumors clinically and histologically. Thus, identifying and differentiating it from its differential diagnoses is important because of its high potential for recurrence and high grade transformation. The treatment of choice is wide local excision with free margin.

Authors' contributions

Seifeddine Ben Hammouda: designed and wrote the article. Amina Chaka and Ibtissem Korbi: specimen contribution. Manel Njima and Leila Njim: conceived and design of the article; also coordination and helped to draft the manuscript. Adnen Moussa: helped to perform immunohistochemical stains and interpretation. Ahlem Bchir: collected clinical information and prepared the figures. Khadija Zouari: helped to draft the manuscript and collect the clinical data. All authors read and approved the final manuscript.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Competing interests

The authors declare that they have no competing interests.

Acknowledgements

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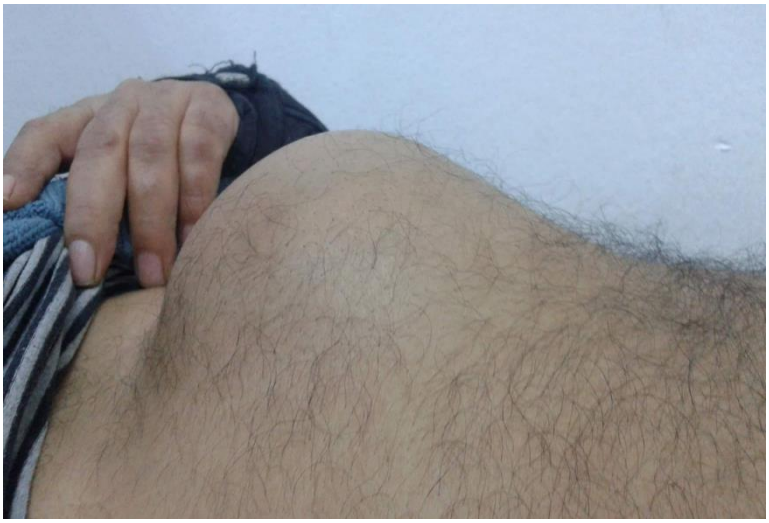

Fig. 1: Physical examination revealed a large subcutaneous mass in the left region of the anterior abdominal wall measuring 10 x 7 cm.

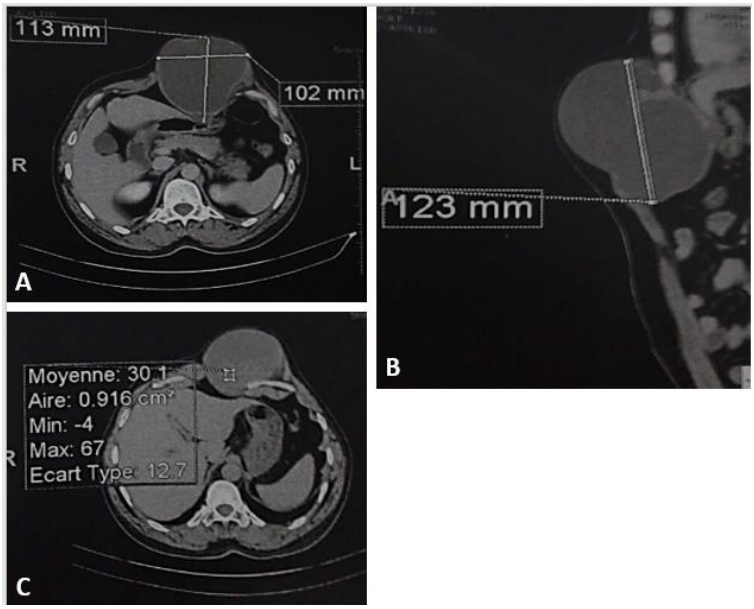

Fig. 2: (A), (B), (C): CECT of abdomen showed oval mass, well-defined, predominantly cystic with a tissue component and measured 12.3 x 11.3 x 10.2 cm.

