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Pleomorphic hyalinizing angiectatic tumor arising in the groin: A case report

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ABSTRACT

INTRODUCTION: Pleomorphic hyalinizing angiectatic tumors are a rare group of tumors that are currently classified as benign tumors of unknown differentiation. To our knowledge, less than 100 cases have been reported in literature. We report a case that presented in the groin – an uncommon location for this rare tumor.

CASE REPORT: A 75 year-old female presented with a seven-year history of painless right groin mass with rapid growth of 2 year duration. On physical examination, a firm and mobile mass was identified in the right groin. It measured 12 cm × 8 cm, with no clinically palpable lymph nodes. Microscopic and immunohistochemical features were consistent with pleomorphic hyalinizing angiectatic tumor.

DISCUSSION: Pleomorphic hyalinizing angiectatic tumor is a rare soft tissue tumor usually diagnosed using microscopic and immuohistochemical analysis to allow for differentiation from other soft tissue tumors. It is treated by wide local excision.

1. Introduction

Pleomorphic hyalinizing angiectatic tumors are a rare group of tumors first reported 2 decades ago by Smith et al. [1,2] Since its first report, less than 100 cases have been documented [3,4]. We report a case that presented as a groin mass with a view to add to the body of evidence on this rare tumor which will guide accurate diagnosis and management.

2. Case report

A 75 year old African-American female who presented with a seven-year history of painless right groin mass. The mass had progressively increased in size over the years but more noticeably in the last 2 years prior to presentation.

She has a past medical history which is significant for arthritis, hypercholesterolemia and hypertension for which she was on Clonidine, Hydralazine, Lisinopril and Metoprolol. She also has a family history which is significant for stomach cancer in her father, lung cancer in two siblings, colon cancer in a sister, liver cancer in a brother, and skin cancer in another brother.

Pertinent finding on physical examination included a firm mass in the right inguinal region. The mass extended toward the right side of the mons pubis. It measured approximately 12 cm in its longitudinal dimension and 8 cm in transverse dimension. It was mobile – not attached to the overlying skin or the underlying structures, non-pulsatile and was not associated with tenderness or erythema. There was no ulceration or enlarged groin lymph node.

Initial laboratory work up showed normal biochemical and hematological profiles. A computerized tomography scan of the abdomen and pelvis was done and showed a 4.9 cm × 7.2 cm × 10.7 cm partly solid and cystic heterogeneous enhancing subcutaneous mass in the right inguinal region (Fig. 1). The patient proceeded to have a core needle biopsy of the tumor and pathology reported a diagnosis of cellular angiofibroma. In view of her family history and recent rapid increase in size of the tumor, a wide local excision was undertaken to treat the tumor.

Pathologic evaluation of the specimen revealed a tumor measuring 12 cm × 7 cm × 5 cm with cystic spaces measuring up to 7 cm. Sections of the mass showed moderately cellular spindle cell neoplasms arranged in a random pattern with variable sized vessels, both ectatic and small hyalinized vessels set within an edematous focally myxoid stroma and intervening thick collagen bundles. There was sparse mitotic activity. There were areas with giant cells showing marked degenerative atypia (Fig. 2). Immunohistochemistry demonstrated cells positive for CD34, vimentin, estrogen and progesterone receptors (Fig. 3) but negative for desmin, SMA, S100 and CD 117 (Fig. 4). Immunohistochemical staining for STAT6

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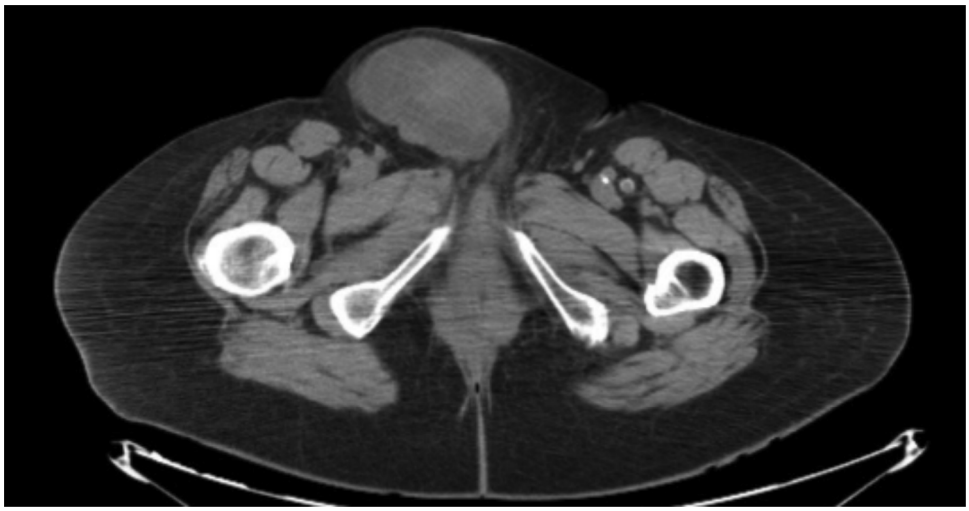

Fig. 1. CT image of pelvis showing the lesion in the right groin.

