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NONIMMUNE HYDROPS FETALIS AND BILATERAL PULMONARY HYPOPLASIA IN A NEWBORN INFANT WITH NUCHAL VASCULAR HAMARTOMA*

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Nuchal vascular hamartoma was found in a newborn premature infant who presented with nonimmune hydrops fetalis, pulmonary hypoplasia due to bilateral pleural effusion and polyhydramnios in utero. The baby died 26 hours after birth despite maximal respiratory and circulatory support. Postmortem examination revealed a vascular hamartoma localized to the left posterolateral region of the neck. We suggest that nuchal vascular hamartoma may be associated with fetal hydrops, probably due to compromised lymph drainage. *Key words: vascular hamartoma, nonimmune hydrops fetalis, pulmonary hypoplasia.*

Nonimmune hydrops fetalis is a relatively rare and complex disorder that requires detailed investigation. A wide range of associations have been reported, but chromosomal abnormalities, cardiac anomalies, pulmonary abnormalities, infection and multiple births are the common causes of the disorder[1,2]. We describe a newborn infant with nuchal vascular hamartoma, who presented with nonimmune hydrops fetalis, bilateral pleural effusion, pulmonary hypoplasia and polyhydramnios in utero. The association between nuchal vascular hamartoma and pleural effusion and pathogenetic mechanisms leading to fetal hydrops are discussed.

Case Report

A 3400 g female infant was born to a 28-year-old multigravida at 36 weeks gestation by cesarean section. Pregnancy was complicated by polyhydramnios; bilateral pleural effusion and ascites were noted during ultrasound examination

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at 33 weeks. Because of hydrops fetalis, a cordocentesis was performed for fetal karyotyping. The fetal blood hemoglobin was 17 g/dl. At birth, the baby was severely asphyxiated and showed nuchal mass and cervical edema, especially in the left posterolateral region of the neck (Fig. 1). She was immediately intubated and mechanically ventilated. Initial blood gases revealed severe respiratory acidosis. Chest x-ray showed bilateral pleural fluid, especially in the left hemithorax, and relatively good aeration of the right hemithorax (Fig. 2). Bilateral thoracocentesis was performed and a sero-hemorrhagic pleural fluid was obtained. Cell count of the pleural fluid revealed 820 leukocytes/ml, with a predominance of lymphocytes. The protein and glucose contents were 3.1 g/dl and 112 mg/dl, respectively. Triglyceride and cholesterol contents were 75 mg/dl and 57 mg/dl, respectively. Lactic dehydrogenase (LDH) content was 750 IU/L. Cultures of pleural fluid were negative as were serologic tests for toxoplasmosis, rubella, cytomegalovirus, and herpes simplex virus infections. Other laboratory investigations revealed normal urinalysis, complete blood count, blood urea nitrogen, creatinine, glucose, liver functions, electrolytes, total protein (4.1 g/dl), albumin (2.6 g/dl), cholesterol (58 mg/dl), triglyceride (24 mg/dl), and lactic dehydrogenase (1542 IU/L) levels. Chromosomal investigation revealed a normal karyotype. Despite maximal respiratory and circulatory support, severe hypoxemia and acidosis persisted, and the baby died 26 hours after birth.