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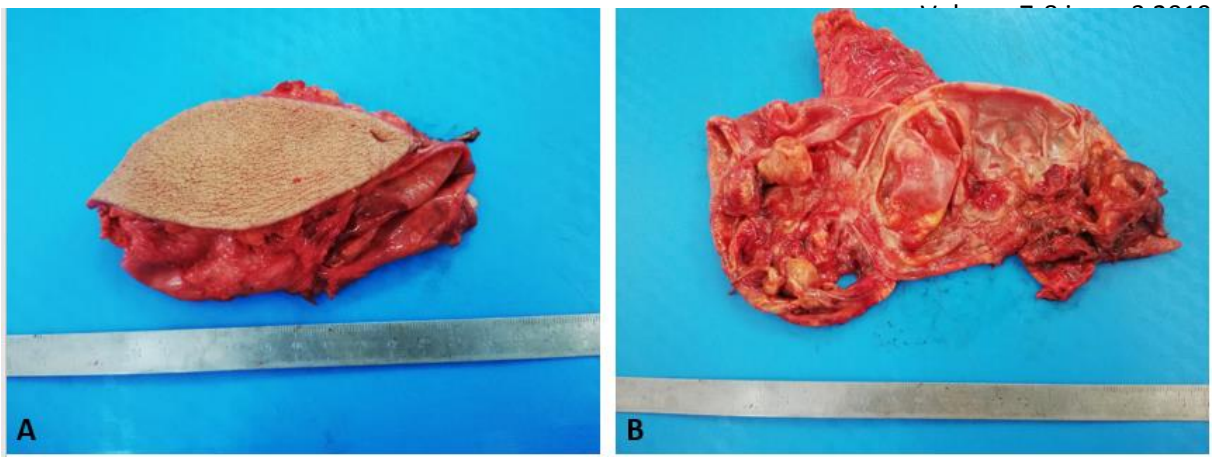

Fig. 3: (A) Gross appearance of the specimen showing oval mass covered by an intact skin. It had soft consistence, maroon color and measured 15 x 8 x 7 cm.
(B) Cut surface of the specimen shows variegated appearance with important cystic changes and several buds, of which the largest measured 2.5 cm of diameter.

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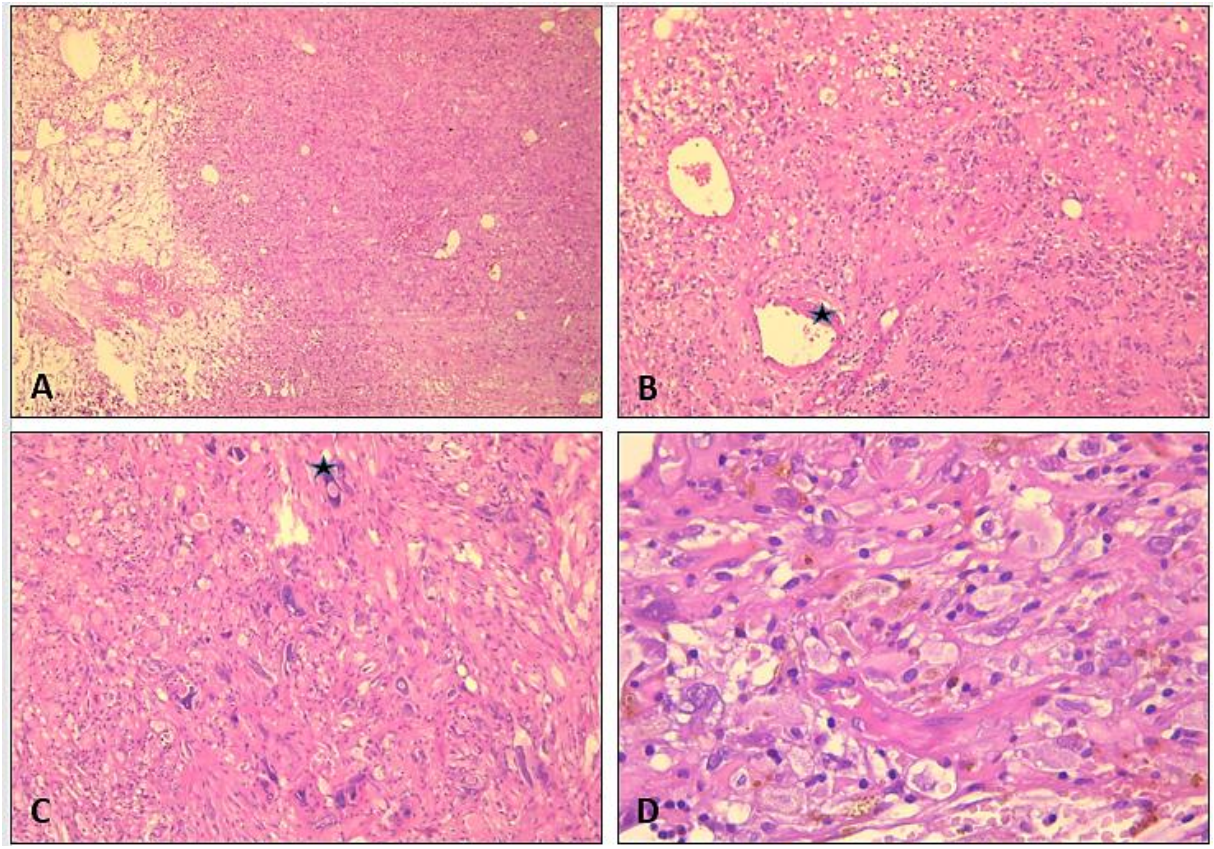

