Case Report

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Giant cervical lymphangioma encompassing the neck great vessels (carotid artery and jugular vein): a case report

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Introduction and importance: Lymphangiomas are benign congenital defects affecting the lymphatic system. These lesions commonly involve the head and neck, predominantly the posterior cervical triangle. Lymphangiomas cause obstructive symptoms in the upper airway and pose an esthetic concern to the patient. Clinically, these lesions are seen as cervical swelling, and a definite diagnosis is made by ultrasonography, computerized tomography scanning, and histopathologic analysis. Herein, the author present an unusual case of an 18-month-old child presenting with a huge cervical swelling on the right side with extension to the carotid triangle (encompassing neck great vessels) as well as unilateral neck and fascial disfigurement. The patient was treated surgically with the complete excision of the mass and followed up with an excellent esthetic result.

Case presentation: An 18-month-old child was brought to our teaching hospital's pediatric surgery department with a huge cervical mass on the right side since birth. Following the completion of work-up with the laboratory and imaging (computerized tomography scan) diagnostic modalities, the patient was prepared for definite treatment. Our team approached the mass through a right neck hockey stick incision, and with the preservation of the neurovascular bundle, the mass was completely excised. The patient was followed up for 12 months on two occasions, with excellent esthetic results and no relapse.

Conclusion: Lymphangiomas confined to the posterior cervical triangle are a common problem in children. Lesions extending to the anterior of the neck, especially those involving the neck neurovascular bundle, are uncommon entities. The decision to proceed with sclerotherapy or surgical excision should be justified, provided that, during the surgical procedure, the neurovascular bundle is preserved and none of the vital organs (neurovascular components) are compensated with the goal of a complete mass excision.

Keywords: Esthetic, excision, lymphangioma, lymphatic system, neurovascular

Introduction

Lymphangiomas, or lymphatic malformations, are isolated regions of lymphatic tissue resulting from the failure of proper lymphatic anastomoses during the development phase[1–3]. Lymphangiomas are considered benign lesions and mostly occur in children[4]. Improper anastomosis could occur between lymphatic tissues, capillaries, veins, and arteries[5]. The problem is thought to result from an abnormal connection between the jugular sac and the peripheral lymphatic system[6]. The incidence is reported to be 1:6000 at birth and 1:750 among spontaneous abortions[7]. The common location of lymphatic vessel malformations is the neck, and the cervical region (75%) accounts for about 25% of all cysts of the neck. They may occur on both the front and rear triangles and may be unilateral or bilateral[8–10]. The majority of cervical masses are asymptomatic, with clinical outcomes ranging from aggressive infiltration into surrounding structures to complete spontaneous regression[11]. In the case of aggressive growth, they will cause considerable disfigurement and consequently lead to management problems[12]. This work has been reported in line with the Surgical Case Report (SCARE) Criteria[13].

HIGHLIGHTS

- Lymphangioma is a benign congenital malformation of the lymphatic system.
- The neck, particularly the posterior cervical triangle, is the most commonly involved site.
- In 80–90% of the cases, the lesion presents within 5 years; however, the presentation may occur as late as adolescence or adulthood.
- Intralesional injection of OK-432, a sclerosing agent, has shown good results in neonates, with the effects of decreasing mass volume and no feeding or respiratory complications.
- Giant cystic lymphangiomas of the neck are best managed by surgical incision.

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A Case report from Afghanistan with literature review

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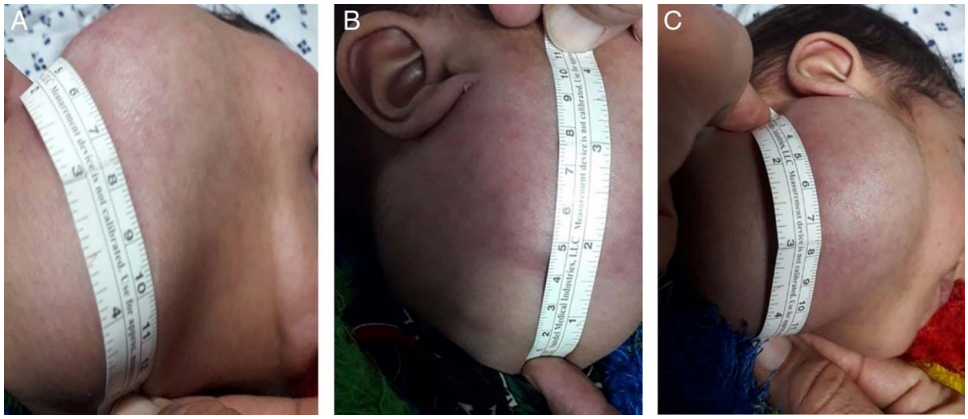

