Aggressive giant cystic lymphatic malformation in a newborn

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Lymphatic malformations are uncommon, benign and congenital malformations of the lymphatic system exhibiting lack of development of communication between the lymphatic and venous circulation. We report the unusual case of rapidly expanding giant lymphatic malformation of the torso, bilateral axillae and left upper extremity of a newborn. As the first-line treatment, aspiration and sclerotherapy with bleomycin were performed. The sclerotherapy failed to cause regression of the mass, and rapid expansion of the malformation necessitated surgery. Partial resection of the mass was performed. Clinical symptoms of respiratory distress resolved in the early postoperative period, and the patient became hemodynamically stable. However, intrathoracic invasion of the mass developed, leading to restriction of thoracic expansion, ending in death. In conclusion, surgical treatment of giant lymphatic malformations remains challenging.

Key words: giant cystic lymphangioma, newborn, surgery, bleomycin, thoracic invasion.

Lymphatic malformations are uncommon, benign and congenital malformations of the lymphatic system exhibiting lack of development of communication between the lymphatic and venous circulation. Typical histological features consist of a mass with numerous dilated, thin-walled vascular channels with aggregates occasionally being lymphatic in nature in the surrounding stroma[1]. Lesions are well-differentiated vessels and channels interconnected in a multicystic pattern[2]. There are two main treatment options for lymphatic malformations: surgical excision and sclerotherapy. If possible, total excision of the lymphatic malformation is the only potential "cure" for the disease[3].

We present the unusual case of a newborn with giant lymphatic malformation extending from the right side of the neck and thoracoabdominal region to the whole right upper extremity.

Case Report

At the 16th week of gestation, a lymphatic malformation was reported on the prenatal ultrasound. The antenatal course was uncomplicated. A 2260 g preterm (30 weeks) male infant was delivered at a university hospital by cesarean section secondary to fetal bradycardia to a 32-year-old G3P3 mother. He required several seconds of positive pressure ventilation (PPV), as his heart rate was less than 100 beats/min. Initially, he did not respond to PPV and after intubation, 0.5 ml intratracheal adrenalin was administered. Responding to the emergency resuscitation, the infant was transferred to the pediatric intensive care unit. The physical examination included lymphatic malformation extending from the right side of the neck and axilla, descending anteriorly to the thoracoabdominal region and posteriorly to the lower back region including the right upper extremity. Thoracoabdominal ultrasonography (USG) and magnetic resonance imaging (MRI) revealed dense fluid containing a giant multicystic lesion extending from the right side of the neck, including the right side of the thoracoabdominal wall, to the right forearm region and the proximal part of the right upper extremity (Fig. 1). The lesion demonstrated no mediastinal, intrathoracic or intraabdominal extension in the MRI, at that moment. As the first-line treatment, aspiration and sclerotherapy with intralesional bleomycin were planned.

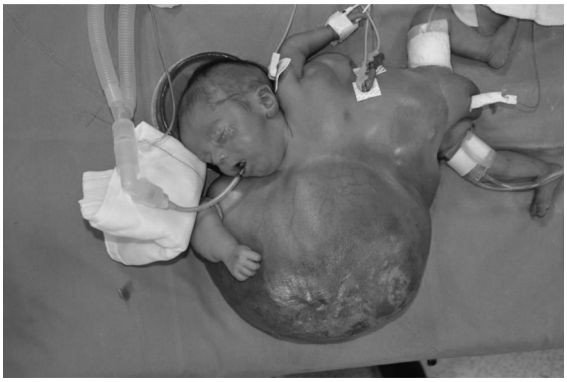

Fig. 1. Intraoperative picture taken before partial resection of the giant lymphatic malformation demonstrating the extent of the mass.

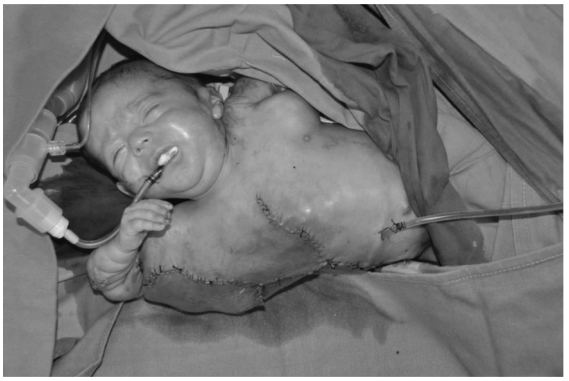

Fig. 3. Early postoperative picture after the partial resection of the mass.

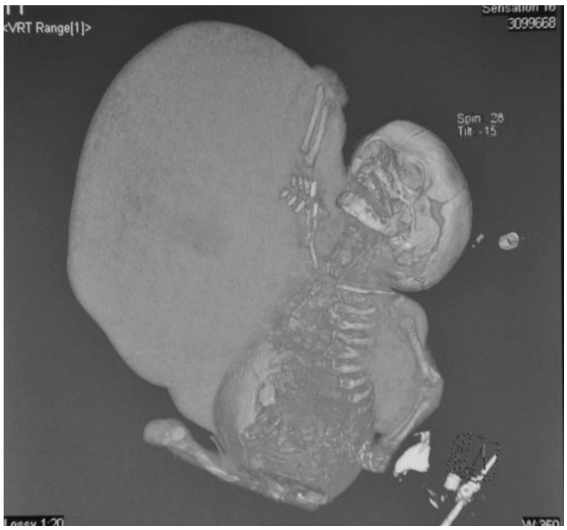

Fig. 2. A 3D CT image of the giant lymphatic malformation involving the torso, bilateral axillae and left upper extremity.