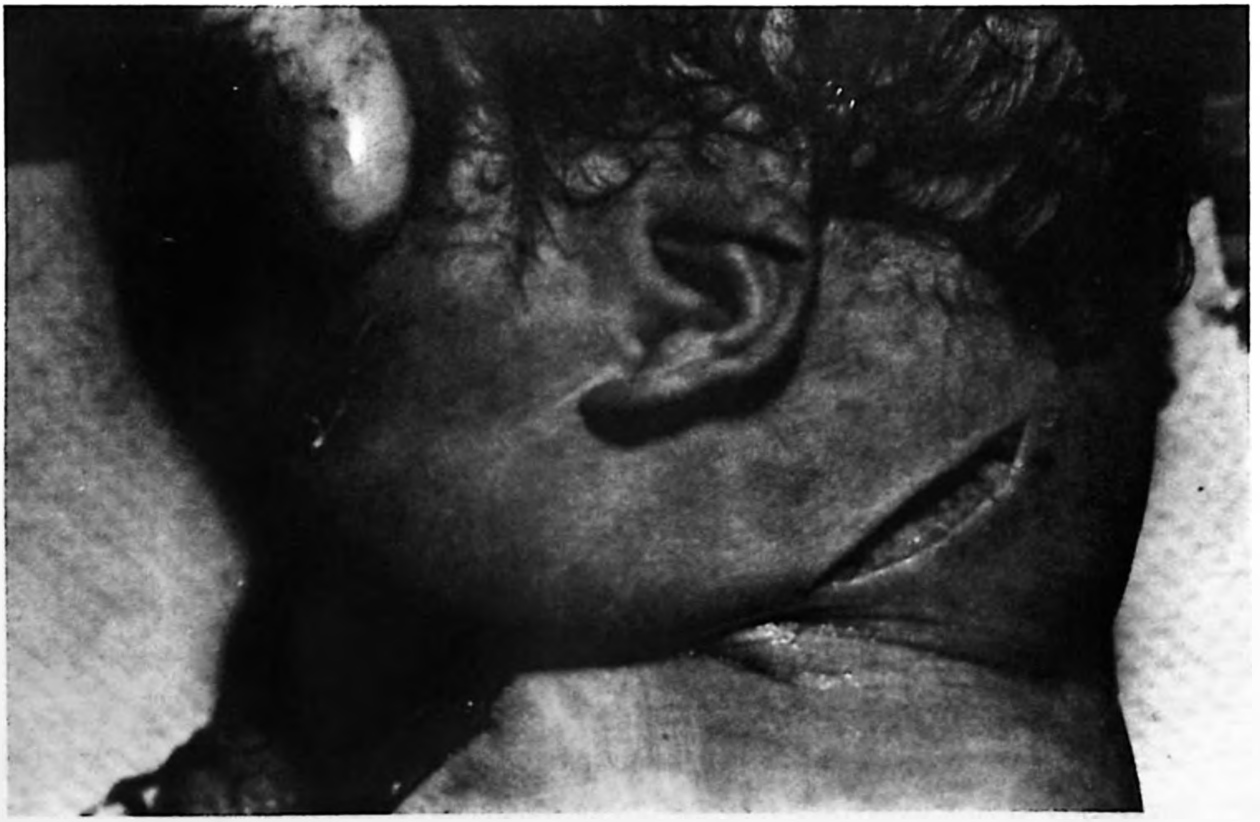

Fig. 1: Nuchal mass and cervical edema in the left posterolateral region (postmortem view).

