Case Report:

Giant lymphatic malformation of the trunk with hemorrhage.

SN Singh, Mohammad Tahazzul, Anita Singh, RK Chaudhary

Department of Pediatrics, Chhatrapati Shahuji Maharaj Medical University, Lucknow-226003, Uttar Pradesh.

Abstract

Lymphatic malformation involving trunk is rare, commomnly being in neck and axillary region. A neonate with giant cystic swelling on left side of trunk since birth with overlying echymosis & abrasion, and severe pallor is presented. Aspiration and sonography suggested diagnosis of cystic hygroma with intralesional hemorrhage. Blood transfusion and sclerotherapy was done followed by surgical excision, but the baby died of sepsis postoperatively.

Keywords: Lymphatic Malformation (LM); Trunk; Hemorrhage

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Introduction

Lymphomatous malformations (LM) are congenital lesions arising from sequestration of lymphatic tissue from the embryonic lymphatic sac [1]. These sequestrated tissues fail to communicate with remainder of the lymphatic or venous system. Later on, dilatation of the sequestrated lymphatic tissue ensues resulting in the cystic morphology of the lesion [2]. Cervical and axillary localization is most frequent [3]. A Doppler USG should be done to detect any vascular compartment of the L.M. The common complication of L.M. includes respiratory obstruction, infection, ulcerated lesions and intracystic hemorrhage. A complete surgical excision of the cyst is preferred whenever possible. Intralesional injection of various sclerozing agents i.e. ok-432 (a killed lyophilized strain of group A streptococcous), 100% ethanol, bleomycin or picibanil may be useful for the involution of giant LM [4,5]. Because of rarity of such huge size chest wall LM with hemorrhage, this case is being reported.

Case Report

A term male neonate, delivered at home presented on day 2 of life with massive cystic swelling covering left side of chest & abdomen up to groin & extending upto the right half of the trunk (Fig. 1). The skin over the swelling had large area of ecchymosis & abrasion at places. The baby had severe pallor and required blood transfusion. The transillumination test was negative. X-ray chest & abdomen showed huge rounded uniform opaque shadow overlapping left hemithorax and abdomen and extending laterally (Fig. 2). On sonography, the lesion showed fluid attenuation with echogenic shadows and was poorly

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Figure 1. Giant cystic swelling of the left side of trunk



Figure 2. X-Ray Chest P.A. view

marginated, suggesting cystic hygroma. There was no flow on Doppler study. Aspirate from the cyst was uniformly hemorrhagic indicating intralesional bleeding. This appearance was most consistent with a diagnosis of giant lymphatic malformation of the trunk. Needle aspiration was done and injection bleomycin was instilled but there was no reduction in size of swelling. Surgical resection of cyst was done at age of 18 days. Postoperatively the baby required ventilatory support but eventually baby died of sepsis.

Discussion

Lymphatic malformation (L.M) is a sponge like collection of abnormal channels and cystic spaces that contain clear fluid. They are usually classified as capillary, cavernous or cystic lymphangioma (cystic hygroma). Most of the LM are apparent at birth as cystic translucent mass, however translucency may get altered when there is infection or hemorrhage. It can manifest anywhere in the body. The common locations are cervico-facial region, axilla, mediastinum, groin and below tongue [3]. Prenatal diagnosis of cystic hygroma using sonography is possible [6]; however, it was missed in this case. The foetus with cystic hygroma can be associated with other anomalies in 62% cases [3]. On sonography, LM generally appears as anechoic cystic mass, but occasionally debris may result in echogenisity [7]. A Doppler study should be done to detect any vascular compartment of the LM. CT or MRI scan may further aid in the identification of the lesion and its extension and are particularly useful, when surgical management of the lesion is contemplated. Some cystic hygromas contains nests of poorly supported vascular channels that are prone to bleeding. The common complication of LM includes respiratory obstruction, infection, ulceration and intracystic hemorrhage [3,8]. Trauma during birth due to huge size of the cyst in present case resulted in ecchymosis and ulceration of overlying skin. Sudden increase in size of the cyst, which often becomes tense, indicates hemorrhage or infection. Aspiration can provide temporary relief in an emergency, such as in the presence of dyspnea, but reaccumulation occurs. A complete surgical excision of the cyst is preferred, however, because of their tendency to infiltrate and surround adjacent tissues and major structures like nerves and vessels radical surgical resection becomes difficult. Recurrence is common with incompletely resected lesions. Because endothelial lining of the cyst is vulnerable to chemical irritant, various sclerosing agents have been tried to treat cystic hygroma. [4,5]. Intralesional injection of bleomycin however, failed to reduce the size in our case.

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Correspondence to:

SN Singh Department of Pediatrics Chhatrapati Shahuji Maharaj Medical University Lucknow-226003, Uttar Pradesh India Phone: +9415202124 Email: drsn.singh@rediffmail.com