Management of A Giant Cervical Cystic Lymphangioma: About A Case
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Abstract

Introduction: Cystic lymphangiomas are dysembryopathies of the lymphoganglion system, responsible for a tumor syndrome by more or less exuberant angio-lymphatic proliferation, but histologically benign. We report a case of giant cervical cystic lymphangioma in an infant with postnatal diagnosis in order to discuss the particularities related to its management. Observation: This was a 7-month-old male child, with no significant pathological history, seen in consultation for a right laterocervical tumefaction observed from birth, evolving gradually. The swelling measures approximately 16x10 cm, painless, of soft consistency, covered with healthy skin, mobile in relation to the superficial plane. The diagnostic hypothesis of a cystic lymphangioma has been raised. Cervical ultrasound and cervico-thoracic scan revealed a right cervical mass with a liquid component sitting between the jugular vein and the trachea on which it exerts a mass effect. The child underwent complete excision of the tumor under general anesthesia. The pathological examination performed on the surgical specimen concluded to a cystic lymphangioma. The postoperative course was simple. The evolution was favorable after a decline of 4 years.

Conclusion: Cystic lymphangioma is a rare benign lymphatic malformation, but potentially serious in its evolutionary characteristics. Keywords: Cystic lymphangioma, malformation, cervical, infant.

INTRODUCTION
Cystic lymphangiomas are dysembryopathies of the lymphoganglion system, responsible for a tumor syndrome by more or less exuberant angio-lymphatic proliferation, but histologically benign. These are rare benign lymphatic malformations, but potentially serious due to their evolutionary characteristics. The cervico-facial region is the site of predilection with a frequency estimated at 75% on average [1]. The symptomatology depends on the size and topography of the cystic formation. Treatment is essentially based on surgery [2]. We report a case of giant cervical cystic lymphangioma in an infant with postnatal diagnosis in order to discuss the particularities related to its management.

OBSERVATION
This was a 7-month-old male infant, with no notable pathological history, seen in consultation for right laterocervical swelling observed from birth with progressive evolution. The general state is preserved. The swelling measured approximately 16x10 cm, painless, of soft consistency. It is nonpulsatile, covered with healthy skin, mobile relative to the superficial plane (Figure 1). There were no palpable cervico-facial adenopathies. The intraoral examination is unremarkable. The diagnostic hypothesis of a cystic lymphangioma has been raised. Cervical ultrasound showed a voluminous multi-partitioned cystic mass with irregular contours, echogenic liquid content in places with liquid level (Figure 2). The cervico-thoracic CT scan showed a right cervical mass with a liquid component sitting between the jugular vein and the trachea on which it exerts a mass effect (Figure 3, 4).

Fine needle aspiration cytology revealed haematic and lymphocyte smear without cytological signs of malignancy. The blood test requested is specific. The procedure is performed under general anesthesia with orotracheal intubation. The tumor was excised by the right laterocervical cutaneous approach which made it possible to highlight a polycystic mass of pink color traversed by numerous vessels, of soft consistency allowing a clear liquid (Figure 5, 6). A meticulous dissection made it possible to detach the posteroinferior and superior pole of the tumour, then to free the adhesions of the right carotid region and the right parotid and submandibular glands. The right internal jugular vein was ligated then sectioned at its entry into the tumour. A one-piece resection is thus performed (Figure 7). The pathological examination performed on the surgical specimen concluded to a cystic lymphangioma. The postoperative course was simple. The evolution was favorable after a decline of 4 years (Figure 8).
DISCUSSION

