ABSTRACT
Cystic lymphangioma is a common benign tumor caused by lymphatic malformation. The scrotum is a very rare site for this tumor and only few cases have been reported in the literature. We herewith present a rare case of cystic lymphangioma of the scrotum in an adolescent who presented with an incidental scrotal swelling with no other abnormality, where the diagnosis was suspected on scrotal ultrasonography.

INTRODUCTION
Lymphangioma is a congenital lymphatic tumor usually recognized at birth or in early childhood with the commonest location being head, neck and axilla.[1] Lymphangioma occurring in the scrotum is very rare.[1][2] It may present at birth, in childhood or in adolescence, as seen in our case.[3] Symptomatology is non-specific with resultant diagnostic difficulties. Inaccurate diagnosis with improper management is associated with high rates of recurrence.[4]

CASE REPORT
An 18-year-old male with incidentally noted, painless, soft swelling of 2-3 months duration located on the right side of scrotum with no other complaints came for ultrasonography. History of trauma was not present. Clinical examination revealed a nontender, extratesticular, soft swelling with slight compressibility. Laboratory tests were unremarkable. High resolution ultrasonography of the scrotum revealed a well defined, compressible, cystic mass with multiple septae and locules located inferolaterally in the right scrotal sac, separate from the normal appearing testis and epididymis [Figure 1]. [Figure 2]. There was appreciable posterior acoustic enhancement. The lesion measured approximately 37.2 × 20.3 × 16.5 mm (6.5 mL in volume). The septae were thin and complete and revealed low velocity and high resistance flow on color doppler imaging [Figure 3]. Presence of low level internal echoes with few mobile echogenic foci was also noted. However, no evidence of any calcification was noted. Minimal free fluid was also noted in the right scrotal sac. Ultrasound of the left scrotal sac and the abdomen was unremarkable. No evidence of any extrascrotal extension into the abdomen was noted even on computed tomography of the lower abdomen.

Fig. 1 - Longitudinal US image of lymphangioma scroti showing a multiseptate, cystic mass with posterior acoustic enhancement.
Based on the clinico-radiological findings, a provisional diagnosis of cystic lymphangioma of scrotum was suggested.

Surgery revealed a cystic mass with multiple septae located in the right scrotal sac, densely adherent to the tunica but separate from the right testis and epididymis. Complete excision of the mass including the adjacent skin was performed with uneventful postoperative period. Histopathology of the excised tissue revealed multiple lymphatic channels lined by endothelium while cytology of the contained fluid confirmed lymph like fluid. No evidence of inflammatory infiltrate or cellular atypia was seen. Six months follow-up period was unremarkable.

DISCUSSION

Lymphangioma occurs as a result of the failure of lymph to drain from sequestered lymphatic vessels with consequent dilatation of the ducts and formation of a cystic, multisepate mass. Majority of the lesions are congenital but may also occur secondary to infection, inflammation or degeneration. Out of the three forms, capillary, cavernous and cystic varieties, the cystic type is the commonest.[2]

Trauma or aspiration of lymphangioma can result in internal hemorrhage, making it difficult to differentiate it from hemangioma on microscopy. However, presence of benign lymphoid aggregates is highly specific for lymphangioma.[5]

Clinical diagnosis of lymphangioma is easier if it is present in usual locations but imaging is crucial in diagnosis at uncommon and rare locations, especially in the scrotum. All the cases reported in the study by Hurwitz et al., were misdiagnosed pre-operatively, further emphasizing the role of imaging.

These lesions usually present as painless, indolent scrotal masses but may sometimes be associated with acute scrotum due to enlargement or pain following infection or hemorrhage.[6],[7] These tumors are frequently infiltrative and may extend in to the abdominal wall, perineum or retroperitoneum.[1],[4],[6] It may be associated with cryptorchidism.[2]

High resolution ultrasonography is highly accurate in delineating the type and extent of the lesions in most cases. Color doppler US gives an idea of the pattern and extent of vascular supply. Computed tomography and magnetic resonance imaging are used only as problem solving tools especially to determine the extension in to the pelvis and retroperitoneum.

High resolution ultrasonography of cystic lymphangioma reveals cystic mass with multiple septations and locules filled with low level internal echoes that may be mobile. The echoes may be due to internal hemorrhage or debris. Septations seen in lymphangioma often shows low velocity and high resistance vascular flow, characteristic of benign lesion.[8]

Complete excision of the lymphangioma including the scrotal skin is the treatment of choice to prevent recurrences. Injection of sclerosing agents, fulguration or local cryotherapy for cystic lymphangioma are associated with high recurrence rates.[1],[3]

The important differential diagnoses of the multicystic, extratesticular scrotal mass include loculated hydrocele/pyocele, hematocoele, varicocele, hernia, spermatocele, cystic lymphangioma of epididymis and epididymal cyst. Presence of septal flow differentiates cystic lymphangioma from hydrocele, pyocele and hematocoele. Absence of color flow in the cystic spaces differentiates lymphangioma from varicocele. Hernia will have an intra-abdominal extension.

To summarize, although scrotum is a very rare site for cystic lymphangioma, it should be considered in the list of differential diagnosis of multiloculated, benign appearing extratesticular lesions with limited vascular supply in a young male. Ultrasonography is decisive in most cases, accurately identifying and determining the extent of these lesions. Correct diagnosis not only avoids mismanagement but also reduces recurrences by institution of the specific therapy early in the course of disease.
REFERENCES


