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# Cystic lymphangioma of the elbow in an infant: A case report

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ABSTRACT

The macrocystic Lymphatic malformations are 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 18 month old infant. Was followed up in our department since the age of 6 months because of congenital subcutaneous soft, painless mass of the right elbow. Ultrasonography and the magnetic resonance imaging (MRI) were evocative of the diagnosis of macrocystic lymphatic malformation. Surgical resection was complete without postoperative recurrence. MRI is useful for the diagnosis and the definition of tumor limits. The Treatment is difficult because of their location and rough delimitation.

## 1. Introduction

Cystic lymphangiomas are benign tumors. They occur exceptionally in the context of chromosomal abnormalities or complex angiodysplasia [1]; more recently known as cystic lymphatic malformations (CLM), are mature, hemodynamically inactive lymphatic malformations, consisting of abnormal lymph vessels and cysts of varying size and shape [2]. 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 $cm^3$ [3]. Some authors prefer to classify them as capillary lymphangiomas, cavernosal and cystic hygromas [4]. Macro cystic lymphatic malformations (MCLM) (or cystic lymphangiomas or cystic hygromas) constitute a circumscribed variant of deep lymphangiomas with easy expansion [5]. We report an original observation by the unusual seat of the CLM of the elbow in an infant and we raise the therapeutic difficulties of these malformations.

## 2. Patient and observation

It was a small infant 18 months old, with no significant pathological history, followed in our department since the age of 6 months for a congenital subcutaneous swelling of the right elbow; this mass gradually increasing in size. Initial clinical examination found a subcutaneous swelling of the antero-internal border of the right elbow crease of 7.5 * 4.5 cm of major axes with a brachial perimeter facing the 14 cm lesion (Fig. 1).

At palpation, this swelling was painless, soft, not swinging, and slightly mobile relative to the deep plane. The remainder of the locomotor and somatic examination was without abnormalities.

The ultrasound of the soft tissues revealed an antero-internal surface of the right elbow fold of a multi-partitioned superficial cystic formation of liquid content, well limited, measuring 64 mm of major axis and 35 mm of transverse diameter, multiple cystic areas (Fig. 2), 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 (Fig. 3).

MRI revealed a multilocular cystic mass in T1 hyposignal and hypersignal T2 (Fig. 4) with a super-internal fleshy component allowing to retain the diagnosis of cystic lymphangioma of the right elbow crease.

No serum therapy session by Ethibloc W had been completed, because we have neither the means or the human expertise in sclerotherapy. Complete surgical excision was performed with an internal approach to the right elbow crease (Fig. 5-a/5-b).

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). 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 was simple. The right upper limb remained warm, with no sensory motor deficit. The infant was reviewed

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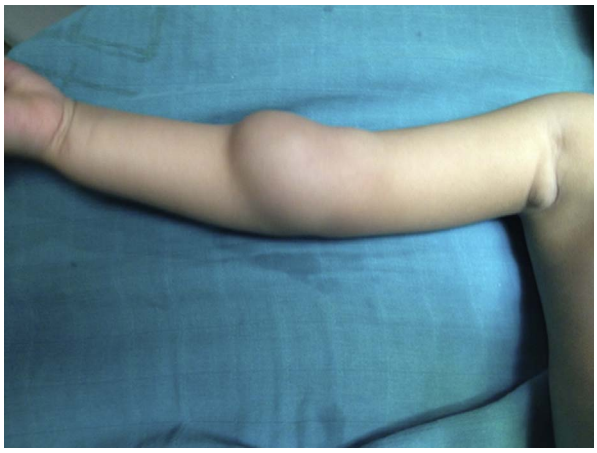

