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RESEARCH ARTICLE

ELBOW CYSTIC LYMPHANGIOMA IN CHILD: A CASE REPORT

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ABSTRACT

Cystic lymphangioma is a rare type of tumor of the lymphatic system, benign and typical during infancy, and are generally congenital. They are basically dilated lymph nodes with an endothelial covering, and serous or chylous contents. This tumor is located in the neck in 75% and axilla in 20% of cases. Involvement of the upper limb and particularly the elbow is very rare. We report on a case of macrocystic lymphatic malformation localized at the elbow. It was a 14 years old adolecent. Was followed up in our department because of congenital subcutaneous soft, painless mass of the left elbow. Ultrasonography and CT scan were evocative of the diagnosis of macrocystic lymphatic malformation. Surgical resection was complete without postoperative recurrence.

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INTRODUCTION

The cystic lymphangioma are benign tumors. They come through chromosomal abnormalities or complex angiodysplasias (Montes, 2007); recently called cystic lymphatic malformations (CLM), are mature lymphatic malformations, hemodynamically inactive, consisting of abnormal lymphatic vessels and cysts size and shapes (Wierzbicka, 2006) Macro cystic lymphatic malformations (MCML) (or cystic lymphangioma or cystic hygroma) is a circumscribed variant of deep lymphangiomas whose expansion is easy (Herbreteau *et al*., 1992).We report an original case of an unusual site of CLM and we raise the therapeutic difficulties of these malformations.

PATIENT & OBSERVATION: It was a 14 years old boy, with no significant pathological history, followed in our department for a congenital subcutaneous swelling of the left elbow; this mass gradually increasing in size. Clinical examination found left elbow antero-internal border subcutaneous swelling mesuring 12* 6 cm with a brachial perimeter facing the 19 cm (Figure 1). At palpation, this swelling was painless, soft, not swinging, and slightly mobile relative to the deep plane limiting the left elbow flexion. The remainder of the locomotor and somatic examination was without abnormalities. The ultrasound of the soft tissues revealed an antero-internal surface of the left elbow fold of a multi-partitioned superficial cystic formation of liquid content, well limited, measuring 94mm of major axis and 47mm of transverse diameter, multiple cystic areas, separated by thick walls with the existence in Doppler mode of a venous flow and a humeral pedicle, was without anomalies passing within the mass at a distance from its wall. CT Scan revealed a multilocular cystic mass with a hypodense liquid content allowing to retain the diagnosis of cystic lymphangioma of the left elbow crease (Figure 2).

Complete surgical excision was performed with an internal approach to the left elbow crease (Figure 3). Examination of the operative piece showed a multi cystic mass with hematic content. Histological study of conjunctivo-adipose tissue; found a proliferation of lymphatic vessels (vascular type).

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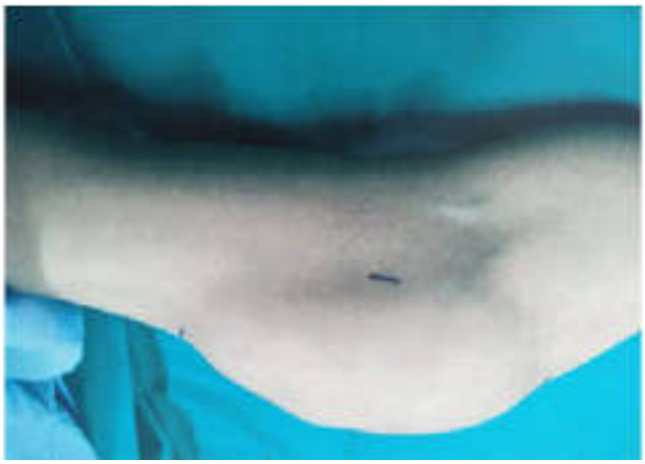

