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Case report

A pelvic pleomorphic hyalinizing angiectatic tumor: a rare case report

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Abstract

Pleomorphic hyalinizing angiectatic tumor (PHAT) is a particular rare entity corresponding to soft tissue neoplasm with low malignant potential. The exact etiopathology responsible for the tumor occurrence is not exactly identified. Knowledge of this condition is necessary to avoid misdiagnosis of the lesion and subsequently, to adopt the proper management. Here we report a case of PHAT originating from soft tissue in the pelvic retroperitoneum. According to the litterature, few articles deal with such pathology and such location. This work illustrates features of the tumor, its characteristics, and the adequate management with the histopathological findings.

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Introduction

Pleomorphic hyalinizing angiectatic tumors (PHATs) were first described in 1996 by Smith *et al.* [1], and are rare mesenchymal tumors of uncertain origin and low potiential of malignancy [2,3]. PHATs are characterized by the presence of amorphus materiel including various clusters of thin-walled characteristic angiectasic vessels that are infiltrated and surrounded by spindle-shaped, round and plump pleomorphic cells. These cells are frequently arranged in sheets or, less commonly in fascicles. The pleomorphic cellular component is constituted of large single and multinucleate cells with enlarged irregular nuclei and abundant cytoplasm. The mitotic activity is rarely found [1,2].

Patient and observation

A 40-year-old woman was referred to the surgery department for evaluation of a pelvic mass. The lesion was incidentally discovered by her gynecologist on ultrasonographic imaging required for 4-month-history of pelvic reccurent pain. Her past medical and surgical history was unremarkable. The physical examination found slight pelvic palpatory tenderness. Laboratory tests were within normal limits. Radiological investigations including ultrasonography (US) and magnetic resonance imaging (MRI) of the pelvic cavity revealed a large irregular and lobulating heterogenous soft tissue lesion measuring 10*7*5 cm and located in the pelvic retroperitoneum (Figure 1). The mass had close contact with pelvic right ureter and right iliac vessels. The patient underwent a surgical complete excision of the lesion. Intraoperatively (Figure 2), the process was irregular with multiple vessels on its surface, the mass was adherent to adjacent tissues and vessels. The tumor was carefully dissected and then completely removed with no bleeding during operation. The specimen (Figure 3) was sent for an histopathological examination. Microscopically (Figure 4), numerous dilated thin-walled vessels with an evident hyalinization of the vessel walls were found. Pleomorphic neoplastic cells were distributed among dilated vessels. Cellularity was variable with hyper and hypo cellular areas and sparse mitotic activity was noted. Furthermore on Immunohistochemistry, the tumor shows a strong positivity for CD34, CD99, CD117 and vimentin. Whereas, S-100 protein, CD56, smooth muscle actin (SMA), Desmin, DOG-1, ALK-1 and actin were negative. Basing on these findings, the definitive diagnosis was consistant with pleomorphic hyalinizing angiectatic tumor (PHAT) of the soft parts. The post-operative course was uneventful, the patient was discharged the 4th day post operatively. The follow-up period was of 6 months with no evidence of local recurrence on follow-up imaging.

Discussion

Pleomorphic hyalinizing angiectatic tumor (PHAT) are defined as rare soft tissue tumors categorized under benign neoplasm with uncertain differentiation and uncertain origin [4,5]. The most common site of such tumors is the subcutaneous tissue and muscles, located mainly in low extremities, less commonly these tumors may have a deep location as the pelvis [6-8]. In the present case, the soft tissue in the pelvic retroperitoneum is affected. The clinical presentation is non-specific with no significant manifestations. It could be incidentally discovered when the mass is relatively large with a local slow growing course [6]. According to the literature, there are few published reports related to the imaging features of PHATs. Typically, PHATs are shown as soft tissue lesions without calcifications (8 10). On MRI, PHATs appear hypointense isointense on T1 weighted sequences, heterogeneously isointense hyperintense on T2 weighted sequences, and show significant heterogenous enhancement following intravenous contrast

