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Perineal and scrotal lymphangioma in a 3-year-old male patient: An unusual case report

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ABSTRACT

Lymphangioma is a rare cause of swelling in the perineal and scrotal regions. This case report describes the clinical, ultrasonographic, and histopathological findings of a 3-year-old pediatric patient with a diagnosis of perineal and scrotal lymphangioma.

Key Words: Lymphangioma; perineum; scrotum; child.

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Introduction

Lymphangioma includes congenital benign lymphatic hamartomas located almost completely in the axillary and mediastinal regions (95%) [1,2]. The perineal and scrotal regions are very rare localizations [2,3]. Perineal lymphangioma, which is an extension of abdominal lymphangioma, is rarely isolated, as reported in a few cases in the literature [3,4]. Scrotal lymphangioma usually involves the scrotal walls, tunics, testes, epididymis, spermatic cord, and Colle fascia [5]. This case report describes the clinical, ultrasonographic, and histopathological findings of a 3-year-old pediatric patient with a diagnosis of perineal and scrotal lymphangioma.

Case report

A 3-year-old male patient was brought to the clinic by his parents, who had noticed a swelling in his perineal region that was present at birth and had gradually increased in size.

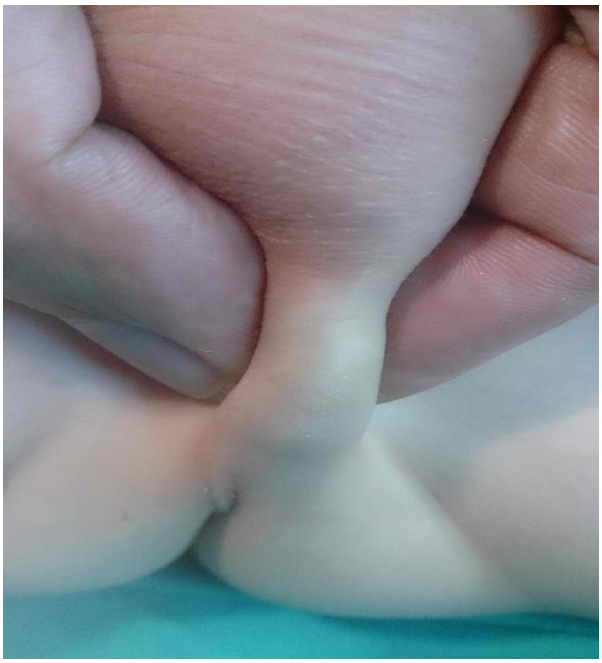

Fig. 1. The appearance of the perineal-scrotal mass in the examination.

His physical examination revealed a non-tender and medium-sized perineal mass 2 × 2 cm in diameter (Fig. 1).

The left testis could be felt separately from the mass. The right scrotum was normal. Ultrasonography revealed a mass with microcystic areas in the perineal area. A diagnosis of lymphangioma was made based on the radiologic and clinical findings. An ultrasound of the abdomen was normal. The patient also had a hemangioma on the right eyelid and on the left elbow (Fig. 2. A, B).

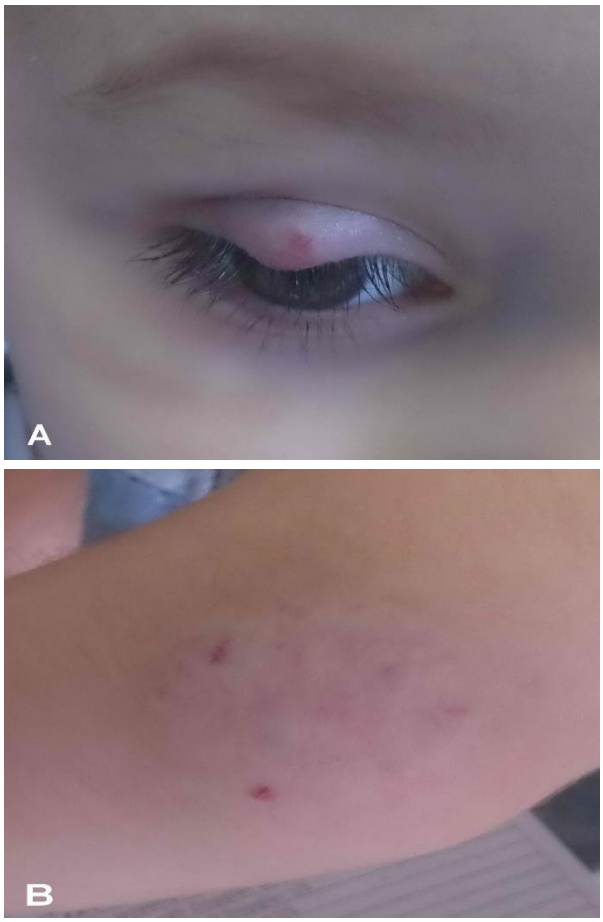

Fig. 2. A, B. The hemangioma on the right eyelid and left elbow.

The patient underwent exploratory surgery after routine examinations. A perineal and scrotal incision revealed a 3 × 3 cm multilocular cystic mass that was adherent to the scrotal wall (Fig. 3). The left testis and the spermatic cord were healthy. The mass, along with the overlying skin, was completely excised, and the wound was closed in layers over a negative suction drain, which was removed on the 2nd postoperative day. The postoperative period was uneventful, and the child was discharged in fair health on the 3rd postoperative day.