**Fig. 1.** Swelling of the right elbow crease in an infant.

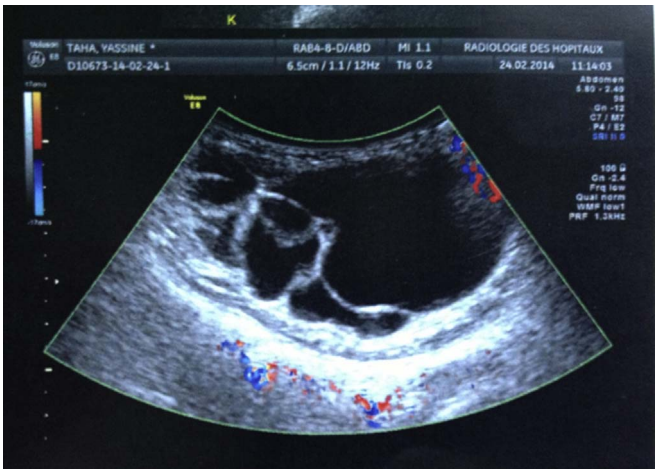


**Fig. 2.** Ultrasound appearance of a superficial cystic formation, multi-partitioned, fluid content and well limited.

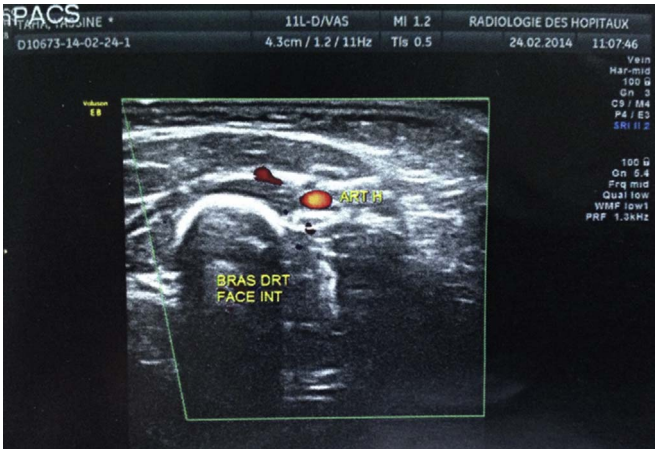


**Fig. 3.** Image of the Doppler showing the humeral pedicle without abnormalities.

regularly on consultation rhythm of 3 months; for a period of 12 months. The lesion had clinically disappeared; with persistence of some clinically palpable indurations in the right elbow fold (Fig. 6).

## 3. Discussion

The MCLM are located in the neck in 75% of the cases and in the axillary hollows in 20% of the cases, with possibilities of communication between these two locations by bridges passing under the clavicle. In 5% of cases, lesions are in the mediastin, retroperitoneum or pelvic region [5]. The involvement of the upper limb, and particularly of the arm, is very rare [6,7].

In the series of Sermon et al. On 19 patients with cystic lymphangiomas, 2 had brachial localization [8]. In that of Kennedy et al. [9], one limb injury was found in 8 out of 72 patients, including 2 in the upper extremity.

MCLM or cystic lymphangiomas are mature, hemodynamically inactive, congenital malformations of abnormal lymph vessels [10]. They produce round or lobulated and compressible masses. Their incidence is about 1 per 12,000 births. They are present at birth in 30–50% of cases and are found in 90% of cases before the end of 2 years [2]. Rarely, they appear in adulthood. MCLMs appear during embryonic development when the primary lymph bags do not fuse with the central venous system. The pathogenesis of MCLM is incompletely elucidated, but advances have been made thanks to molecular biology. Recent studies have shown the role of many regulatory genes including that of endothelial vascular growth factor receptors, VEGFR3 and VEGFR2, whose ligand is VEGF-C. These receptors intervene in the growth of the lymphatic vessels in the skin without influencing the development of the blood vessels [8,9]. Moreover, it has been observed that the endothelial cells of the MCLM secrete a large quantity of basic fibroblastgrowth factor (bFGF) which is an inducer of angiogenesis. Furthermore, the level of an inhibitor of angiogenesis (thrombospondin), secreted by the cells of the lymphatic malformations is lowered. Thus, the malformations would result from a runaway angiogenesis [11,12].

The diagnosis of cystic lymphangioma is made on the basis of a clinical aspect of the lesions realizing soft, lobulated, renitent swellings, not attached to the underlying skin plane and not very mobile with respect to the deep planes. Ultrasound has an interest in positive and sometimes differential diagnosis [13].

It shows multilocular cystic masses with septas of variable thickness. The content is anechoic, hypoechoic or hyperechoic depending on whether the lymph fluid is infected, hemorrhagic or hyperlipidic. The MRI reveals the characteristic aspect of hyposignal in T1 and hypersignal in T2. It also allows us to better appreciate the extension of the tumor according to different cuts (sagittal, transverse) and the achievement of adjacent clinically non-suspect structures thus providing a valuable aid to surgery. 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 [12,14]. In the first intention, the MCLM are rather a treatment by sclerotherapy.

Several sclerosing agents have been tested: hypertonic salty serum, lipiodol, bleomycin and boiling water. Ethibloc W, the most widely used sclerosing agent in France, seems to give good results in the treatment of MCLM [15]. This agent polymerizes rapidly in contact with the blood and is at the origin of biodegradable emboli which resorb in 4–6 weeks after injection. It thus leads to a giganto-cellular reaction necessary for the collapse of the walls of the cyst. It hardens and disappears in 2–6 months. The most frequent complications are febrile inflammatory reactions and oozing ulcers. The results of this method are excellent or good in 60% of the cases.

The therapeutic choice must take account of the benign character of the tumor, the symptoms, the operative complications and the risks of recurrence [10]. Surgical treatment is reserved for microcystic malformations. Complete surgical excision was performed in our case with rigorous postoperative surveillance for one year [2]. 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.

## 4. Conclusion

In conclusion, the authors report in light of this observation the