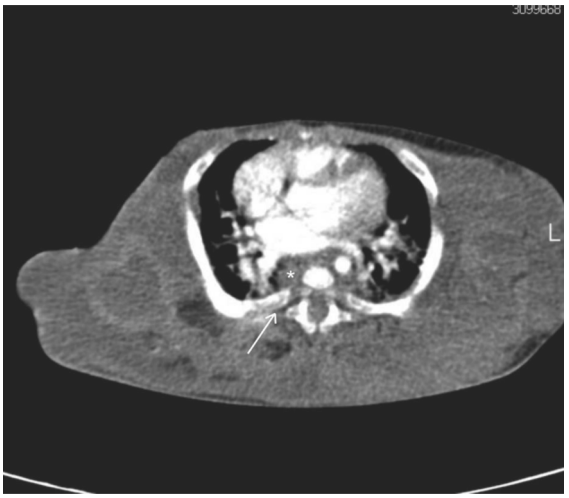

Fig. 4. CT image demonstrating the aggressive behavior of the mass. The intrathoracic invasion (asterisk) and site of entry (white arrow) to the thoracic cavity at the posterolateral border are marked. Note the close proximity of the intrathoracic portion of the mass to the aorta and pericardium.

Bleomycin (0.25 mg/kg) was administered followed by ultrasound-guided aspiration of 210 ml of cyst fluid (lymph and old blood). The interval between injections was four weeks. After four weeks, the same dose of bleomycin was administered following aspiration of 560 ml fluid. Although regression of the lymphatic malformation was expected, computerized tomography (CT) scan demonstrated a rapid increase in the size of the lesion (Fig. 2). The right humerus was luxated from the glenoid fossa and circulation of the overlying skin of the lesion was also threatened by the rapid expansion. Right-sided preexisting lesion of the left axilla and the left upper extremity expanded distally. The patient was hemodynamically unstable, and blood transfusion and fresh frozen plasma support were required periodically. Since the sclerotherapy failed to cause regression of the mass, surgery was decided. Partial resection of the mass involving the right thoracoabdominal, axillary region and upper extremity was performed (Fig. 3). Resection of the mass had to be stopped due to hypothermia that occurred during the operation. Clinical symptoms of respiratory distress resolved in the early postoperative period, and the patient became hemodynamically stable. The patient was transferred from the intensive care unit to the infant inpatient ward, and the second stage of excision of the remaining mass was planned. In the interim, intrathoracic invasion of the mass developed, leading to restriction of

thoracic expansion (Fig. 4), massive atelectasis, and finally to nosocomial pneumonia resulting in sepsis and exitus of the patient in the postoperative 10th week.

Discussion

Lymphatic malformations are classified as truncal versus extratruncal, microcystic (<2 cm) versus macrocystic (>2 cm) according to the diameter of the largest cystic cavity, or combined[4].

Although lymphatic malformations can be seen in any anatomic location, approximately 95% of lymphatic malformations occur in the neck and axillary region, and the remaining cases occur in the superior mediastinum, chest wall, mesentery, retroperitoneal region, pelvis, and lower limbs[5]. As lymphatic malformations are congenital anomalies, they are present at birth but not always evident. There is a great variability in the clinical presentation of the malformations depending on the anatomical localization of the lesion. Lymphatic malformations involving the head and neck region may present with airway obstruction, dysphagia, speech pathology, loss of vision, and cosmetic problems such as loss of oral hygiene and dentition, and finally prognathism and malocclusion by affecting the skeletal growth. Trunk and extremity malformations may lead to functional impairment, limb/girth discrepancies, and lymphedema[4]. The giant thoracoabdominal lymphatic malformation of our patient blocked thoracal expansion, which required invasive ventilation, threatened the circulation of the skin over the lesion, and luxated the right humerus from the glenohumeral joint.

Surgical removal is the method of choice for the treatment of lymphatic malformations. Other treatment modalities include aspiration, radiation and injection of sclerosing agents[6]. Sclerotherapy has emerging value in the current literature and may be an alternative to surgical removal for treatment of selected cases[7]. Bleomycin, tetracycline, inactivated OK-432, and 100% ethanol are used as first-line treatments through an intralesional percutaneous route for sclerotherapy[8]. There are reports regarding the success of bleomycin injection for the treatment of lymphatic malformations[8,9]. It is especially emphasized that bleomycin injection was more effective when administered for the treatment of macrocystic lymphatic malformations[9]. However, in this particular case, although the patient was suffering from giant multiple macrocystic lymphatic malformations, USG-guided application of bleomycin did not result in regression of the mass as expected. It has been observed that the size of the macrocystic malformation rapidly increased after the injections because of bleeding into the cystic space. Chronic hemodynamic instability of the patient required blood and fresh frozen plasma transfusion secondary to bleeding. This unpreventable condition and respiratory impairment became the major indications for surgical intervention.

Despite the risks of nerve and vascular damage, massive bleeding, scarring, and recurrence, surgery is the mainstay treatment modality for lymphatic malformations. However, traditionally giant lesions have been removed in staged resections[3]. Hong et al.[8] reported having partial resection as the only remaining option due to the extensive involvement of a lymphatic malformation of the tongue. Takamatsu et al.[10] reported a challenging case of giant lymphatic malformation involving the bilateral mediastinum.

Despite its benign histopathological character, the clinical behavior of the mass in the presented case was more like that of a malignant tumor. The unexpected response to sclerotherapy, rapid growth and intrathoracic invasion of the mass were remarkable.

Surgical treatment of giant lymphatic malformations is challenging, and the problems related to duration of the operation, closure of large defects and blood loss may determine the extent of the surgery and contribute to patient survival in newborns.

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