Figure 1. The figure shows appearance of the right elbow mass

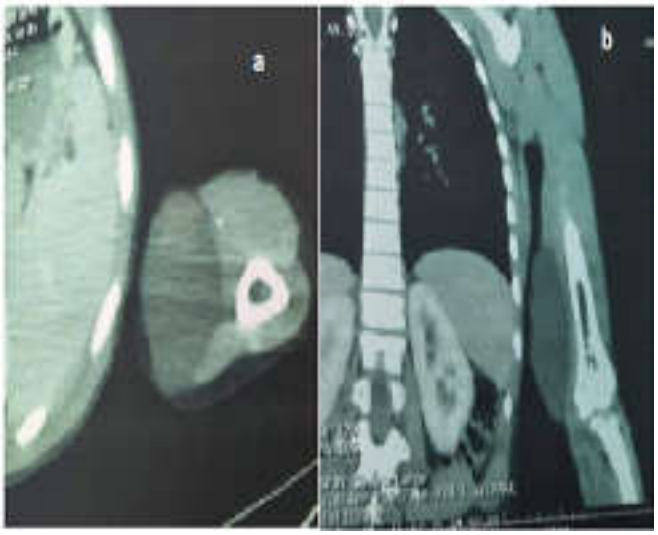

Figure 2. (a) axial, (b) coronal planes showing a low fluid density tumor

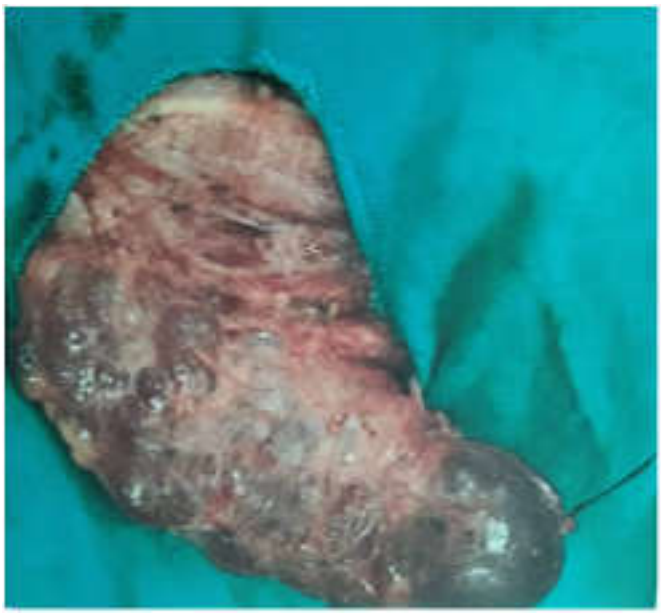

Figure 3. Perioperative image of the cystic lymphatic malformation

This proliferation is organized in cystic cavities of variable size lined by a flattened endothelial epithelium largely abraded and supported by a connective tissue seat of myxoid rearrangements: aspect of cystic lymphangioma. The immediate surgery care was simple. The left upper limb remained warm, with no sensory motor deficit. The infant was reviewed regularly on consultation rhythm of 3 months; for a period of 12 months. The lesion had clinically disappeared.

DISCUSSION

Lymphangioma is a rare benign vascular malformation of the lymphatic system of the childhood composed of cystically dilated lymphatics (Wierzbicka, 2006). Their incidence is reported to be 1.2-1.8 per 1000 of new births (Filston, 1994). There are several classifications; the most commonly used divide them in macro cystic, micro cystic and mixed lesions, depending on whether the volume of the cystic spaces is less than or greater than 2 cm3 (Herbreteau, 1992). Some authors prefer to classify them as simple or microcystic (formed by lymphatic capillaries), cavernous (formed by bigger lymphatic vessels with a fibrious adventicia), and cystic lymphangiomas (CL) also known as cystic hygromas.

Cystic lymphangioma noncommutating masses range from millimeters till centimeters in size (Grasso *et al.*, 2008; Bill, 1965). These tumors are very rare in the extremities (Rossi, 2004). In a recent and large series study, 11 of 186 patients (6%) were presented with lymphangiomas of the upper limbs (Wierzbicka *et al.*, 2006). Mirza *et al.* reported 2 cases of cystic lymphangioma of the sternum and a case of upper extremity CL in a 2-month-old male (19). Furthermore, Greenbaum *et al.* reported a case of a 2-year-old female with an elbow lymphangioma (Greenbaum, 2004).

The diagnosis of cystic lymphangioma is made on the basis of the tumor clinical aspect realizing soft, lobulated, renitent swellings, not attached to the underlying skin plane and not very mobile with respect to the deep planes (Sheth, 1987). Ultrasound is very useful and sensitive in detecting cystic masses. It is superior in terms of compliance and avoidance of the use of anesthetics needed in MRI or CT. It is also useful in assessing postoperative complications and recurrences (Murase, 1992). CT scan shows a low fluid density tumor (10-36 HU) but the partitions are sometimes revealed only after injection of the contrast agent (Wierzbicka, 2006; Poyraz *et al.*, 2004).

Magnetic resonance imaging would appear to be useful for exploring this tumor, reveals the characteristic aspect of hyposignal in T1 and hypersignal in T2. Only histology allows a diagnosis of certainty (Montes, 2007; Wierzbicka, 2006).
Therapeutically, several means are available such as sclerotherapy, surgery, laser and radiotherapy. The indication depends mainly on the micro or macro cystic type of the lymphatic malformation and its anatomical seat (Rossi *et al*., 2005; Rossi, 2005).

The therapeutic choice must take account of the benign character of the tumor, the symptoms, the operative complications and the risks of recurrence (Hadj-Henni, 2008). In the first intention, the MCLM are rather a treatment by sclerotherapy. Surgical treatment is reserved for microcystic malformations. Complete surgical excision was performed in our case with rigorous postoperative surveillance for one year (Wierzbicka, 2006). The removal of cystic lymphangioma can sometimes be difficult because of the anatomical seat of the lesion or its extension. Thus, the complete removal of the lymphangiomas sometimes requires several interventions.

CONCLUSION

Cystic lymphangiomas are uncommonly present in the upper extremities. The diagnosis is usually made by imagery, but histology is indispensable for confirmation. Treatment is surgical as these lesions rarely regress spontaneously especially if they are symptomatic and causing musculoskeletal dysfunction.

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