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RESEARCH ARTICLE

CONGENITAL CYSTIC LYMPHANGIOMA: ANUNUSUAL PRESENTATION

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ABSTRACT

Lymphangioma is a condition of congenitallymphatic tissue malformation presenting as benign, hamartomatous growth. Lymphangiomas can occur in the skin and mucous membranes in any part of the body. It is diagnosed clinically by transillumination test positivity and radioimaging. Treatment is surgical excision or sclerotherapy or combination of both. Here we report a case of giant cystic lymhangioma involving both side of the trunk symmetrically.

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INTRODUCTION

Cystic lymphangioma is a congenital malformation ,resulting from failure of a primary lymphatic sac to establish drainage into the venous system (Fonnkaisurd, 1986) . The incidence of the disease is approximately 1 in 12,000 births and are the second most common benign vascular tumors in children (Faul *et al*., 2000). Most are located in head and neck region. There is no gender predisposition. Several treatments of cystic lymphangioma have been reported (Cynthia E Herzog, 2015). We presenthere a rare patient with lymphangioma involving both side of the chest and abdominal wall.

Case report: Aterm, 3.7 kg baby boy was born of a non-consanguineous parentage by caesarian section following an uncomplicated pregnancy of a mother of 20 years age. The baby breathed spontaneously after birth. The baby was born with bilateral symmetrical swelling at both side of trunk. (Fig 1). Antenatal history revealed that mother was a booked case, taken iron and folic acid tablet regularly, no history of teratogenicity or radiation exposure. No specific history suggestive of congenital infection was there. Antenatal USG at 30th week of gestation showed bilateral loculatedcystic mass of the chest and abdominal wall. On examination, there was a unusual symmetrical swelling involving both sides of chest and abdomen extending from axilla above to groin below. Swelling was soft cystic in nature without any tenderness. Trans-illumination test was positive (Fig 2).Other systems were normal on examination. USG of the swelling showed cystic anechoic swelling outside the bony rib cage, with multi-septation and echogenic debris inside- suggestive of Cystic Lymphangioma (Fig 3). Underlying lung tissue and pleural space was normal. Chest X-ray showed bilateral clear lung fields. USG abdomen & Echocardiography was also normal. Aspiration from cystic swelling showed, straw colored fluid with485/mm^3 cells, mostly are lymphocytes (96%). Protein was 70 mg/dl with increased cholesterol- all these are suggestive of lymphatic origin of the swelling. Umbilical cord blood taken at the time of delivery showed normal karyotype (46 XY). Breast feeding was started within few hours after birth. Percutaneous sclerotherapy followed by Excision of the swelling was planned and baby was discharged on day 4 of life in stable condition with advice to follow up after 14 days. But unfortunately the baby was brought to hospital by parents on day 12 after discharge with clinical features of septicemia. Swelling was increased in size, became tense and tender, hemorrhage and necrosisoccurred of overlying skin with increased local temperature. Despite immediate intensive supportive care and I.V antibiotics, we lose the baby within 12 hours due to uncorrectable shock.

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DISCUSSION

Congenital lymphangioma, alymphatic malformations, occur when developing lymphatic tissue fails to anastomose or

improperly anastomose with capillaries, veins, and arteries after the 6th week of gestation. Although 75% of lymphatic malformations are found in the head and neck region, it can occur anywhere (Chervenak *et al*., 1983).

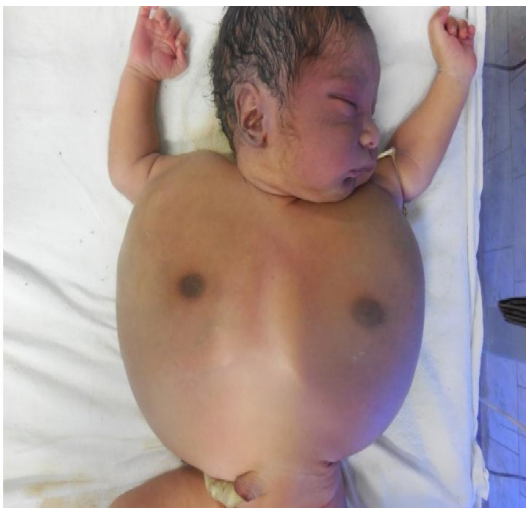

Fig 1. Boy with bilateral symmetrical swelling involving both side of trunk. Swelling was soft, cystic in nature

