Pleomorphic Hyalinizing Angiectatic Tumour of Soft Parts
A Case Report and Review of the Literature

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ABSTRACT

Pleomorphic hyalinizing angiectatic tumour (PHAT) is a recently described, rare, low-grade soft tissue neoplasm. The lesion is characterized by clusters of hyalinized and thrombosed ectatic vessels alternating with a variably cellular stroma composed of atypical cells, many with intranuclear pseudoinclusions. Other features are inflammatory cell infiltration, haemosiderin deposits, focal calcification and minimal to absent mitoses. No metastases have so far been described; however, the local recurrence rate has been found to be high.

To date, approximately 60 such cases of PHAT and its precursor, "early PHAT", have been described in the world literature. We report the first known case of PHAT from this institution which occurred in the left loin of a 77-year old woman. Three years previously, a smaller lesion excised from the same location had been called an ancient schwannoma on histology. This is the most common differential diagnosis offered for this entity even though the two differ in immunohistochemical profile. 'Early PHAT' was also identified on the periphery of the recurrent lesion.

Tumor Pleomórfico Hialinizante Angioectásico de las Partes Blandas
Reporte de un Caso y Revisión de la Literatura

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RESUMEN

El tumor pleomórfico hialinizante angioectásico (TPHA) – entidad rara, de reciente descripción – es un neoplasma del tejido blando, de bajo grado. La lesión se caracteriza por la presencia de racimos de vasos ectásicos trombosados e hialinizados, que alternan con un estroma celular variable compuesto de células atípicas, muchas de ellas con pseudoinclusiones intranucleares. Otras características son: la infiltración celular inflamatoria, los depósitos hemosiderínicos, la calcificación focal, y la mitosis mínima o ausente. Hasta el presente no se han descrito metástasis. Sin embargo, se ha hallado que la tasa de recurrencia local es alta. Hasta la fecha, aproximadamente 60 de estos casos de TPHA y su precursor el "TPHA temprano", han sido descritos en la literatura mundial. Reportamos el primer caso de TPHA conocido de esta institución – una anciana de 77 años de edad, a quién se le presentó en la región lumbar izquierda. Tres años antes, una lesión más pequeña extirpada del mismo lugar, hubiera sido llamada un schwannoma antiguo en histología. Este es el diagnóstico diferencial más común ofrecido para esta entidad, aun cuando los dos difieren en cuando a perfil imunohistoquímico. El TPHA temprano fue identificado también en la periferia de la lesión recurrente.

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INTRODUCTION

Pleomorphic hyalinizing angiectatic tumour (PHAT) is a rare, recently recognized soft tissue neoplasm of uncertain histogenesis. It was first described by Smith *et al* in 1996 (1) in a series of fourteen cases. The largest published study of this entity to date was done by Folpe *et al* (2) in 2004 in a review

of forty-one cases of PHAT and "early PHAT", a partially myxoid lesion that now appears to represent a precursor lesion. Variable recurrence rates have been reported but not metastases.

We present the case of a 77-year old woman who three years previously had a 12 cm subcutaneous lesion excised from the left loin. The recurrence three years later measured 17 cm in widest diameter.

Case Report

A 77-year old woman presented in 2001 with a two-year history of a left loin mass which was slowly increasing in size. Examination of the mass revealed an 8×6 cm soft to firm subcutaneous mass and excision biopsy was recommended. The patient however defaulted and re-presented eight months later at which time the mass had increased in size to 12 cm. Excision biopsy was done and the pathological diagnosis offered was that of an ancient schwannoma. Resection margins were not commented on at that time. Three years later the mass recurred in the same location. Wide local excision was performed.

Grossly, the lesion had a maximum dimension of 17 cm. It was described as an irregular, firm, tan mass which on cut sections revealed a variegated tan, lobulated surface with multiple foci of haemorrhage and necrosis.

Microscopically, a fairly well circumscribed but unencapsulated lesion was identified composed of alternating vascular and cellular areas. The latter consisted of sheets and fascicles of round to spindled, pleomorphic cells arranged for the most part in a compact fashion. In the less cellular areas, the stroma displayed variable myxoid change. The constituent cells had eosinophilic cytoplasm with hyperchromatic nuclei, some of which contained prominent pseudoinclusions (Fig. 1). Mitoses were rare, numbering less than two

