

CASE REPORT

Giant cervical lymphangioma in a pediatric patient: a case report

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ABSTRACT

Lymphangiomas are rare benign congenital malformations of the lymphatic system. They vary in size and can cause compressive symptoms, potentially putting the patient's life at risk. Surgery is considered the most effective therapeutic option. We present the case of a one-year-old patient from Pucallpa, Peru, admitted to Hospital Amazónico with a deforming right cervical mass consistent with cystic lymphangioma and signs of local necrosis. The patient exhibited symptoms of dysphagia, spontaneous crying, pain, hyporexia, cough, and fever. Open surgery was performed, achieving complete excision of the mass while preserving the adjacent nervous and vascular structures. Pathological analysis confirmed the diagnosis of lymphangioma. The patient evolved favorably, being discharged 8 days post-surgery. No complications were observed during the one-year follow-up period. In this case, pathological examination of the mass established the diagnosis of lymphangioma, and open surgery allowed for complete tumor removal without major complications. It is hoped that the clinical experience presented will aid in the effective management of pediatric lymphangioma cases with similar presentations.

Keywords: Lymphangioma/surgery; Infant (Source: MeSH)

Linfangioma cervical gigante en paciente pediátrico: un reporte de caso

RESUMEN

Los linfangiomas son alteraciones congénitas benignas poco frecuentes del sistema linfático. Su tamaño es variable, pudiendo causar síntomas compresivos y poner en riesgo la vida del paciente. La cirugía es considerada la mejor opción terapéutica. Presentamos el caso de un paciente de un año de edad procedente de Pucallpa, Perú, que ingresó al Hospital Amazónico por presentar masa cervical derecha deformante compatible con un linfangioma quístico, con presencia de necrosis local. El paciente cursó con disfagia, llanto espontáneo, dolor, hiporexia, tos y fiebre. Se realizó una cirugía abierta con escisión completa de la masa y preservación de los componentes nerviosos y vasculares. La masa se caracterizó como un linfangioma según los hallazgos patológicos. El paciente evolucionó de manera favorable siendo dado de alta 8 días después de la cirugía. No se observaron complicaciones durante el año de seguimiento. En el caso presentado, el análisis patológico de la masa permitió establecer el diagnóstico de linfangioma; mientras que la cirugía abierta permitió extirpar el tumor sin mayores complicaciones. Se espera que la experiencia clínica presentada sea útil para el manejo efectivo de casos pediátricos de linfangioma con presentación similar.

Palabras clave: Linfangioma/Cirugía; Infante (Fuente: DeCS)

INTRODUCTION

Lymphatic malformations are rare, benign congenital anomalies, with 90% of cases presenting before the age of two (1). These lesions most commonly occur in the neck, particularly in the posterior cervical triangle (2). Although often asymptomatic, these conditions can lead to obstructive symptoms and pose a serious risk to the patient's life by compressing the airway or causing significant bleeding (3). Diagnosis is primarily clinical, supported by imaging techniques (ultrasound or tomography) and laboratory tests (2). Surgical intervention remains the treatment of choice (2). This case report describes the surgical management of a disfiguring, infected neck mass causing obstructive symptoms in a pediatric patient.

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CLINICAL CASE

A one-year-old male patient from a rural area in Pucallpa, Peru, was admitted to the emergency department of Hospital Amazónico with a right cervical mass. As detailed, this mass had been present since birth and had gradually enlarged over time. In the past three months, the mass had grown and prevented him from swallowing food. Two weeks before hospital admission, the patient developed spontaneous crying episodes, mild respiratory distress, pain, hyporexia, cough, and fever.

On examination, a soft tumor measuring 14 cm × 12 cm × 7 cm was identified, extending from the right lateral-anterior cervical region to the ipsilateral preauricular area, with signs of phlogosis. The mass caused a localized deformity that hindered cervical movements and elicited moderate tenderness on palpation (Figure 1A). In addition, the hemogram results showed 31.78×10^9 leukocytes/L with a left shift. Ultrasound revealed an extensive cystic mass with multiple septa, suggestive signs of cellulitis, and an abscess measuring 22 mm × 13 mm. The tumor area was tender during the ultrasound examination. A presumptive diagnosis of a right giant abscessed cystic lymphangioma with right cervical cellulitis was made.

The mass was successfully removed through surgical intervention at the right cervical region, with total resection of the tumor and preservation of the internal jugular vein, spinal nerve, and laminar drainage (Figure 2). Based on the results of pathological anatomy, the diagnosis of cystic lymphangioma with the presence of localized necrosis was confirmed. The patient was hospitalized in the general surgery department with favorable evolution (Figure 1B). He was started on a large liquid diet six hours postoperatively, with analgesia and antibiotic coverage with ceftriaxone and clindamycin for seven days. On the third postoperative day, there was scarce serohematic secretion, so the laminar drain was removed. The patient was discharged on postoperative day 8. He evolved favorably and did not present any complications during the year of follow-up.



Figure 1. Patient diagnosed with cervical lymphangioma. A (left): patient with a mass in the right cervical region. B (right): Patient on the third postoperative day.