Figure 1. A, B, and C: Physical views of the right cervical mass with its two-dimensional measurement (13 × 12 cm).

Case presentation

Herein is presented an 18-month-old child with a giant cervical mass from the northern Faryab province of the country. The patient was born to a consanguineous couple with no positive history for the mentioned problem or others. She has had a small swelling in the right cervical area since birth. Initially, the parents took their patient to the district hospital and got conservative advice with no result, but the mass was growing. Thereafter, the parents visited a general surgeon in a private clinic. The surgeon examined the patient and punctured that using a small-bore needle with a bloody result, therefore referring the patient to our teaching hospital pediatric surgery department.

Our team examined the patient physically and noted a huge mass measuring (13 × 12 cm; Fig. 1A, B, and C) in its transverse and craniocaudal diameters. We admitted the patient to our pediatric surgery ward and advised all routine and biochemical laboratory tests, along with computerized tomography scan (CT-Scan) imaging. A blood test revealed leukocytosis with a high-grade fever, and a CT-Scan showed a mass occupying the entire posterior and much of the anterior cervical triangles, extending to the carotid triangle (Fig. 2A and B).

The patient was discussed, and we decided to schedule her for surgery. As the patient's clinical picture was not fit for operation, we kept her in our ward and put her on antibiotic therapy. After a week on appropriate antibiotic therapy, the fever subsided and the white blood cells returned to normal. The patient was prepared for surgery, and we approached the lesion through a right cervical transverse incision (typically in the shape of a hockey stick). Following a skin incision, dissection of the platysma muscles along with the superficial cervical fascia were done using diathermy. The mass was found to extend to the supraclavicular region on the right side, inferiorly to the right zygomatic arch, superiorly with mass effect over the trachea and extension into the retropharyngeal, and encompassing the neck great vessels (external and internal carotid arteries).

The lesion was meticulously dissected from its above-mentioned boundaries using scissors, artery forceps, and bipolar

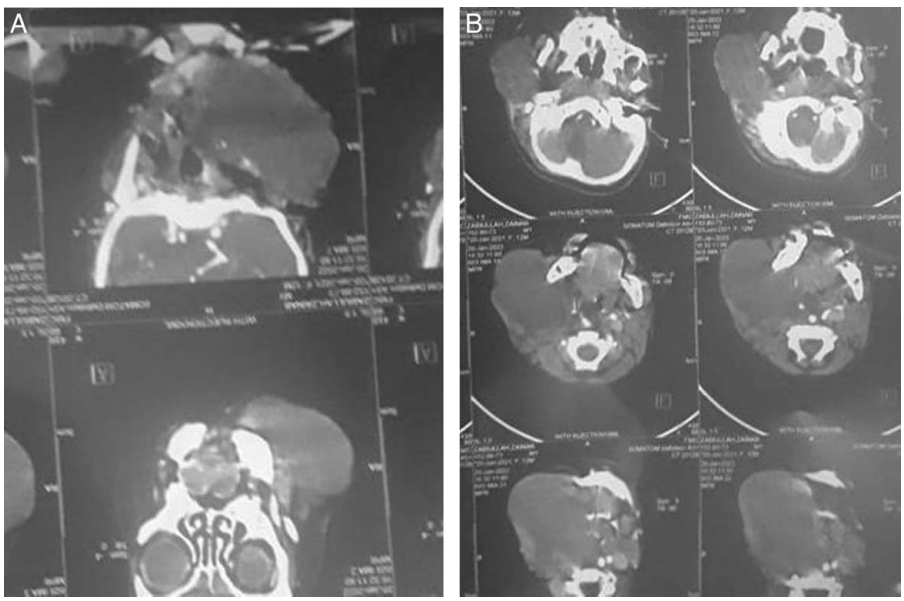

Figure 2. A and B: Computed tomography scan images of the lesion, revealing the cystic nature of the mass.

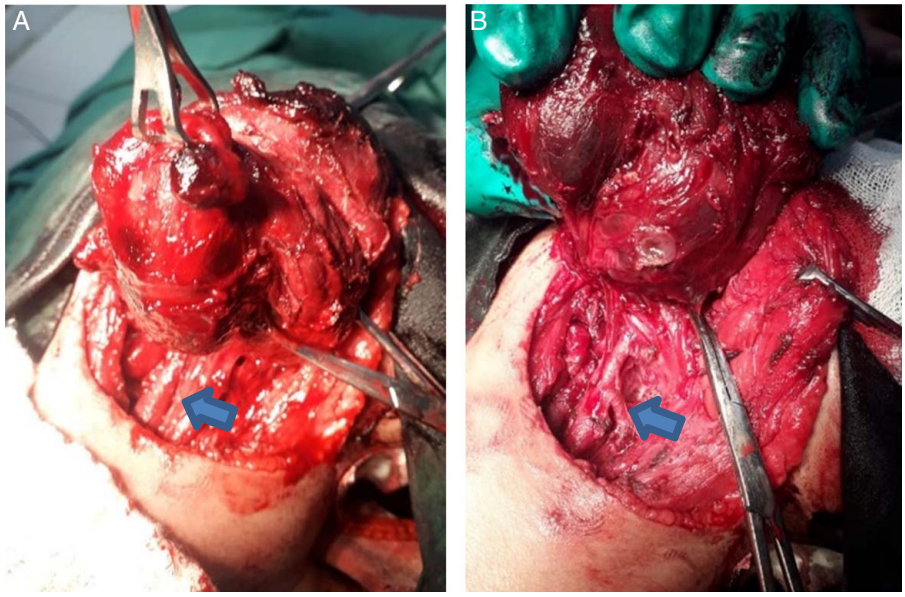

Figure 3. A and B: Operative steps of the mass (visible neck great vessels, arrows).

cautery. The most difficult part of the dissection was also completed successfully without causing any damage to the neurovascular bundle (branches of the carotid artery, jugular vein, hypoglossal, and vagus nerves). The wound was repaired layer by layer with leaving a closed-suction drainage inside, and the operation ended uneventfully (Fig. 3A and B).

The 24 h wound drainage was 25 cc on the first two occasions, 10 cc on the next two occasions, and 5 cc on the fifth day. We removed the drain, and the patient was discharged in satisfactory condition following a one-week hospital stay in the pediatric surgery ward. The patient was followed on two occasions (6th, and 12th months) with acceptable cosmetic and functional results (Fig. 4A and B).

Discussion and conclusions

Lymphangiomas or cystic hygromas are hamartomatous congenital malformations of the lymphatic system, most commonly seen in the cervico-facial regions (especially in the posterior cervical triangle), the axilla, the mediastinum, the groin, and the oral cavity. More than 95% of cases occur in the head and neck, with the remaining 5% occurring elsewhere, including the abdomen. Occasional sites are the liver, spleen, kidney, intestine, and other rare sites[14–18]. Cystic lymphangioma can be associated with chromosomal abnormalities (trisomies 13, 18, and 21; Noonan syndrome and Turner syndrome). Lymphangiomas are estimated to occur at a rate of 1.2 to 2.8 per 1000 live births[19]. The reported incidence by Gupta was 1/4000[20], and by Nicholls *et al*[21], 1/6500 live births. No racial or ethnic predispositions have been reported, and, in most reviews, the male to female ratio is equal[22–25], although some authors describe male predominance[26,29]. In 50–70% of the cases, the lesion is visible at birth or prenatally, and in 80–90% of the cases, it presents within 5 years[22,27,28]. However, presentation may occur as late as adolescence or adulthood. Natural resolution is uncommon but has been reported in up to 15% of cases[27,29].

