Case Report

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Lymphangioma of Male Exogenital Organs

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Lymphangioma manifesting in the genitourinary tract is an uncommon disease. Cystic lymphangioma is a congenital lymphatic hamartoma known as lymphatic malformation. Herein we report 2 cases of lymphangioma of male exogenital organs. After complete excision of the tumor and subsequent follow-up for 6 months, both of them were free of recurrence. Ultrasonography and computed tomography scans are very useful in the differential diagnosis of this cystic disease.

Key Words
genitourinary tract;
lymphangioma;
male

Lymphangioma is a congenital lymphatic hamartoma, which is an uncommon disease entity in the genitourinary tract. There are case reports of scrotal cystic lymphangioma in the English literature, but case reports related to perineal cystic lymphagiomas are rare.[1-6] Between January 1990 and December 2000, a total of 144 cases were diagnosed as lymphangioma at our hospital. Only 3 (2.1%) of them came from male genital organs. Herein, we report clinical presentations, image studies, prognosis and management among 2 of them.

CASE REPORTS

Case 1

A 23-year-old man had suffered from a painless mass over the perineum for 4 months. Two months before admission to our hospital, he noted painful swelling of the mass. Physical examination revealed a 4 × 6 cm mass in size over the perineal region extending to the lower margin of the scrotum. Ultrasonography showed a complex, septated cyst with internal echogenicity (Fig. 1). Lymphangioma was diagnosed and he received excision of the cystic mass. A multilobulated cystic mass over the perineal region with its major part between anus and scrotum, and the origin of the tumor stalk originating near the anus was found. The mass did not communicate with the scrotal content. All of the lymphatic vessels were transected and ligated. The pathologic finding was a thin-walled and dilated lymphatic space with the lumens filled with eosinophilic proteinaceous fluid (Fig. 2), compatible with lymphangioma. The subsequent course of the disease was uneventful and the patient was discharged. There was no evidence of recurrence 6

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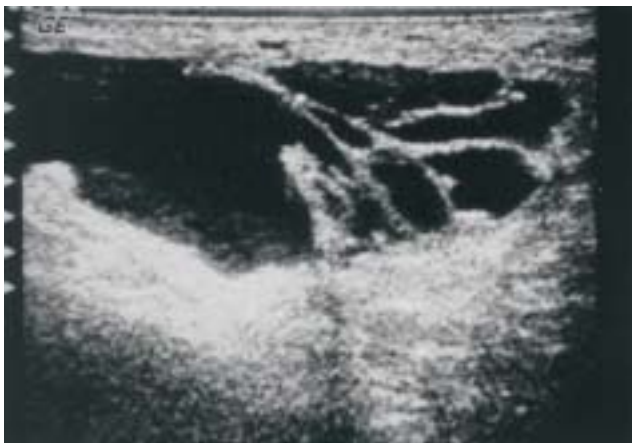

Fig. 1. Ultrasonography revealed a complex septated mass with internal echogenicity.

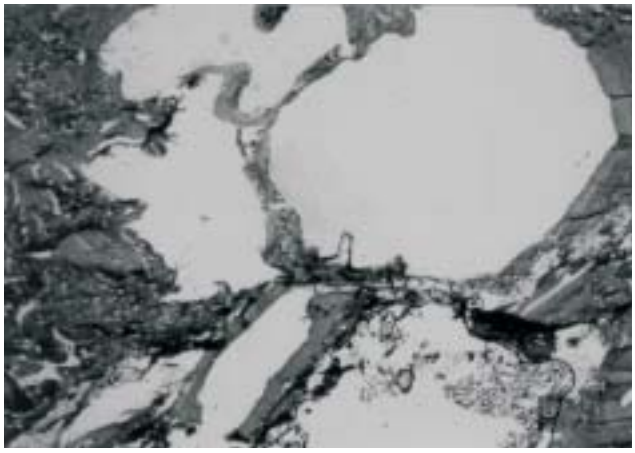

Fig. 2. The pathologic finding was a thin-walled and dilated lymphatic space with lumens filled with eosinophilic proteinaceous fluid.

months after surgery.

