SCROTAL LYMPHANGIOMA IN ADULTS: “PROS” AND “CONS” MORPHOLOGICAL DIAGNOSIS

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ABSTRACT

According to WHO Classification of Tumors of Soft Tissue and Bone, lymphangioma is defined as a benign, cavernous/cystic vascular lesion composed of dilated lymphatic channels, that characteristically occurs in newborns or early infancy without gender or race predilection. The scrotum is one of the least affected sites. We present a case of 43-year-old man with tumor mass, gross and histologically similar to the scrotal lymphangioma, in combination with acute purulent orchitis and venous occlusion. The authors describe histopathological findings and discuss relevant issues in terminology.

Key words: lymphangioma of the scrotum, lymphatic malformation, histological diagnosis.

INTRODUCTION

The human lymphatic system is only rarely affected by neoplastic growth, producing tumor masses. According to WHO Classification of Tumors of Soft Tissue and Bone, lymphangioma is defined as a benign, cavernous/cystic vascular lesion composed of dilated lymphatic channels, that characteristically occurs in newborns or early infancy without gender or race predilection (4). Lymphangiomas are most commonly found in the head and neck area and with decreasing frequency axilla and groin, trunk and extremities, visceral locations (abdomen and thorax) (3). The scrotum is one of the least affected sites with main clinical features including painless, indolent scrotal masses; sometimes may be associated with acute scrotum due to hemorrhage or infection (1–2, 6–8). There is a wide spectrum of differential diagnoses: hydrocele, hematocoele, spermatocele, varicocele, inguinal hernia and dilatation of the rete testis, to name a few. High resolution ultrasonography is the first imaging assay to resolve the diagnostic problem. Complete surgical excision of the lymphangioma is curative, preventing further recurrences. Surgical pathology examination of the specimen provides correct diagnosis. In the older age group, lymphangiomas are thought to arise secondary to concurrent infections or trauma.

CASE PRESENTATION

43-year-old man with no significant past medical history was admitted in our hospital for diagnostic evaluation of incremental painful swelling of the left scrotal sac, accompanied by mild fever (38.2°C) lasting for several days. On palpation the left testicle was tender with firm spermatic cord. Laboratory studies revealed mild leucocytosis (12.9 G/l), increased sedimentation rate and CRP. Ultrasonography showed multilocular cystic mass of the testicular coverings and orchiectomy was performed. The specimen was submitted for histologic evaluation. Grossly, the whole sample measured 5.5x6 cm with 5 cm spermatic cord attached. The sample consisted of atrophic testis measuring 4x2.5 cm, adhered firm, whitish coverings and comma-shaped tumor mass with small cystic spaces filled with clear fluid. The tumor occupied half of the testicular circumference and was inserted between fibrotic tunica albuginea and tunica vaginalis (Fig. 1A). Histologically, the testicular parenchyma was destroyed by heavy neutrophil infiltrate, consistent with acute purulent orchitis. Sections from the spermatic cord were found to contain small to medium sized veins that
were completely obliterated by organized thrombi accompanied by acute inflammatory cells (Fig. 1B). The multilocular cystic lesion was composed of fibrous septa with thickness variation lined with inconspicuous endothelial cells with no atypia. The cystic spaces contained pale pink material and scattered neutrophil leucocytes (Fig. 1C, D). Despite the gross and histological similar appearance to the scrotal lymphangioma, patient’s age and evidence for vascular obstruction precluded the diagnosis. In the final pathological report the lesion was interpreted as tumor-like remodeling of the testicular lymphatic channels.

DISCUSSION
In the current definition of lymphangioma, WHO experts have agreed on the word “lesion” instead of neoplasm probably due to lack of consistent data to provide evidence for tumor nature (4). The pathogenesis of lymphangioma was first proposed by Whimster in 1976, who assumed the pathological basis is in the failure of primitive lymphatic cisterns to communicate effectively with the rest of the lymphatic system with consequent dilatation of the lymphatic channels (9). This theory connects the lesion with embryonic stage of lymphatic system development and should be considered as congenital malformation. There is alternative hypothesis reflecting the origin of lymphangioma supported by Ozeki et al. observations that certain blockers of endothelial growth could trigger regression in thoracic lymphangiomatosis (5). This implies a possible tumor nature engaging some degree of endothelial hyperplasia. At this time both terms lymphangioma/lymphatic malformation can be used interchangeably.

The histopathological diagnosis is usually straightforward and hardly ever requires further investigation by immunohistochemistry (4). Based on the relative diameter of the vessels lymphangiomas are subclassified as capillary, cavernous and cystic, although a combination of the three patterns is not unusual with in a single lesion. The endothelial lining is often flattened or absent, frequently surrounded by lymphocytic aggregates. Endothelial cells consistently express D2-40 and CD31, variably CD34. The main differential diagnosis is adenomatoid tumor, but both tumors show same immunohistochemical profile and share excellent prognosis due to absence of documented malignant transformation. Outside the early infancy patient’s group, pathological cause for stop of the lymph drainage can always be found and should be carefully looked for. In our opinion, in older patients the usage of both terms - lymphangioma/lymphatic malformation – should be avoided and replaced by lymphatic remodeling in the setting of evident cause for obstruction.
Fig. 1. A: Gross specimen. B (H&E): Spermatic cord. C, D (H&E): Multilocular cystic formation.

REFERENCES: