

# Rare lymphatic malformation in an extreme premature infant: answer

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Lymphatic malformation, cystic lymphangioma, extreme prematurity, congenital mass.

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## Answers

1. Lymphatic malformation: cystic lymphangioma.
2. I. Chest X-ray to rule out possible lymphatic malformation in thorax.  
II. Sonographic study of abdomen to exclude possible involvement of liver, spleen, kidney, and intestine.

III. Genetic screening to rule out possible association with trisomies 13, 18, and 21, Turner syndrome, or Noonan syndrome.

3. General surgery, genetics.

## Introduction

Lymphatic malformations are benign, congenital proliferations of lymphatic tissue.

They can present anywhere in the body, but most commonly (> 95%) can be found in cervico-facial and upper torso regions [1, 2].

These malformations may be associated with trisomies 13, 18, and 21, Turner and Noonan syndromes, hydrops, or other structural anomalies [3]. Up to 70% of these lesions are present at birth, and there is a mild male predominance among reported cases [4].

## Clinical course

We present a case of massive cystic lymphangioma in an extreme preterm infant, which was initially diagnosed by prenatal ultrasound and confirmed by fetal MRI. Physical examination and postnatal ultrasound confirmed diagnosis of cystic lymphangioma. No other anomalies were detected, genetic screening for infant was normal. Despite of size of lymphatic malformation at birth, it was an isolated finding with minimal vascularity, thus the prognosis for self-resolution was very good.

No surgical intervention was required other than skin care. The lesion started to decrease in size from day 5, and showed signs of gradual involution from the 2<sup>nd</sup> week of life (**Fig. 1**).

At 37 weeks of corrected gestational age (GA) lymphatic malformation underwent full resolution, with minimal pigmentation and scarring on the site of lesion (**Fig. 2**).

## Discussion

Lymphatic malformations are hamartomatous lesions that are usually presented and diagnosed at birth. In the last few decades more cases of lymphangiomas are diagnosed by prenatal sonography. These lesions are rare – incidence of lymphatic malformations reported to be between 1:4,000 and 1:6,500 [1, 2, 5]. Origin and pathogenesis of these lesions are not fully understood, and are explained by anomalous sequestrations of lymphatic tissues and vessels during embryogenesis. These lymphatic mal-



**Figure 1.** Involution of mass on day of life 15.



**Figure 2.** Picture taken at the age of 4 months, showing minimal pigmentation and scarring.

formations are known to be associated with other anomalies and genetic syndromes [2, 5].

Lymphatic malformations are clinically divided into capillary lymphangioma, cavernous lymphangioma and cystic lymphangioma. Usually capillary lesions are small, and cystic lymphangiomas are huge in size. Structurally cystic lymphangiomas could be divided into microcystic (smaller than 2 cm), macrocystic (larger than 2 cm) and mixed (multiple cysts separated by septa) [6]. Most of these lesions are benign, and do not create clinical concerns unless they compress vital organs and cause life-threatening complications. Large cystic lymphangiomas are at risk of rupture and hemorrhage. Another common complication of these lesions is a secondary infection.

Our patient was diagnosed prenatally with lymphatic malformation, and despite the extreme prematurity and large size of lesion he had no

clinical issues caused or aggravated by this congenital mass. He was thoroughly assessed after delivery, and no associated anomalies or dysmorphic features were found. His chest X-ray did not reveal any unusual masses in the thorax; cranial and abdominal ultrasounds were normal as well. His karyotype was 46XY, and his microarray was reported as normal.

In available literature, spontaneous resolution of similar lesions was reported as uncommon [1, 2, 5-7]. Most of the authors identified leakage from the lesion or infection as the most common indications for surgical excision. Being born as extreme preterm carries additional risks for infection: skin of infant born at 26 weeks of GA has undeveloped structure (immature stratum corneum) and insufficient barrier function [8]. We were fortunate not to have these complications, which contributed towards spontaneous resolution of cystic lymphangioma in our patient, with first signs of improvement noted in the 2<sup>nd</sup> week of life and full recovery at 37 weeks of corrected GA .

#### Declaration of interest

The Authors declare that there is no conflict of interest.

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