Cystic lymphangiomas are rare malformations and represent 0.8% of benign tumors and 0.1% of benign cervico-facial tumors. They represent approximately 6% of soft tissue tumors and 25% of tumors of vascular origin in subjects under 20 years of age [3]. They can be observed at all ages, however about 50% are apparent from birth, as was the case with our observation, and 70 to 90% are apparent before the age of 2 years [4, 5]. The predominance of one sex varies according to the studies [3, 6] and the present observation concerned a male child. Of ubiquitous location, the preferential location of these tumors remains the posterior cervical triangle with the possibility of mediastinal extension in 10% of cases [4]. Other locations have also been mentioned: the location of the mesentery [7, 8], the scrotum [9], the orbit [10]. Three theories currently exist to explain the origin of congenital lymphangioma [3]. Some explain it by a defect of connection between certain peripheral lymphatic vessels and the lymphatic bags or between the lymphatic bags and the venous system [3, 5]. For others, the lymphangioma is a hamartomatous malformation that arises from the anatomical and functional sequestration of normal lymphatic cells that develop independently of the rest of the lymphatic system [11]. Finally, the sometimes locally aggressive and recurrent behavior has led some to consider the lymphangioma as a real benign embryonic neoplasia, with a capacity for lymphangioproliferation and active secretion.
The clinical symptomatology depends on the size of the tumor and the topography of the cystic formation. Apart from the palpable cervical mass, cystic lymphangiomas have no clinical specificity. Thus the circumstance of discovery of cervical cystic lymphangiomas is sometimes a revealing symptomatology such as cervical mass as observed in our case.

Antenatal ultrasound can detect cystic lymphangioma in two-thirds of cases [4, 12, 13] and locate the swelling in relation to the cervical spine. It appears as a polylobed mass with thin walls, including anechoic cysts of variable size. Cystic contents may be heterogeneous. In this observation, no concept of antenatal diagnosis was reported by the parents. Postnatal ultrasound is particularly interesting in cervical forms of cystic lymphangioma. Indeed, it makes it possible to distinguish the cystic lesions from the solid lesions, to evoke the diagnosis, to specify the volume, the limits and the reports/ratios of the tumor, in particular compared to the vessels of the neck, thanks to the echo-Doppler. She can also look for a mediastinal extension and specify the macro or micro cystic character of the lymphangioma. The data provided by the scanner or the MRI make it possible to consider the most suitable therapeutic method and total excision when the surgical option is chosen. The scanner reveals a multi-partitioned mass, with a hypo-dense liquid density, which does not capture the contrast product except at the level of the walls [2, 14]. MRI seems to give more specific images with a hyper signal in a T2-weighted sequence reproducing the different partitions and thus delimiting the extent [14]. It was not performed in this observation. The high cost of this examination limits its realization in our context. Three histological criteria characterize cystic lymphangiomas: it is a cystic formation; containing septa with connective stroma, the thickness of which varies according to the age of the cyst, provided with lymphoid tissue and smooth muscle, a key element for diagnosis; with endothelial coating, attaching the tumor to a lymphatic vascular origin [3, 4]. We thus find the two macroscopic types, micro and macrocystic lymphangiomas, and a third microscopic type: capillary lymphangioma comprising small vessels with a narrow lumen [3, 4, 14].

The treatment is essentially surgical allowing the complete excision of the tumor essential to have a complete cure. There remains a debate as to the most opportune age for this surgery. In this case, the intervention was performed at 7 months. We are in agreement with many authors who consider that it is necessary to wait until the age of 6 to 9 months, so that general anesthesia carries less risk and to allow scheduled surgery after a complete assessment [3]. Others believe that early surgery is preferable, before inflammatory flare-ups make dissection difficult due to fibrosis and neovascularization. Surgical excision obyes the rules of lympho-nodal surgery of the neck, which is always long and meticulous, all the more difficult since it involves surgery in children. In the reported case, the surgery lasted about 5 hours. The mortality rate after surgical resection ranges from 0 to 7% depending on the series [3,5]. The main causes of death are airway obstruction, including accidental obstruction of the tracheostomy tube, and blood loss. Some authors observed in their series 54% of relapses in the first 3 months, and all the recurrences observed, took place before 14 months [15]. No lasting complication was noticed in this observation after a follow-up of 4 years.

Sclerotherapy may be indicated as an alternative to surgery. The principle is to inject a product into the cysts which will generate a giganto-cellular type inflammatory reaction, which subsequently turns into sclerosis or fibrosis responsible for stabilizing the development and then the virtual disappearance of the tumor [3]. It can be indicated in mono or paucicystic forms of cystic lymphangiomas and in diffuse macrocystic forms, whereas it has no place in microcystic forms. This technique, probably somewhat or less effective than surgery, has fewer side effects and complications [4]. It was not performed in this observation due to the large volume of the tumor.

CONCLUSION

Cystic lymphangioma is a rare benign lymphatic malformation, but potentially serious in its evolutionary characteristics. Surgical treatment with complete excision is the treatment of choice and is the only guarantee to avoid recurrences.

REFERENCES