ORIGINAL RESEARCH PAPER
Volume - 9 | Issue - 6 | June - 2020 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

A RARE CASE OF A CHEST WALL LYMPH CYST – A CASE REPORT

Surgery

Jai Durairaj Paramasivam — Assistant Professor, Dept. of Paediatric Surgery, Saveetha Medical College & Hospital, Thandalam, KanchipuramDist – 602105.

Nikitha Moorthy — Resident, Dept. of General Surgery, Saveetha Medical College & Hospital, Thandalam, KanchipuramDist – 602105.

Sivarahini Ganesh — Resident, Dept. of General Surgery, Saveetha Medical College & Hospital, Thandalam, KanchipuramDist – 602105.

Surya Rao RaoVenkata Mahipathy* — Professor & Head, Dept. of Plastic & Reconstructive Surgery, Surgery, Saveetha Medical College & Hospital, Thandalam, KanchipuramDist – 602105. *Corresponding Author

ABSTRACT

Lymphangiomaand its variants are benign congenital malformations.We present an atypical case of a cystic hygroma of the chest wall in a 10month old child. The child came with a painless progressive swelling in left lateral chest wall.Examination revealed a swelling of 7 x 5cm along the left anterior axillary line from third to seventh intercostals spaces, ovoid in shape, mobile,tense but painless.Ultrasonogram revealed a cystic lesion in left chest wall. MRI showed a well defined cystic lesion with septations in the subcutaneous plane with intermuscular extensions favouring a lymphangioma.Surgical excision of the cyst was performed. Histopathology revealed as lymphatic cyst (macrocystic variant of lymphangioma).

KEYWORDS

Lymphangioma, Chest wall, Rare, Surgical excision

INTRODUCTION

Lymphangioma and its variants are congenital malformations originating from lymphatic hyperplasia. They are localised or extensive lesions of malformed lymphatic vessels in skin and deeper soft tissues, occurring due to sequestration of segments of primitive lymphatic vessels. They represent about 5-6% of all childhood tumours. They are most frequently located in head, neck and axilla, but isolated cystic hygroma of chest wall is a rare condition. Based on histology, the lymphatic malformations (LM) are divided into three subtypes: macrocystic LM, microcystic LM and mixed LM. Here, we present a rare case of cystic hygroma chest wall that underwent surgical excision.

CASE REPORT

A 10month old child was brought to us by the mother with complaints of painless swelling on the left lateral chest wall since birth. The swelling was spontaneous in onset and gradually progressive. On clinical examination,the swelling was 7 x 5cm along the left anterior axillary line from third to the seventh intercostals spaces, ovoid in shape, mobile, fluctuant& painless.**(Fig. 1)**No other swellings were noted in the neck or axilla.Transillumination was positive. Ultrasound showed a large cystic lesion with internal septations with no internal vascularity noted.A lesion of size 8 x 4 cm noted in subcutaneous plane in left lateral chest wall with no obvious intrapleural extension. Ultrasound of the neck and abdomen were normal. MRI revealed a well defined cystic lesion with septations in the subcutaneous plane with intermuscular extensions favouring a lymphangioma.**(Fig. 2)**After pre anaesthetic work up, the patient was planned for surgical excision.An elliptical incision was made over the swelling. A 10 x 7 cm cyst was seen in the subcutaneous plane extending to the intermuscular plane. Cyst wall along with the adherent muscle fibres were excised in toto and sent for histopathological examination.**(Fig. 3)**The cyst contained 50-60ml of lymph fluid which was drained. Hemostasis was secured and wound closure was done in layers. The postoperative period was uneventful and patient was discharged on the fifth post-operative day.**(Fig. 4)**Histopathologic examination showed fibrocollagenous tissue with congested blood vessels and areas of hemorrhage and dilated and collapsed lymphatic vessel lined by single layer of endothelium and filled with eosinophilic fluid material and suggestive to be lymphatic cyst. The patient is asymptomatic and on regular follow up without relapse.