administration [2]. Microscopally, PHATs have polymorphotic appearance with characterized thin-walled, ecstatic vessels lined with a layer of hyaline substance mostly composed of fibrin. These pleomorphic neoplastic cells are immersed in an amorphus materiel of proliferating spindle and inflammatory cells containing hyperchromatic, pleomorphic nuclei and inter-nuclear cytoplasmic inclusions. Furthermore, few mitotic activity is rarely found [9,10]. Immuno-histochemical stains are useful for better tumor characterization because this kind of neoplasm has several histologic similarities with other soft tissue tumors and could be easily misdiagnosed. Differential diagnosis include; solitary fibrous tumors, fibrous histiocytoma, cellular angiofibroma and schwannoma. Thus, PHATs are characteristically strongly positive for CD34 and vimentin but show negative staining for desmin and S-100 [9]. Complete surgical excision of the tumor with macroscopic healthy soft tissue margins avoid the local recurrence. However, metastases have not been recorded till date.

Conclusion

We report a rare case of PHAT with an unusual deep location, which is accidentally discovered. The diagnosis is established basing on microscopic and immunohistochemical analysis. The prognosis related to such tumors is relatively good except a low risk of local recurrence when the excision is not complete.

Competing interests

The authors declare no competing interests.

Authors' contributions

All the authors have read and agreed to the final manuscript.

Figures

Figure 1: radiological findings on MRI of the pelvis showing a well-defined lobulated soft tissue mass (white arrow), measuring 10x7x5 cm and located in the pelvic retroperitoneum. The signal intensity was inhomogenous. The lesion has significant heterogenous enhancement with fat saturation following intravenous contrast administration

Figure 2: intraoperative photo demonstrating the tumor during dissection. The mass was carefully separated from adjacent tissues and vessels

Figure 3: the specimen corresponding to a white-yellow lobulated mass

Figure 4: numerous ectatic thin-walled vessels with an evident hyalinization of the vessel walls. Pleomorphic neoplastic cells were distributed among dilated vessels and are immersed in an amorphus materiel of proliferating spindle and inflammatory cells. Cellularity was variable with hyper and hypo cellular areas

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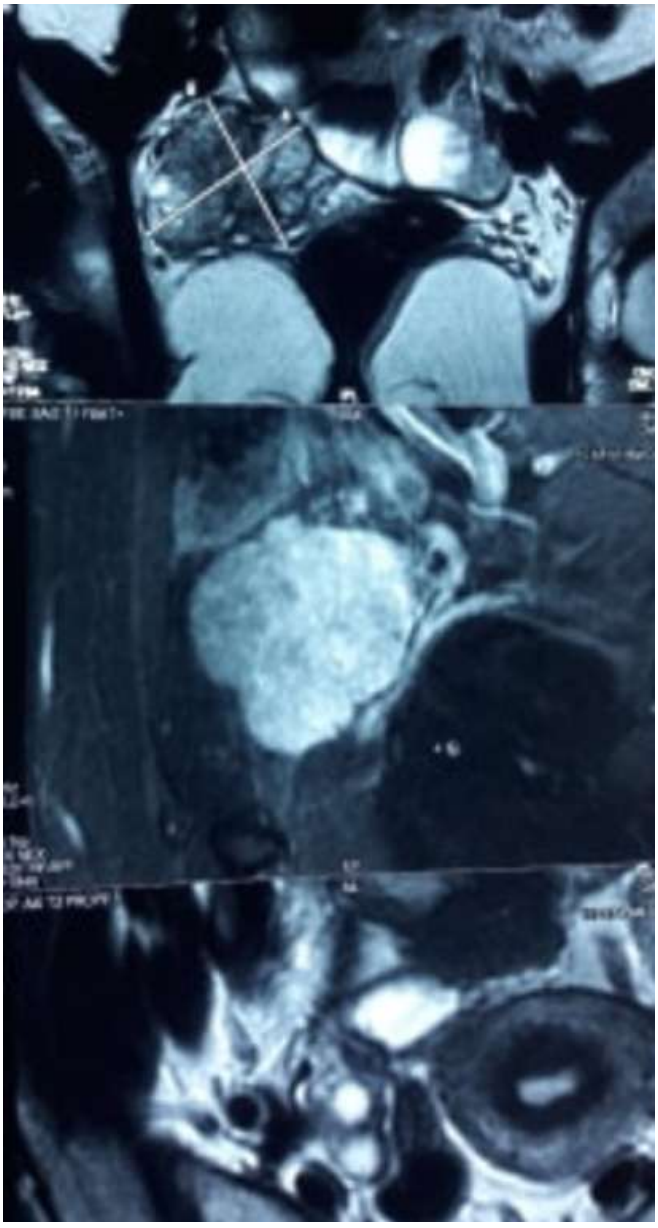