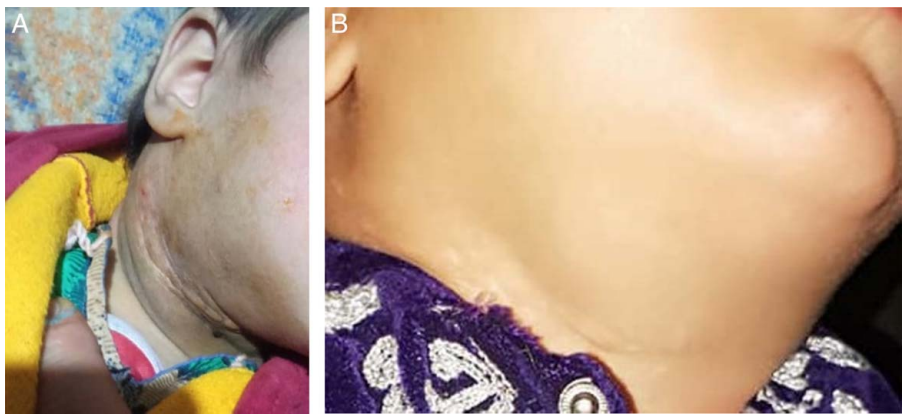

Figure 4. Postoperative views: (A) sixth postoperative month outcome. (B) 12th postoperative month outcome.

Lymphangiomas result from the failure of lymphatic tissues' and/or vessels' connection to the normal draining vessels during intrauterine life[14,15]. The origin of the abnormality is explained by three theories; the first theory suggests the arrest of the primitive lymphatic channels during embryogenesis, the second theory suggests that primitive lymphatic sacs fail to reach the venous system; while the third and most advanced theory suggests that during embryogenesis the lymphatic tissue lies in the wrong site[29]. Lymphangiomas are classified into three types: simplex (capillary), cystic, and cavernous, of which the last one is referred to as a cystic hygroma. Based on size, the lymphangiomas are classified into three types: microcystic (less than 2 cm), macrocystic (more than 2 cm), and mixed, which has variable sizes and may be composed of single or multiple cystic lesions[14,15]. The lesion of our patient measured 12×13 cm and was of a multicystic nature.

The common sign of lymphangioma is the presence of a mass, and depending on its size, it will cause respiratory obstruction and feeding problems. Cystic hygroma of the giant type will cause respiratory distress and dysphagia. Clinically, cystic hygromas are large, cystic, soft, and nontender masses. They transilluminate, unless complicated by hemorrhage or infection[14–17]. Airway and swallowing problems may persist after neck surgery due to mucosal edema, enlargement of internal lymphangiomas, and pharyngeal loss of innervation[30]. Lymphangioma could be diagnosed prenatally and postnatally. Ultrasonography (USG) diagnoses the lesion in mentioned both periods of life, but CT-Scan is superior to USG since the last modality is useful in the differential diagnosis and extent of the lesion to the deeper structures of the neck, mediastinum, and thoracic cavity[12,14,15,17,18,31].

Treatment indications for lymphangiomas are disfigurement, large size, rupture, infection, etc.[14,15,18]. Respiratory distress and dysphagia are conditions that require emergency treatment. The best treatment modality for curing cystic lymphangiomas is complete surgical excision. Simple drainage, aspirations, steroids, sclerotherapy (OK-432, monoclonal antibody, bleomycin), radiation, laser excision, radio-frequency ablation, and cauterization are other treatment options, each with advantages and disadvantages. Our decision to proceed with the surgical procedure was based on respiratory distress and disfigurement.