DISCUSSION

The incidence of lymphatic malformations is 1 in 2000-4000 births. Lymphangiomas account for 5-6% of all paediatric neoplasms(1). Cystic lymphangiomas occur 65% at birth and 90% by the end of second year(1,4). In our case, we have diagnosed lymphangioma at 10months of age. Most common site of occurrence is 75% in head and neck,20% in axilla, 2% in retroperitoneum, intraabdominal, 2% in limbs and bones and 1% in mediastinum (3,5). Here we report this rare case of chest wall lymphangioma.Lymphangiomas occur due to lymphatic growth arrest, failure of lymphatics to connect to the venous system(2) or sequestrated lymphatic rests that retain their embryonic growth potential. It is a congenital anomaly, but can also occur due to trauma and inflammation. Chest wall lymphangiomas are mostly asymptomatic.Diagnosis can be made by physical examination, and transillumination differentiates cystic lymphangioma from solid tumours. USG with Doppler reveals cystic nature of lesions and adjacent blood vascular component of cystic lymphangiomas(6). MRI is highly sensitive and the best modality to diagnose lymphangiomas. It is used to determine the size of the lesion and infiltrations. T1 weighted images show hypointense and T2 with gadolinium shows a thin peripheral & septal enhancement. T2 weighted images show hyperintensity with thin septae(7,8,9). Cytogenetic study for determining aneuploidy and array comparative genomic hybridization can be done.

Immunohistochemistry markers LYVE-1 lymphatic vessel endothelial receptor-1 and D2-40 can be used to differentiate lymphangioma from hemangioma.Early diagnosis prevents further growth and infiltration of structures. The definitive treatment is complete resection of the lesion by preserving critical vascular and neural structures. Closed suction drainage system is necessary.Recurrence occurs when incomplete resection is done (10). Non surgical treatment includes chemotherapy and interferon- alpha (11,12). Intralesionalsclerotherapyis effective in treating and resolving macrocystic lesions but with less efficacy in microcystic lesions.100% ethanol is the most effective sclerosant used in localisedmacrocystic lesions. It causes rapid cellular dehydration of lymphatic endothelial cells, but is contraindicated in lesions adjacent to major nerves or cutaneous lesions for the risk of neuropathy and necrosis.OK-432 causes inflammatory response resulting in cytokine production by leukocytes and is used in macrocystic lesions. Bleomycin inhibits DNA synthesis and is also used in macrocystic lesions.Sodium tetradecylsulphate is a surfactant sclerosant. It damages the endothelial lining resulting in thrombosis, inflammationand subsequent fibrosis of the lesion and is used for cutaneous lesions. Doxycycline is used in extensive lesions. Adverse reactions after sclerotherapy include soft tissue oedema, skin necrosis & neuropathy. Lasers like carbondioxide laser, pulsed dye lasers and Nd-YAG laser are used to treat superficial

lymphatic malformations with good results. Relapse is common in incomplete removal of lesions and usually occurs within 3months. A case of chest wall lymphangioma that relapsed 19 years after surgery has also been reported(13). Hence, long term follow up of patients is very important to prevent relapse.

CONCLUSION

Appropriate prenatal evaluation &early postnatal counselling will result in better outcomes. A good physical examination with appropriate imaging is essential and complete resection will prevent relapse.

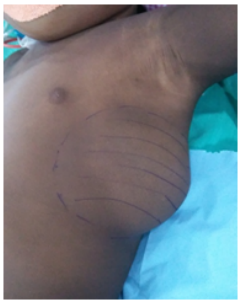
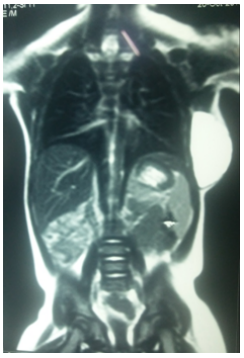


Fig. 1 – Clinical photograph showing the chest wall lymph cyst



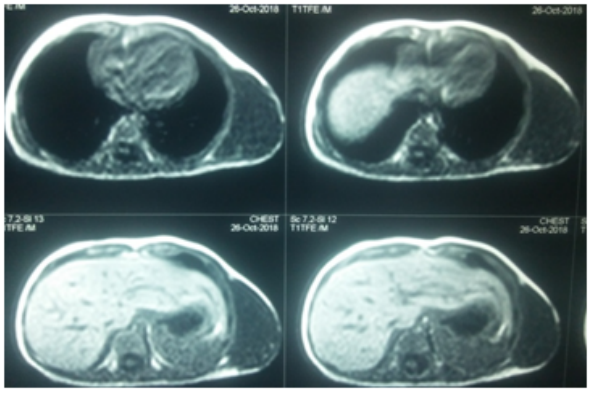


Fig. 2 – MRI showinga well defined cystic lesion with septations in the subcutaneous plane with intermuscular extensions favouring a lymphangioma