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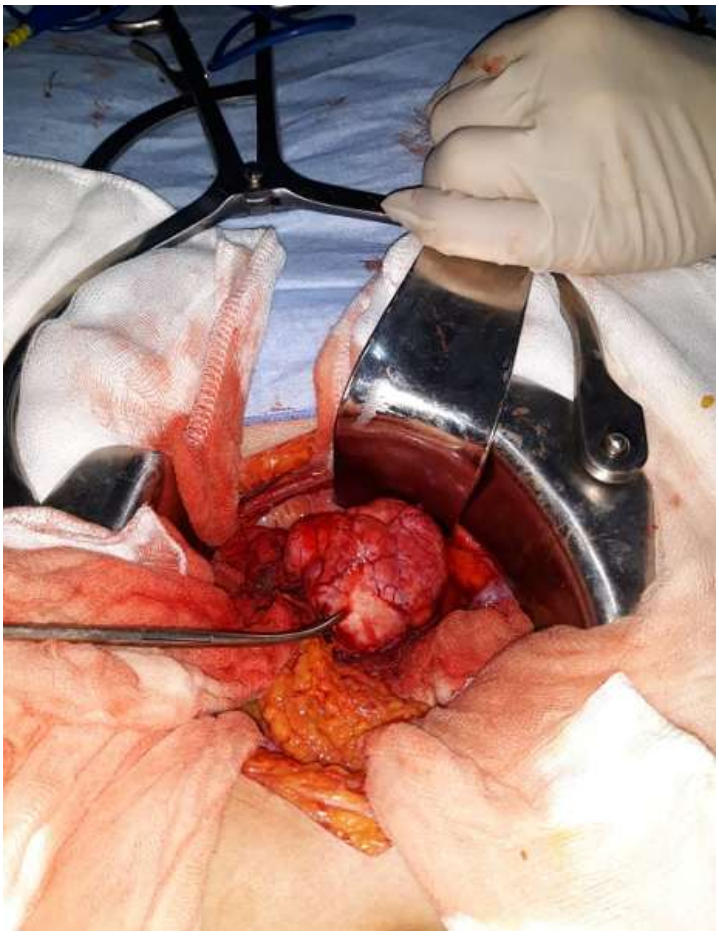