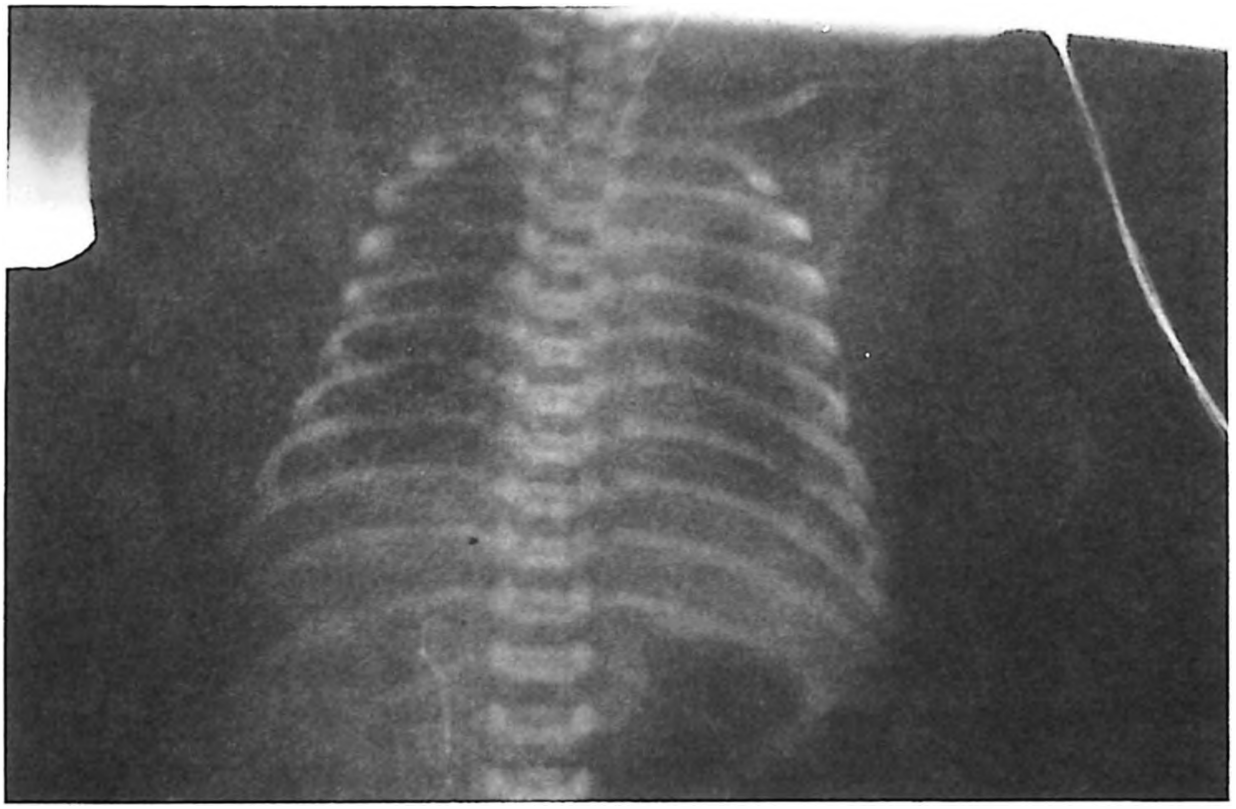

Fig. 2: Chest x-ray two hours after birth. Bilateral pleural fluid, especially in left hemithorax, and relatively good aeration of the right lung.

Upon postmortem examination, 25 ml sero-hemorrhagic pleural fluid was found. Both left and right lungs were hypoplastic, weighing 15 g each. In addition, external examination revealed a left cervical subcutaneous mass, 3.5 x 3 x 3 cm and firm, that was well-circumscribed and partially congested. Histologically, the lesion consisted of many vascular spaces lined by endothelial cells (Fig. 3). The lumen of some spaces contained red blood cells. A collection of lymphoid tissue was seen in the interstitial area. The histopathological diagnosis was vascular hamartoma (hemangio-lymphangioma).

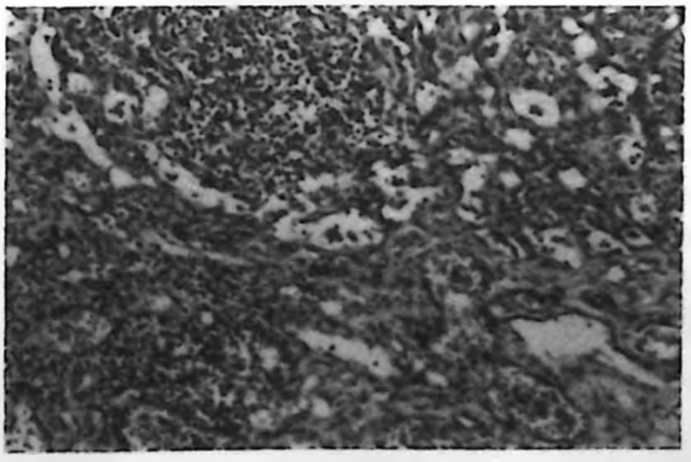

Fig. 3: Microscopic appearance of the nuchal mass. The lesion was composed of vascular spaces with a lymphoid stroma (x 100).

Discussion

Vascular hamartomas in the head and neck region are very unusual in infancy and childhood[3,6]. This case is unique because nuchal vascular hamartoma is associated with bilateral pleural effusion and hydrops fetalis. No report of this association was found in international literature for the past 20 years. There may be a coincidental association or it may be the cause of the hydrops.

The lymphatic channels in each uper quadrant drain into the jugular lymphatic sac. These jugular sacs eventually form communications with the venous system. We suggest that this communication and lymph drainage may be compromised by nuchal hemangio-lymphangioma in this case. Fetal pleural effusion and progression to hydrops is probably due to disturbed lymph drainage[2,7]. Based on the composition of pleural fluid before feeding, congenital chylothorax was considered to be the most likely diagnosis in our patient[8,9]. Pleural effusion may have caused pulmonary hypoplasia in our patient because significant pleural effusions before the alveolar phase of lung development irreversibly damage fetal lungs[10].

This report suggests that vascular hamartoma may be one of the many causes of non-immune hydrops fetalis.

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