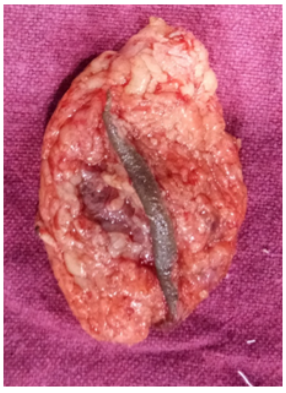


Fig. 3 – Specimen after excision

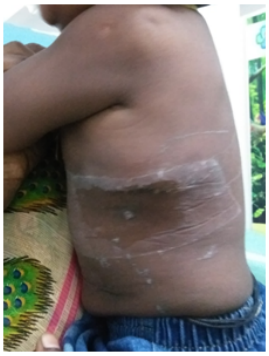


Fig. 4 – Photographon the 5th post-operative day

REFERENCES

1. Faul JL, Berry GJ, Colby TV, Ruoss SJ, Walter MB, Rosen GD, Raffin TA. Thoracic lymphangiomas, lymphangiectasis, lymphangiomatosis, and lymphatic dysplasia syndrome. Am J RespirCrit Care Med. 2000
2. Fonnkaisurd EW. Disorders of the lymphatic system. In: Welch KJ, Randolph JG, Ravitch MM, O'Neill JA Jr, Rowe MI, editors. Paediatric Surgery. Vol. 2. Chicago: Year Book Medical Publishers; 1986. p. 1506.
3. Robbins SL, Cotran RS, Kumar V. Blood vessels. In: Robbins SL, Cotran RS, Kumar V, editors. Robbins Pathologic Basis of Disease. Philadelphia: Saunders; 1984.
4. Güemes I, Ibáñez V, Barrios JE, Esteban MJ, Miranda J, Sáenz P, Vila-Carbó JL. Acute abdomen: other initial symptom of abdominal lymphangioma (in Spanish) Cir Pediatr. 1998;11:73–75.
5. Albanese CT, Wiener ES. Cystic hygroma. In: Spitz L, Coran AG, editors. Pediatric Surgery. London: Chapman & Hall; 1995. pp. 94–95
6. N. Yokoigawa, M. Okuno, and A.-H. Kwon, "Cystic lymphangioma of the chest wall: A case report," Case Reports in Gastroenterology. 2014; 8(3): 393-397.
7. Y. Nakazato, Y. Ohno, Y. Nakataa et al., "Cystic lymphangioma of the mediastinum," American Heart Journal. 1995; 129(2): 406-409.
8. D. Lu, Y. Wang, W. Zeng, and B. Peng, "Giant fetallymphangioma at chest wall and prognosis: Case report and literature review," Taiwanese Journal of Obstetrics and Gynecology. 2015; 54(1): 62-65.
9. J. Hoffman-Tretin, M. Koenigsberg, and M. Ziprkowski, "Antenatal demonstration of axillary cystic hygroma," Journal of Ultrasound in Medicine. 1988; 7(4): 233-235.
10. J. G. Park, M.-C. Aubry, J. A. Godfrey, and D. E. Midthun, "Mediastinallymphangioma: Mayo Clinic experience of 25 cases," Mayo Clinic Proceedings. 2006; 81(9): 1197–1203.
11. C. Turner and S. Gross, "Treatment of recurrent suprahyoidcervicofaciallymphangioma with intravenous cyclophosphamide," American Journal of PediatricHematology/Oncology. 1994; 16(4): 325-328.
12. M. A. Reinhardt, S. C. Nelson, S. F. Sencer, B. C. Bostrom, S. C. Kurachek, and M. E. Nesbit, "Treatment of childhood lymphangiomas with interferon-α," Journal of PediatricHematology/Oncology. 1997; 19(3): 232-236.
13. B. P. Flanagan and E. B. Helwig, "Cutaneous Lymphangioma," Archives of Dermatology.v1977; 113(1): 24-30.