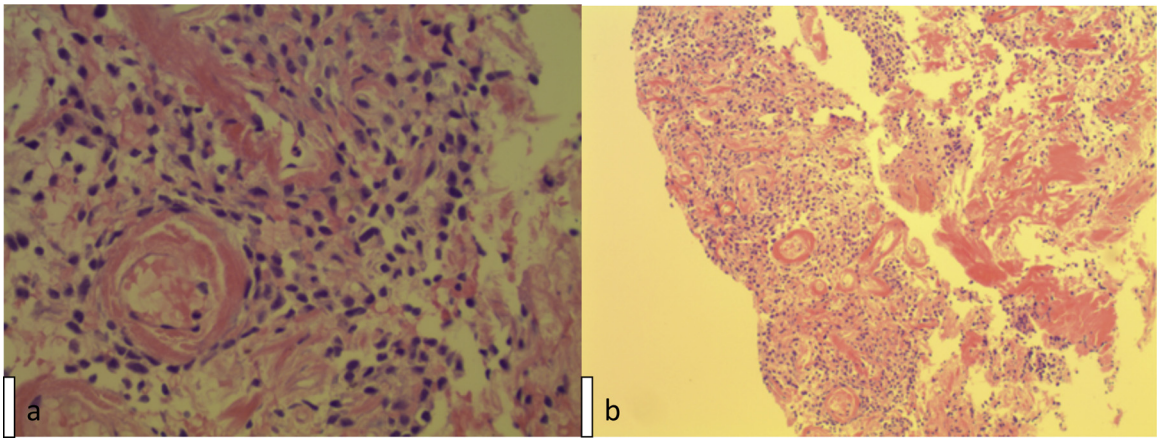

Fig. 2. a) High power view of the tumor. b) Low power view showing low grade spindle cell neoplasm with prominent vascular component and perivascular hyalinization.

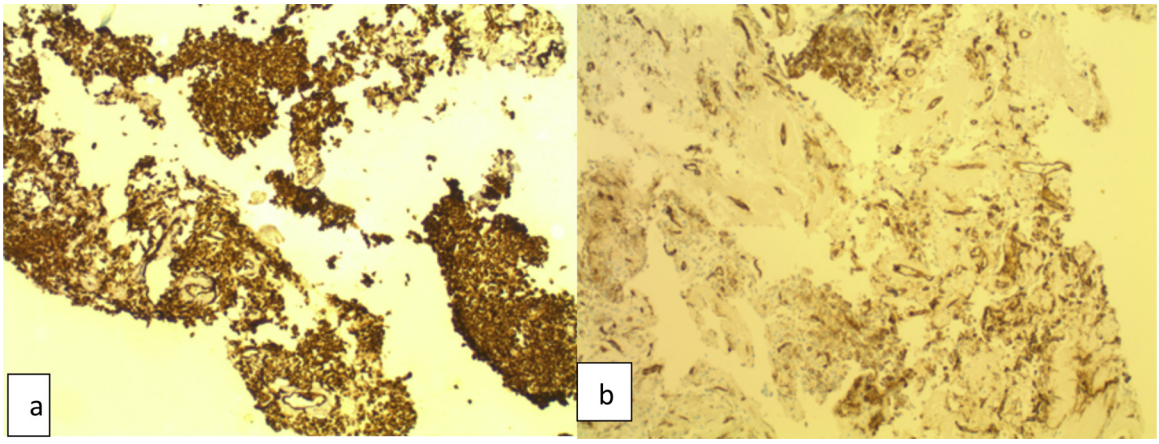

Fig. 3. Immunohistochemistry, (a) Vimentin, positive, (b) CD 34, strong positivity.

was also negative. The final pathologic diagnosis was Pleomorphic Hyalinizing Angiectatic Tumor (PHAT).

3. Discussion

Most described cases of pleomorphic hyalinizing angietatic tumors have been noted to involve the lower limbs while breast, upper limbs, buttocks, and trunk are less frequently affected [4,5]. In the above patient, the lesion was located in the groin which is an uncommon location. Demographically, PHAT tumors are more prevalent in older women – as it is the case for this patient.