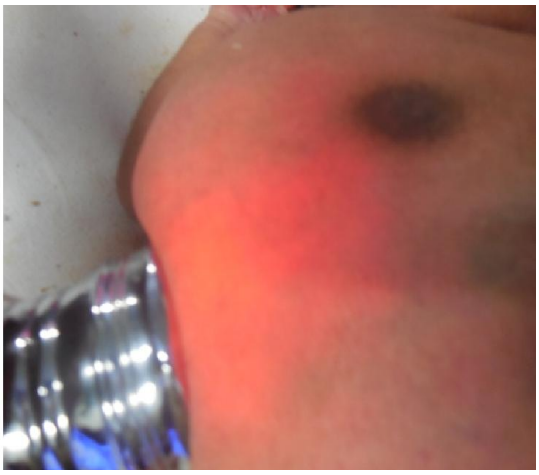

Fig. 2. Transillumination test positive

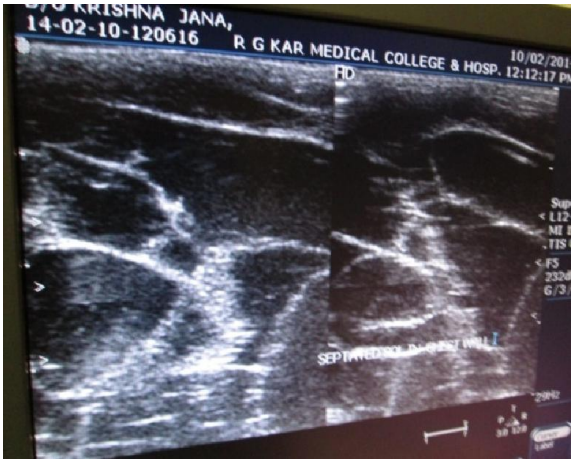

Fig. 3. USG of the swelling showed, cystic anechoic lesion with multi-septation and echogenic debris inside

Amongst them, chest wall lymphangioma is a very rare condition, only a few case report are there in literature. After a thorough search we do not find any case of bilateral symmetrical lymphangioma involving both chest and abdominal wall (Ardenghy *et al*., 1996) . Lymphangiomas are subdivided into three pathologic categories: capillary lymphangioma (lymphangioma simplex), cavernous lymphangiomas (microcystic) and cystic lymphangiomas (cystic hygromas) (Macro-cystic) (Faul *et al*., 2000; Parakh *et al*., 2002). Among these three subtype this case is of cysticlymphangiomas which are large, well-circumscribed, loculated, lymph fluid-filled spaces occur in areas where expansion is possible, as in our case. These lesions are apparent in 50–70% children at birth or prenatally, and most presenting by 2 years of age (Cynthia E Herzog, 2015). Though cystic hygromas have been found to be associated with chromosomal abnormalities such as Turner syndrome and Down syndrome (Gallagher *et al*., 1999), the chest wall lymphangioma are usually not associated with other chromosomal or structural anomalies. Its outcome is relatively favourable (Goldstein *et al*., 2006) .

Diagnosis is principally made on the basis of clinical appearance and imaging. Aspirated cystic fluid material shows, proteinaceous fluid with few lymphocytes. Ultrasound imaging is particularly useful during the perinatal/neonatal period (Davies *et al*., 2000)**.** MRI isnow considered to be the most accurate imaging modality for evaluation specially to distinguish lymphangiomas and other vascular malformations. Cystic hygromas identified in a fetus are especially concerning. The fetus is assessed for additional abnormalities that would increase the risk of fetal death or poor postpartum prognosis such as chromosomal abnormalities, hydropsfetalis, and large cyst volumes. When diagnosed prenatally, the overall prognosis is poor (Nadel *et al*., 1993).

Postnatally, surgical excision is effective, particularly in macrocystic malformations, and can be combined with sclerotherapy. But complete excision may prove technically difficult, because lymphangiomas may surround large blood vessels, airways, and mediastinal organs (Cynthia E Herzog, 2015). Treatment by injection of sclerosing agents, laser therapy and systemic interferon has also been used. Bleomycin, OK-432 (picibanil) may be used as sclerosing agent (Okazaki *et al*., 2007). Incomplete resection, or sclerosis, can result in recurrence of the lymphangioma and a return of symptoms. Before surgical exploration and excision, it is prudent to investigate for other lymphangiomatous lesions and associated congenital anomalies.

Conclusion

Congenital lymphangioma though mostly found in head and neck region, they may present as bilateral symmetrical swelling over both side of the trunk. Simple transillumination test can differentiate it from other vascular malformation.

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