

Pleomorphic Hyalinizing Angiectatic Tumor of the Scrotum

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INTRODUCTION

The pleomorphic hyalinizing angiectatic tumor (PHAT), first described by Smith and colleagues,⁽¹⁾ is a rare mesenchymal tumor with intermediate malignant potential. This unique tumor occurs principally in the superficial soft tissues of the distal extremities, and features ectatic, fibrin-containing vessels with prominent circumferential hyalinization, spindled and pleomorphic stromal cells with intranuclear inclusions, and a variable inflammatory component.⁽²⁾ Although the number of reported cases is limited and there is disagreement regarding the line of cellular differentiation, the pathologic features of PHAT have been well-delineated.⁽³⁾ The purpose of this report is to present the case of PHAT encountered within the scrotal sac and add this rare tumor to the differential diagnosis of the scrotal swelling.

CASE REPORT

A 68-year-old man was referred to our department for evaluation of a right scrotal mass that had been enlarging slowly for several years causing mild discomfort. Bimanual examination of the scrotum confirmed the presence of a painless nontender mass of the right hemiscrotum inseparable from the right testicle. The left hemiscrotum

and testicle were unremarkable and with no evidence of disease.

Scrotal ultrasonography revealed a homogeneous echo-poor mass of the right hemiscrotum involving the testicle, measuring approximately 180 mm × 150 mm and consistent with the ultrasonographic appearance of a testicular tumor. Computed tomography of the pelvis exhibited an inhomogeneous mass with calcifications, suggesting an enhancing soft tissue component, measuring approximately 160 mm × 150 mm and not involving the right testicle (Figure 1). There was no evidence of metastatic disease or lymphadenopathy on imagings. Tumor markers, including alpha-fetoprotein, lactate dehydrogenase, and beta-human chorionic gonadotropin assays, were all within normal limits.

The clinic, ultrasonographic, and radiographic appearances were suspected to be malignant, but were not proved conclusively. Therefore, a surgical exploration was performed. Peri-operative distinguishment between the scrotal mass and the testicle was impossible. Subsequently, a right inguinal radical orchiectomy with high ligation of the spermatic cord was performed.

On pathological examination, the

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Figure 1. Computed tomography of an inhomogeneous mass with calcifications, suggesting an enhancing soft tissue component, measuring approximately 160 mm × 150 mm and not involving the right testicle.

excised mass lacked sufficient surrounding tissue to thoroughly examine the presence of positive or negative surgical margins. Gross findings revealed a well-circumscribed 160 mm × 150 mm × 120 mm nonencapsulated mass, with cystic components and diffusely infiltrative borders, and maroon in color.

Microscopic examination exhibited a moderately cellular lesion with a lobular pattern demarcated toward the periphery by a thin pseudocapsule featuring infiltrative margins and was characterized by the presence of alternating vascular and cellular areas. The most prominent feature was the various sized thin-walled ectatic hyalinized blood vessels (Figure 2). The tumor

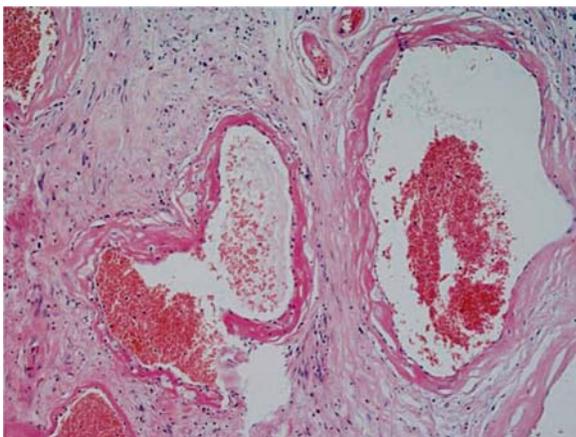


Figure 2. Thin-walled ectatic blood vessels with red blood cells in the vascular lumen (Hematoxylin and Eosin, original magnification ×100).

cells consisted of a mixture of spindled and rounded pleomorphic cells that were arranged in sheets without obvious differentiation, had a distinct cytoplasmic border, and were eosinophilic, with some intranuclear inclusions or cytoplasmic hemosiderin deposits.

Immunohistochemically, the neoplastic cells were strongly positive for vimentin and CD34 and negative for S-100 protein, CD31, desmin, smooth muscle actin, and cytokeratin.

The right testicle was unremarkable and with no evidence of disease. The patient has been currently followed up for 1 year and there is no evidence of clinic or radiographic recurrences so far.

DISCUSSION

The differential diagnosis of scrotal swelling includes tumor, epididymitis, epididymo-orchitis, epididymal cyst, testicular torsion, and less commonly, hernia, hydrocele, spermatocele, varicocele, hematoma, and hamatocele. To the best of our knowledge, this report presents a rare tumor entity that has never been before documented as a primary scrotal lesion.

Pleomorphic hyalinizing angiectatic tumor is a rare soft tissue neoplasm that occurs mainly in the 4th to 7th decades, with a slight female preponderance.⁽²⁾ Approximately, 80 cases have been reported to date.⁽⁴⁾ In most of the cases, patients had the mass for more than 1 year before operation.⁽⁵⁾ The main location of occurrence is the lower limb; however, other anatomic sites have been reported, such as the trunk, upper extremities, inguinal triangle, buttock, oral cavity, and mesorectal soft tissue.^(6,7) Although most of the patients are cured with local excision and no metastases have been reported so far, a high rate of local recurrences (33% to 50%), sometimes necessitating amputation for local control, has been observed.⁽¹⁾

Grossly, PHATs are well-circumscribed, but unencapsulated lesions with a lobulated growth pattern and measure up to 20 cm in diameter.⁽⁴⁾ Histologically, they are of low to moderate cellularity, with a combination of sheets of spindled and pleomorphic cells associated with

an ectatic, partially hyalinized vasculature. The variously sized dilated blood vessels are scattered and clustered, and their walls are lined by amorphous eosinophilic material.

Immunohistochemically, the tumor cells show positive staining for vimentin in almost all cases, with variable CD34 and CD99 positivity.⁽⁴⁾ Other antigens, such as S-100 protein, actin, desmin, cytokeratin, and CD31, are generally negative.⁽³⁾

The differential diagnosis of PHAT includes malignant fibrous histiocytoma, schwannoma, solitary fibrous tumor, myofibroblastoma, spindle cell metaplastic carcinoma, and Kaposi sarcoma.⁽⁸⁾

With the increasing use of ultrasonography and computed tomography, other examples of scrotal PHAT are expected to be discovered. Awareness of the possibility that PHAT may occur in the scrotum should be kept in the differential diagnosis of scrotal swelling. The recognition of this rare tumor entity could avoid unnecessary radical surgery.

CONFLICT OF INTEREST

None declared.

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