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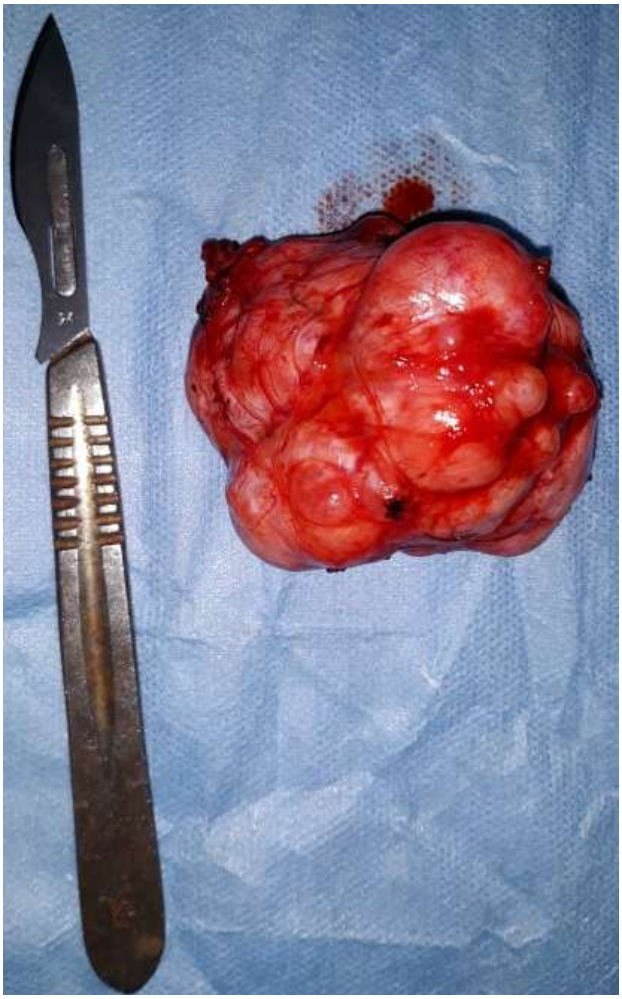

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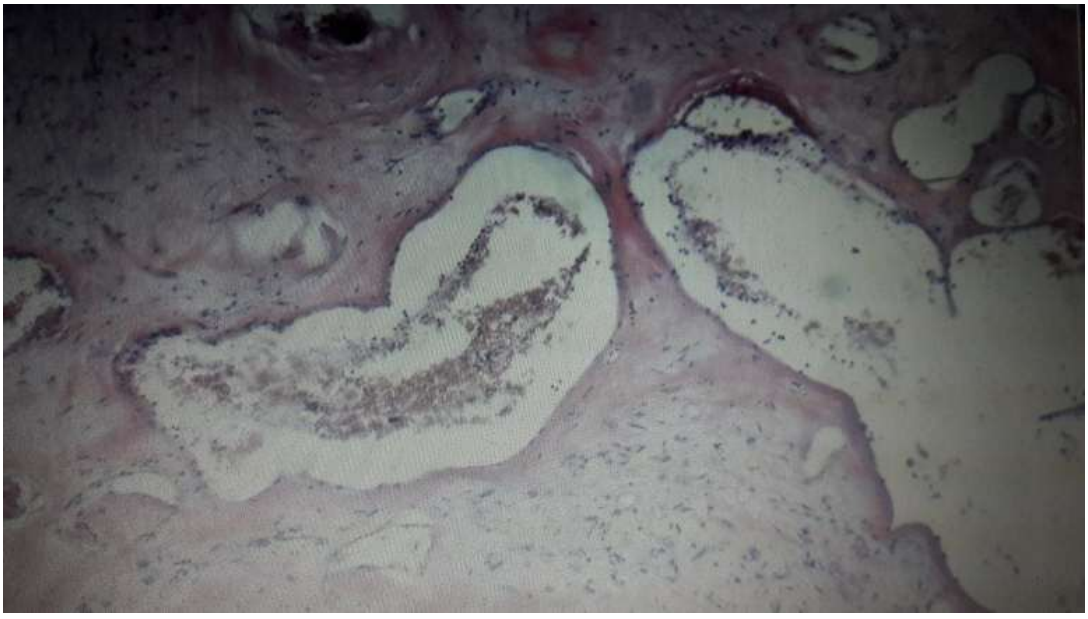

Figure 4: numerous ectatic thin-walled vessels with an evident hyalinization of the vessel walls. Pleomorphic neoplastic cells were distributed among dilated vessels and are immersed in an amorphus materiel of proliferating spindle and inflammatory cells. Cellularity was variable with hyper and hypo cellular areas