Fig. 4: Histological findings, hematoxylin and eosin stain. (A) Low power view (X40) showing spindle cell neoplasm with prominent vascular component and perivascular hyalinization. (B) Medium power view (x100) showing vascular areas comprised of many clusters of ectatic, fibrin-lined and thin-walled vessels. High power view showing spindled to pleomorphic tumor cells that contain intranuclear inclusions (C) and fine hemosiderin granules (D).

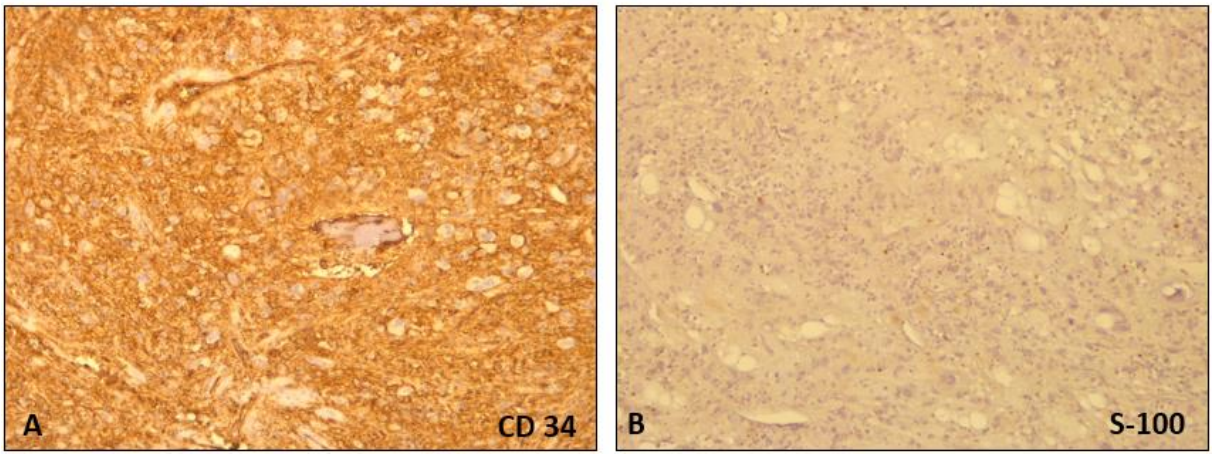

Fig. 5: Immunohistochemistry, Tumor cells showed intense positivity for CD34 (A) and were negative for S-100 protein (B) (X100).

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