Since lymphangiomas are benign conditions, the vital organs should not be injured or sacrificed during surgical resection. Generally, complete excision may be impossible in a single setting and may require additional operations[14–18,32]. Recurrence depends on the complexity of the lesion and the perfection of the excision. Simple lymphangiomas are completely excised and rarely recur, but complex cases that are completely excised recur in 10–27% of the cases, whereas incompletely resected lesions recur in 50–100% of the cases[33]. In case of large neck mass causing airway obstruction, the Ex-utero intrapartum treatment procedure with intralesional injection of OK-432 sclerosing agent is a proper choice because it has no feeding or respiratory complications. Giant cystic lymphangiomas of the neck, head, and oral cavity could be successfully treated by surgery with satisfactory results, but great care must be exercised during complete excisions[34].

Our case was a nontender, semi-mobile, multicystic mass with ecchymotic overlying skin, occupying the right cervical area, extending to the anterior of the neck, and forwarding to the carotid triangle, as well as encompassing the neck great vessels (branches of the carotid artery and jugular vein). During the ante-natal period, the patient was not under regular check-up by an OBS-GYN specialist since the family was living in a remote province far from the capital, where there was not a specialized pediatric center. In Afghanistan, OK-432 sclerosing agent is not available, and our justification for surgical intervention was respiratory problems and disfigurement. The optimal surgical result depends on a well-equipped surgical setting (adequate light, a cooperative assistant, a microsurgical instrument, and an experienced surgeon). In our case, the art of surgery was the successful mobilization of the lesion from the great vessels and its complete excision by careful and nontraumatic tissue handling, paying attention to the neck anatomy (especially the neurovascular buddle), and patiently approaching the case. The toughest part of the operation was the adherence of the great vessels inside the lesion, which were meticulously separated using small artery forceps, and the cauterization of the small interconnecting capillaries using bipolar cautery to minimize bleeding. During follow-up, the patient exhibited excellent cosmetic and functional results over a 6–12-month period of time. The limitations of our study included a lack of readily available diagnostic modalities in the public hospital, which would have allowed us to diagnose the lesion concisely and logically determine our preoperative plan with contrast material.

Ethical approval

Not applicable.

Consent to publish

Written informed consent was obtained from the patient's parents for the publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Author contribution

The entire process of the article from conceptualization to publication (including surgical procedure) was conducted and observed by Turyalai Hakimi.

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The author declares that he has no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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References