Surgical intervention was performed, involving total resection of the tumor from the right cervical region, while preserving the internal jugular vein, spinal accessory nerve, and laminar drainage (Figure 2). Histopathological examination confirmed the diagnosis of cystic lymphangioma with localized necrosis. Postoperatively, the patient was admitted to the general surgery ward and showed favorable progress (Figure 1B). He was started on a clear liquid diet six hours after surgery, along with analgesia and a seven-day course of antibiotics (ceftriaxone and clindamycin). On postoperative day three, minimal serohematic drainage was noted, leading to the removal of the laminar drain. The patient was discharged on postoperative day eight. He demonstrated favorable recovery and remained complication-free during a one-year follow-up period.



Figure 2. Tumor mass removed during surgery

DISCUSSION

Lymphangiomas are rare hamartomatous tumors that develop from failure in lymphatic anastomoses during the embryonic period and may be associated with chromosomal abnormalities (1,4). They have an incidence of 1 in 6,000 live births, with a male-to-female ratio of 1:1 (5,6). Although 90% of cases occur within the first two years of life, occurrences in adolescents and adults have also been reported (1). Seventy-five percent of these tumors originate in the cervical region (lateral or posterior), as observed in this case. They can also manifest in other body regions, such as the axilla (20%), mediastinum (5%), and abdomen (kidneys, liver, spleen, and intestines) (2).

Lymphangiomas are classified into three histological subtypes: capillary, located in the subcutaneous tissue; cavernous, located in the tongue and mouth; and cystic. These tumors vary in size, grow slowly (2), and may present with obstructive symptoms or infectious complications. At the cervical level, lymphangiomas may manifest with dysphagia or dyspnea (3). In the case presented here, a disfiguring tumor that impaired cervical movement and caused dysphagia was reported, which later developed an abscess, leading to mild respiratory distress.

While tomography is the preferred technique to support the diagnosis, its use was not feasible in this case due to limited resources. Ultrasonography, a more accessible and cost-effective alternative, was utilized to support the diagnosis in this case (3,5). Surgical excision is the treatment of choice for lymphangiomas and can be performed using either an open or laparoscopic approach (5). Alternative treatments described include sclerosing agents in neonates to reduce tumor size, laser excision, and radiofrequency ablation (1,3,7). In this case, an open surgical approach was selected, achieving complete tumor excision while preserving nerve and vascular structures. This approach can be associated with

various complications, including hemorrhage, hematoma, seroma, nerve injury, and infection, which can occur in 10-15% of cases (2,8).

In this case, the patient experienced difficulty swallowing due to pressure, mild respiratory distress, hypoxemia, pain, cough, and fever, all of which resolved satisfactorily after surgery. During hospitalization, the patient showed adequate progress and oral intake. There were no signs of recurrence or postoperative complications at the one-year follow-up.

We present this case report of a pediatric lymphangioma diagnosed with ultrasound, an alternative more readily available than tomography. Pathology results allowed us to identify the mass as a lymphangioma, and surgery allowed us to remove it without major complications.

Author contributions

Conceptualization: ER, ZCC; data collection, management, and curation: RE, ZCC; data analysis: SR, ZCC; original version drafting: SR, ZCC; final version drafting and revising: SR, ZCC

Conflicts of interest

The present study was conducted in accordance with the ethical standards outlined in the Declaration of Helsinki. The authors declare no conflicts of interest related to the material presented in this manuscript.

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Ethical considerations

The authors declare that informed consent was obtained from the patient's parents or legal guardians for the use of the patient's information.

REFERENCES

1. Teissier N. Cervicofacial lymphangiomas: imaging, diagnosis, and treatment. *EMC Otorhinolaryngol.* 2017;46(2):1-15. doi: 10.1016/S1632-3475(17)83976-X
2. Ghritlaharey RK. Management of giant cystic lymphangioma in an infant. *J Clin Diagn Res.* 2013;7(8):1755-6. doi: 10.7860/JCDR/2013/5418.3256
3. Wittekindt C, Michel O, Streppel M, Roth B, Quante G, Beutner D, et al. Lymphatic malformations of the head and neck: introduction of a disease score for children, Cologne Disease Score (CDS). *Int J Pediatr Otorhinolaryngol.* 2006;70(7):1205-12. doi: 10.1016/j.ijporl.2005.12.013
4. Gallagher PG, Mahoney MJ, Gosche JR. Cystic hygroma in the fetus and newborn. *Semin Perinatol.* 1999;23(4):341-56. doi: 10.1016/S0146-0005(99)80042-1
5. Hakimi T. Giant cervical lymphangioma encompassing the neck great vessels (carotid artery and jugular vein): a case report. *Ann Med Surg (Lond).* 2023;85(5):1923-7. doi: 10.1097/MS9.0000000000000452
6. Li J, Zhong W, Geng X, Liu X, Zhang X, Wang Y, et al. Ultrasonographic diagnosis, classification, and treatment of cervical lymphatic malformation in paediatric patients: a retrospective study. *BMC Pediatr.* 2020;20(1):441. doi: 10.1186/s12887-020-02337-w
7. Okazaki T, Iwatani S, Yanai T, Kobayashi H. Treatment of lymphangioma in children: our experience of 128 cases. *J Pediatr Surg.* 2007;42:386-9. doi: 10.1016/j.jpedsurg.2006.10.012
8. Mirza B, Ijaz L, Saleem M, Sharif M, Sheikh A. Cystic hygroma: an overview. *J Cutan Aesthet Surg.* 2010;3(3):139-44. doi: 10.4103/0974-2077.74488