Histologically, Pleomorphic hyalinizing angiectatic tumors possess dilated, thin-walled vessels lined with a layer of hyaline substance composed mostly of fibrin. These are immersed in a

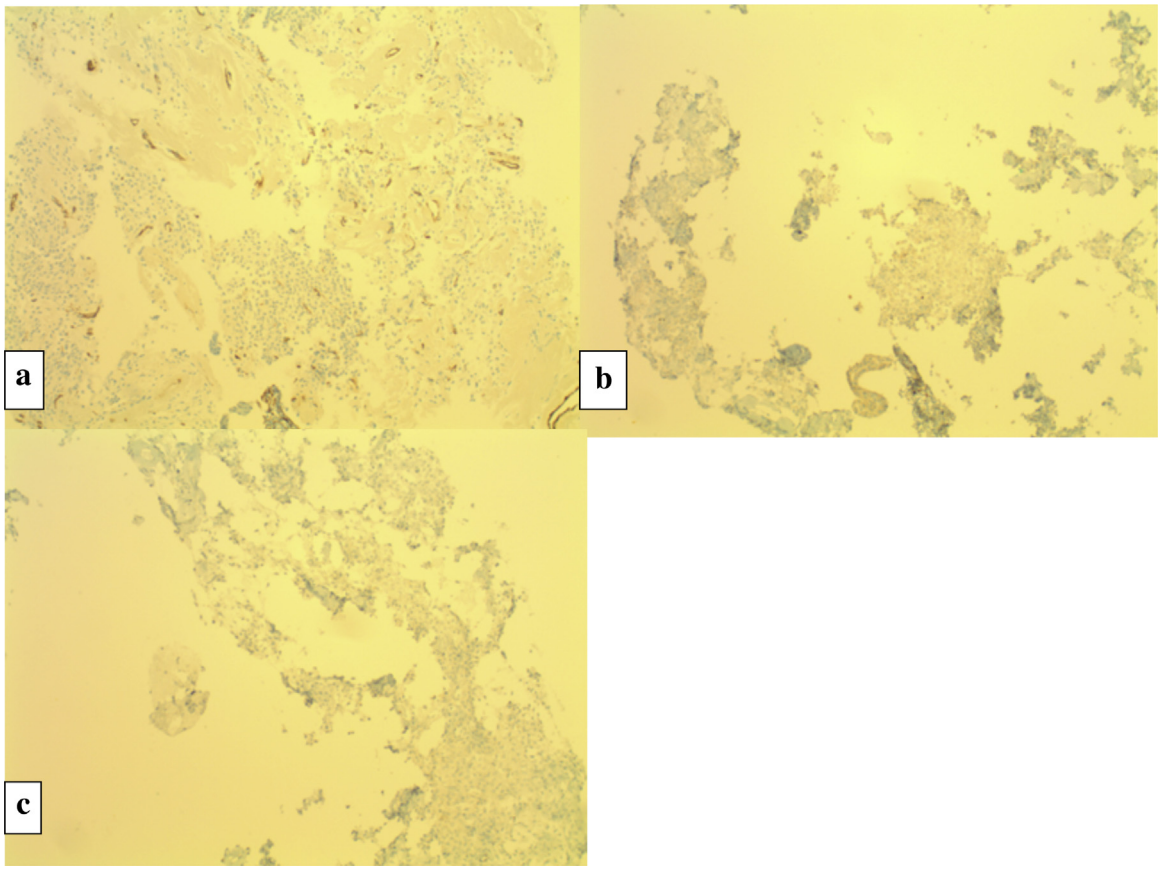

Fig. 4. Immunohistochemistry, (a) SMA, Negative (b) CD117, Negative (c) S100, Negative.

network of proliferating spindle and inflammatory cells [1,2,4–9]. These cells possess hyperchromatic, pleomorphic nuclei and internuclear cytoplasmic inclusions [5,6,9]. In spite of the degree of atypia noted on histology, there is usually few mitotic activity [6,7,9].

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 neurolimma, low grade malignant fibrous histiocytoma, ancient schwannoma, cellular angiofibroma, solitary fibrous tumors, and undifferentiated pleomorphic sarcoma [4]. 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 [5,7,9]. 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 [5,7,9].

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% [3,4]. Although majority of the cases recur as PHAT, rare recurrences with sarcomatous components have been reported. [3,5,6,8] No metastases however have been recorded till date [1,3–5]. Due to the biology of these tumors, some experts recommend that they be viewed as locally aggressive, low grade tumors. However, according to the WHO classification of soft tissue tumors, pleomorphic hyalinizing angietactic tumors are benign tumors of uncertain differentiation [4,9].

4. Conclusion

We report a case of a rare tumor – pleomorphic hyalinizing angiectatic tumor (PHAT) – which presented as a groin mass, and we reviewed the clinicopathologic features of this tumor on the platform of our reported case. Considering the rarity of this tumor, a detailed review of all diagnosed cases will help to define the features of this tumor in order to facilitate accurate diagnosis and prompt management.

Conflict of interest

None.

Consent

Consent was obtained from the patient prior to writing the manuscript.

Author contribution

Chibueze Onyemkpa: Study concept and writing the paper.
Tolutope Oyasiji: Study concept, review and revision of the paper.

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