Case 2

A 57-year-old man presented with a painless, swelling mass over his right scrotum that had been there for 2 months. Physical examination revealed a hard, non-tender mass 2 × 3 cm in size over the right scrotum that was transparent to light illumination. Beta-human chorionic gonadotropin (β-HCG) of serum was less than 5 MIU/mL, and α-fetoprotein (AFP) of serum was 4.92 ng/mL. The computed tomography (CT) scan of the pelvis revealed no retroperitoneal lymphadenopathy. Under the tentative diagnosis of a right testicular tumor, he received radical right orchidectomy. A cystic mass under the tunica albuginea with septation and clear fluid was found during the operation. The pathologic finding was large irregular spaces lined by thin endothelial cells and separated by a variable amount of connective tissue. The endothelial cells were positive for Ulex europaeus agglutinin 1 (UEA1) and factor 8 immunostains. Cytokeratin stain was non-contributory. The pathologic diagnosis was compatible with lymphangioma. The subsequent course of the disease was uneventful and the patient was discharged. Six months after the surgery, there was no recurrence.

DISCUSSION

Cystic lymphangioma is a congenital lymphatic hamartoma commonly known as lymphatic malformation, cystic hygroma or hygroma. Fifty percent of the cases are present at birth,[1] and 90% of them are evident before the age of 2 years. It occurs mostly in the neck or axilla, and the remaining 5% are in the mediastinum, mesentery, retroperitoneum, bone, breast, buttocks, and thigh, *etc*.[2-4,6] Scrotum and perineum are the least frequent sites. The most common manifestation is a painless, gradually growing mass.[2] As soon as sudden onset of enlargement and pain are found, hemorrhage from minor trauma, excessive formation of lymph and infection should be considered.[2] Physical examination usually reveals a multilocular mass which is transparent to light illumination. If tenderness or a bluish or purplish appearance is evident, infection or hemorrhage should be suspected.[2,3]

Ultrasonography is very useful in the differential diagnosis of this cystic disease. The typical finding is a multilocular, septated, cystic mass containing echogenic material.[2,5,6] CT scan of the abdomen or pelvis is indicated for patients who have suspicious extensions of the cystic lesion to the retroperitoneum, pelvis or mesentery.[2,4] Complicated lymphocutaneous fistulae and a lymphocele can be detected by Tc-99m antimony sulfate lymphoscintigraphy.[8] Aspirated fluid usually shows thin, watery, clear to straw-colored fluid containing lymphocytes, monocytes and occasional polymorphonuclear cells.[2]

The differential diagnoses include inguinal hernia, hydrocele, hematocele, varicocele, spermatocele, dermoid cyst, infection, trauma and testicular torsion.[2,4] Missed

diagnoses, which are common before the operation as in our case 2, usually lead to improper surgical approaches, poor exposure, incomplete excision of the tumor and resulting recurrence.[2,3]

Extrascrotal or extraperineal extension to adjacent inguinal, abdominal and pelvic regions is possible. Surgical approaches should be based on preoperative image studies about tumor extension.[1-4] Postoperative radiation was reported for control of recurrence and prevention of keloid formation. Some small lymphangiomas were observed to regress spontaneously.[4]

Abara *et al*. presented a preoperative complication of torsion of a stalk of a cavernous lymphangioma.[7] The most common postoperative complication was tumor recurrence, which happened in 25% to 50% of patients within 3 months. Other complications were edema, prolonged lymphatic drainage and local infection. Associated elephantiasis, filariasis and lymphangioma circumscriptum were also documented.

The pathologic features consisted of endothelial lined canaliculi, connective tissue stroma filled with lymphocytes, monocytes and polymorphonuclear cells. Lymphangiomas are classified into 3 subtypes. Capillary lymphangioma contains dilated lymphatic vessels with rich cellular connective tissue stroma, connected with normal lymphatics. Cavernous lymphangioma contains dilated lymphatic sinuses in an actively growing lymphoid stroma, connected with normal lymphatics. Cystic lymphangioma contains multiple cysts of varying size with serous, serosanguineous or chylous fluid, intercommunicated or separated by fibrous septa.[1,4]

In conclusion, cystic lymphangioma is seen infrequently. Only 5% of patients have them in regions other than neck and axilla. Adult scrotal or perineal cystic lymphangioma is rather uncommon, but multicystic septated mass is a typical finding in ultrasonography. Preoperative workups for the extension of the cystic lesion are important in order to have it excised completely. Awareness of this illness, careful investigation of associated adjacent organs, and complete excision of this cystic tumor will achieve a good prognosis.

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