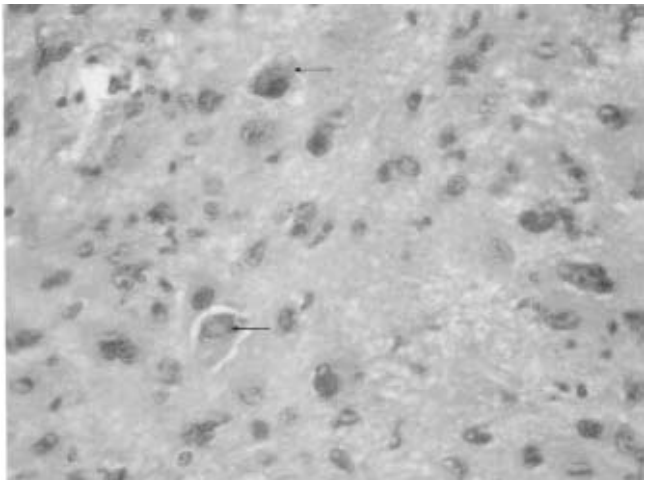

Fig. 1: High power view showing pleomorphic cells containing prominent intranuclear psuedoinclusions (arrows) (H&E).

per 50 high power fields. Foci of necrosis were scattered throughout and in these areas the cells were arranged in a more prominent storiform pattern and contained large eosinophilic nucleoli.

The vascular areas were composed of clusters of ectatic blood vessels of variable size, many of which were surrounded by hyalinized material which in places emanated from between the surrounding cells. Many of the blood vessels contained fibrin thrombi whilst some displayed papillary endothelial hyperplasia (Fig. 2). Some clusters were completely obliterated. Large numbers of patent blood-filled vessels of varying calibre were scattered throughout the remainder of the lesion. Other features included foci of haemosiderin deposition in close association with the vascular aggregates and a variable mixed inflammatory cell infiltrate. In one area, "early PHAT" as described by Folpe *et al* (2) was identified on the periphery of the main lesion.

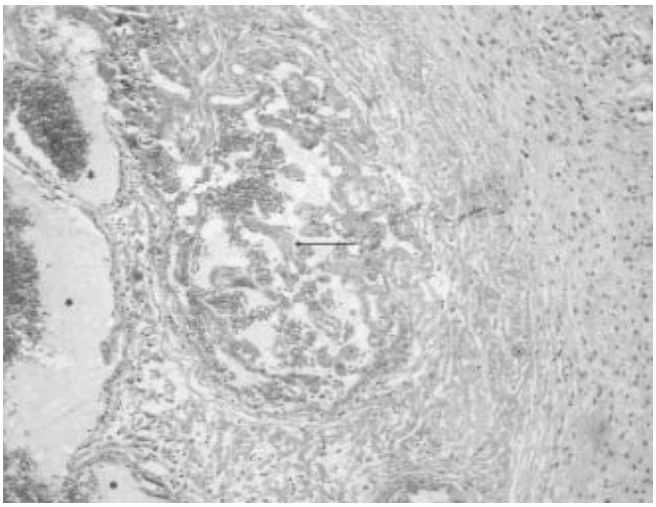

Fig. 2: High power view showing variably-sized, congested, ectatic blood vessels exhibiting papillary endothelial hyperplasia (arrow) (H&E).

This lesion appeared within 0.5 mm of the deep resection margin. Immunohistochemistry showed negative staining of S-100 protein and strong positive staining for vimentin. CD 34 was unavailable.

DISCUSSION

Pleomorphic hyalinizing angiectatic tumour of soft parts (PHAT) is a rare, recently described entity. From the study by Folpe *et al* (2) in 2004, it was observed that PHAT occurred primarily in adults with a median age of 51 years (ages ranging from 10–79 years). It occurs in both genders with a slight predominance in women. The maximum size of the lesion quoted in that study was 19.7 cm and all were subcutaneous in location. However, the previous reports showed that up to 16% of cases can occur intramuscularly (3). Folpe *et al* also described, for the first time, a distinctive myxoid precursor lesion of PHAT, which usually occurs concomitantly with the mature entity (2). That study also showed that when "early PHAT" recurs it does so as classic PHAT.

PHAT is characterized histologically by a unique combination of hyalinized clusters of ectatic vessels alternating with a pleomorphic cellular stroma including large cells with prominent intranuclear pseudoinclusions. Other features include inflammatory cell infiltration, primarily mast cells, haemosiderin deposits, focal calcifications and minimal to absent mitotic activity. Groisman *et al* (6) in 2000 described the presence of numerous, proliferative, non-hyalinized vessels, some of them showing permeation of their walls by minute capillaries.

Pleomorphic hyalinizing angiectatic tumour is considered today to be a mesenchymal tumour of intermediate malignancy, with a high rate of local recurrence. Folpe *et al* (2) described one case in which the recurrent lesion did not contain typical PHAT but instead resembled a high grade myxoid sarcoma. The original lesion was also noted to be the only one of their series which contained a small focus of necrosis.

The index case is the first known case of PHAT to be reported at this institution. The recurrence exhibited large areas of haemorrhage, cystic degeneration and necrosis. "Early PHAT" was also identified on the periphery of this lesion. However, the remainder exhibits all the classic features of PHAT described so far in the literature and immunohistochemistry was supportive of the diagnosis. There was no evidence of malignancy in the recurrence and the resection margins appeared free of tumour. Close follow-up of this patient is recommended.

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