[1] Faul JL, Berry GJ, Colby TV, *et al*. Thoracic lymphangiomas, lymphangiectasis, lymphangiomatosis, and lymphatic dysplasia syndrome. Am J Resp Crit Care Med 2000;161:1037–46.
[2] Fujita Y, Satoh S, Nakayama H, *et al*. In utero evaluation and the long-term prognosis of living infants with cystic hygroma. Fetal Diagn Ther 2001;16:402–8.
[3] Gallagher PG, Mahoney MJ, Gosche JR. Cystic hygroma in the fetus and newborn. Sem Perinatol 1999;23:341–56.
[4] Kennedy TL, Whitaker M, Pellitteri P, *et al*. Cystic hygroma/ lymphangioma: a rational approach to management. Laryngoscope 2001;111(11 pt 1):1929–37.
[5] Davies D, Rogers M. Morphology of lymphatic malformations: a pictorial review. Aust J Dermatol 2000;41:1–7.
[6] Cheng LH, Wells FC. A multidisciplinary approach to recurrent cervicothoracic cystic hygroma in an adult. Br J Oral Maxillofac Surg 2004;42:66–8.
[7] DeCou MD, Jones DC, Jacobs HD, *et al*. Successful ex utero intrapartum treatment (EXIT) procedure for congenital high airway obstruction syndrome owing laryngeal atresia. J Pediatr Surg 1998;33:1563–5.
[8] Naidu SI, McCalla MR. Lymphatic malformations of the head and neck in adults: a case report and review of the literature. Ann Otol Rhinol Laryngol 2004;113(3 pt 1):218–22.
[9] Enjolras O. Classification and management of the various superficial vascular anomalies: hemangiomas and vascular malformations. J Dermatol 1997;24:701–10.
[10] Suzuki N, *et al*. Al prenatally diagnosed cystic lymphangioma in infantsJ. Pediatr Surg 1998;33:1599–604.
[11] Sinard RJ, Welling DB. Cervical lymphangioma with simultaneous skull base invasion and soft tissue regression. Ann Otol Rhinol Laryngol 1995;104:662–4.
[12] Siegel MJ, Glazer HS, Amour TE St, *et al*. Lymphangiomas in children: MR imaging. Radiology 1989;170:467–70.
[13] Agha RA, Franchi T, Sohrabi C, *et al*. for the SCARE Group. The SCARE 2020 guideline: updating consensus Surgical CAse REport (SCARE) guidelines. Int J Surg 2020;84:226–30.
[14] Fonkalsrud EW. Cystic hygroma: an overview. J Cutan Aesthet Surg 2010;3:139–44.
[16] Liu DK, Ma YC, Guo XN, *et al*. Surgical treatment of cervical giant cystic lymphangioma in children. Zhonghua Zheng Xing Wai Ke Za Zhi 2011;27:415–7.
[17] Kushwaha AS, Ghritlaharey RK, Budhwani KS, *et al*. Giant retroperitoneal cystic lymphangioma in a seven-months-old girl. J Indian Assoc Paediatr Surg 2007;12:161–6.
[18] Rani DV, Srilakshmi R, Malathi S, *et al*. Unusual presentation of a retroperitoneal lymphangioma. Indian J Paediatr 2006;73:617–8.
[19] Filston HC. Hemangiomas, cystic hygromas and teratomas of the head and neck. Semin Pediatr Surg 1994;3:147–59.
[20] Gupta B. Incidence of congenital malformations in Nigerian children. West Afr Med J 1969;18:22–3.
[21] Nicholls EA, King PA, McMullin ND. A decade of pediatric lymphangiomas. Pediatr Surg Int 1991;6:421–4.
[22] Hancock BJ, St-Vil D, Luks FI, *et al*. Complications of lymphangiomas in children. J Pediatr Surg 1992;27:220–4.
[23] Uba AF, Chirdan LB. Management of cystic lymphangioma in children: experience in Jos, Nigeria. Pediatr Surg Int 2006;22:353–6.
[24] Adeyemi SD. Management of cystic hygroma of the head and neck in Lagos, Nigeria; a 10year experience. Int J Pediatr Otorhinolaryngol 1992;23:245–51.
[25] de Serres LM, Sie KC, Richardson MA. Lymphatic malformations of the head and neck. Arch Otolaryngol Head Neck Surg 1995;121:577–82.
[26] Ameh EA, Nmadu PT. Cervical cystic hygroma: pre-, intra-, and postoperative morbidity and mortality in Zaria, Nigeria. Pediatr Surg Int 2001;17:342–3.
[27] Alqahtani A, Nguyen LT, Flageole H, *et al*. 25 years' experience with lymphangiomas in children. J Pediatr Surg 1999;34:1164–8.
[28] Fonkalsrud EW. Congenital malformations of the lymphatic system. Semin Pediatr Surg 1994;3:62–9.
[29] Kennedy TL. Cystic hygroma–lymphangioma: a rare and still unclear entit. Laryngoscope 1989;99:1–10.
[30] Grasso DL, Pelizzo G, Zocconi E, *et al*. Lymphangiomas of the head and neck in children. ACTA Otorhinolaryng Ologica Ita Lica 2008;28:17–20.
[31] Kaminopetros P, Jauniaux E, Kane P, *et al*. Prenatal diagnosis of an extensive fetal lymphangioma using ultrasonography, magnetic resonance imaging and cytology. Br J Radiol 1997;70:750–3.
[32] Seashore JH, Gardiner LJ, Ariyan S. Management of giant cystic hygromas in infants. Am J Surg 1985;149:459–65.
[33] Dillon P. Lymphatic and venous disorders. In: Oldham KT, Colombani PM, Foglia RP, editors. Surgery of infants and children: scientific principles and practice. Lippincott-Raven, Philadelphia. 1997: pp. 1727–1743.
[34] Mikovic Z, Simic R, Egic A, *et al*. Intrauterine treatment of large fetal neck lymphangioma with OK-432. Fetal Diagn Ther 2009;26:102–6.