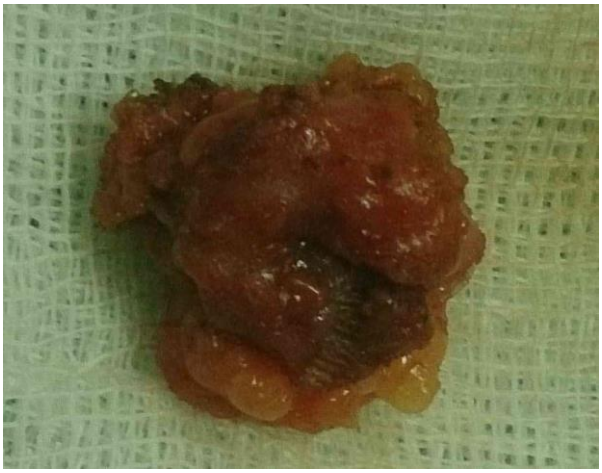

Fig. 3. The appearance of the excised cystic masses.

The cystic lesion was totally excised. Microscopically, dilated lymphatic channels, attenuated endothelial cells, lymphatic fluid and several lymphocytes within the lymphatic canal and smooth muscle fibers in the vessel walls were observed. The endothelial cells had no atypia or mitosis. Immunohistochemical staining revealed positive staining of the endothelial cells for CD31 and CD34. In addition, the tissue samples were stained weakly in the patchy style with vWf and negative with calretinin (Fig. 4. A-C). The histopathological investigation and immunohistochemical staining results were consistent with the preliminary diagnosis of cystic lymphangioma. During the 1-year follow-up, no problem was encountered.

Discussion

Lymphangioma is a benign congenital malformation resulting from a communication

failure between the primitive lymphatics and venous channels, and it leads to the proliferation of lymphatic vessels and the formation of a cystic structure [1,6]. Histologically, lymphangiomas are divided into three subtypes: cystic, capillary, and cavernous.

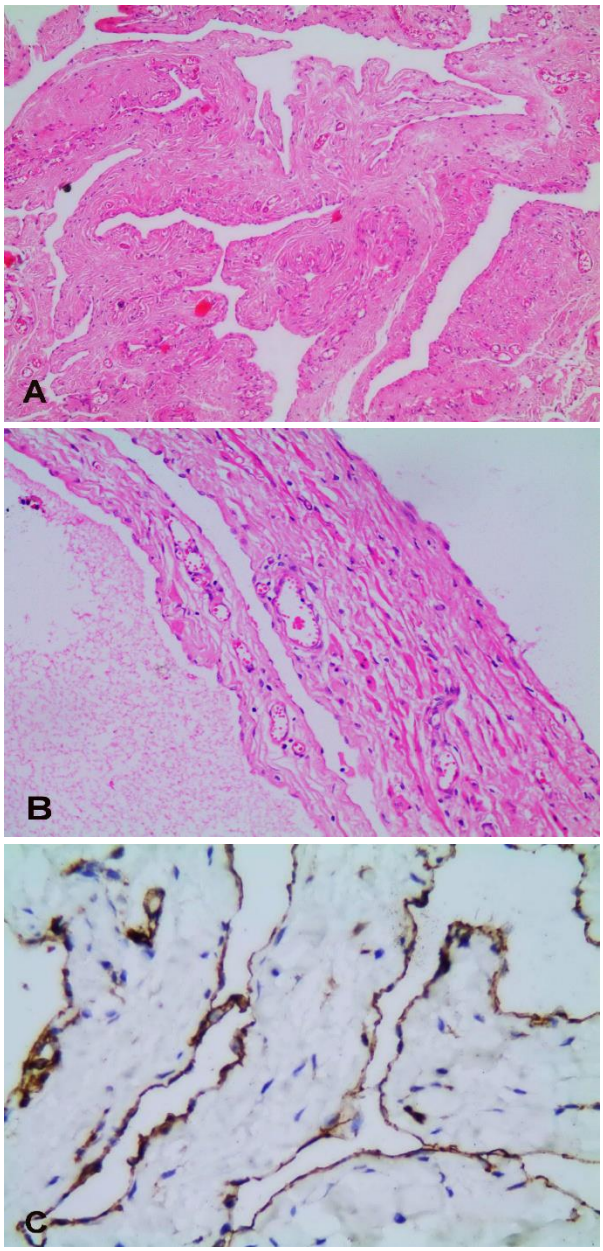

Fig. 4. A. Dilated lymphatic channels, lined by attenuated endothelial cells (H&Ex400). **B.** Lymphatic fluid within the lymphatic canal and the presence of several lymphocytes (H&E x200). **C.** CD31 expression in the endothelial cells, x400.

The cystic type is the most common type of lymphangioma [7]. Lymphangioma localized in the perineal region is usually an extension of an abdominal lymphangioma. However, isolated perineal lymphangiomas have been reported in several cases in the literature [3].

The scrotum is a very rare site for lymphangioma [1,8,9]. Differential diagnoses of perineal lymphangioma include dermoid cyst, cystic hemangioma, lymphocele and mesothelial cyst [3,10]. Differential diagnoses of scrotal lymphangioma include hernia, hydrocele, varicocele, wide epididymal cyst, acute scrotal conditions and filariasis [2, 11,12]. Ultrasonography is an important diagnostic tool for the identifying cystic lymphangioma and shows a multicystic mass with internal septae [1,2,8].

Treatment of cystic lymphangioma involves the complete removal of the entire mass, together with the skin. Recurrence is infrequent, but incomplete excision may result in a relapse. An isolated perineal-scrotal lymphangioma has not been reported previously in the English literature. In our case, total excision of the mass was performed, and no recurrence developed during the long-term follow-up.

Compliance with ethical statements

Conflicts of Interest: None.

Financial disclosure: None.

Consent: All photos were taken with parental consent.

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