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MSD Global Journal of Clinical and Medical Case Reports

Case Report

Received: 09 October, 2024
Accepted: 21 October, 2024
Published: 22 October, 2024

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Citation: Livrinova V, Komina S, Kostadinova KS, Aneta S, Aliji N, Veljanoski D (2024) Congenital Pulmonary Airway Malformation Type II and Challenge in the decision-making process in developing countries-a case report. MSD Glob J of Clin med Case Rep. 2(1): 003-005.
DOI: 10.37179/msdgjcmcr.000004

Congenital Pulmonary Airway Malformation Type II and Challenge in the decision-making process in developing countries-a case report

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Abstract

Congenital Pulmonary Airway Malformation (CPAM) is a congenital lung disease that can be present from prenatal to childhood. The overall incidence of CPAM is reported to be 1 in 10,000 to 1 in 35,000 births, making it the most common type of congenital lung lesion. This malformation results from the interruption of lung development during various stages of embryogenesis. This case report aims to illustrate the disease through ultrasound imaging, emphasize its characteristics, and discuss parental counseling and decision-making regarding the continuation of pregnancy. It is important to highlight the limitations of prenatal diagnostic precision in developing countries and the need for additional diagnostic tools to improve parental decision-making options.

Case Report

Case Presentation: A 25-year-old multiparous woman (G2.P1) presented to the clinic with her second pregnancy. She had undergone three antenatal checkups before being referred to our clinic for second-trimester screening for congenital abnormalities, with her previous checkups indicating a normal pregnancy. She was referred to at 22 weeks of gestation with a singleton pregnancy confirmed by abdominal ultrasound examination. The ultrasound revealed a single fetus with normal fetal movements and biometrics corresponding to the gestational age (23 weeks). The amniotic fluid level was normal, and the placenta had a normal appearance and insertion. No structural defects were detected in the kidneys or other organs.

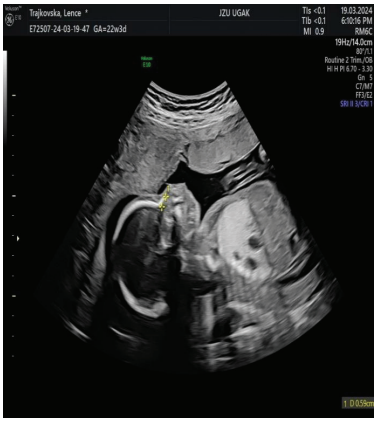
The ultrasound showed normal heart structures, but a mediastinal shift to the right side of the chest was observed, caused by several cystic changes

in the lung tissue on the left side. The largest cystic formation measured 12x10 mm. In the transverse section of the chest, an additional three cystic formations were visible, with a total of approximately five cystic lesions (Pictures 1 and 2). Color Doppler mapping of the lungs was performed (Picture 3). The Congenital Pulmonary Airway Malformation Volume Ratio (CVR) was calculated to be 1.2 cm^2.

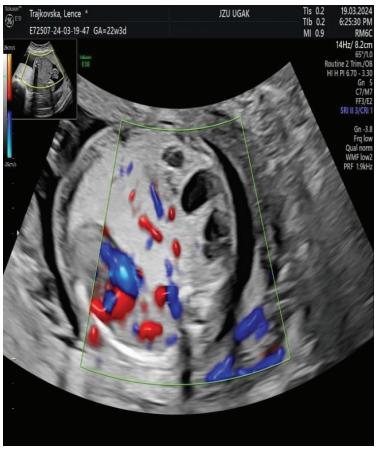
Intervention and outcome: The limitations and diagnostic capacity of the ultrasound examination were explained to the patient. We discussed the possibility of expectant monitoring of the pregnancy and the risk of intrauterine fetal death (heart failure). The patient and her husband declined fetal MRI. Due to a lack of resources, genetic testing for the HOXB5 gene was unavailable. They opted to terminate the pregnancy. A histopathological examination of the fetus revealed the following results, as shown in (Pictures 4 and 5).

Histopathological Diagnosis: Abortus inductus indicatus propter malformationem foeti (generis masculini, ponderis 600g, longitudinis 31 cm) Malformatio Congenita Viae Aeriae Pulmonis typus secundum (congenital pulmonary airway malformation type II). Immaturitas organorum internorum, stasis universalis. haemathoma retroplacentare.

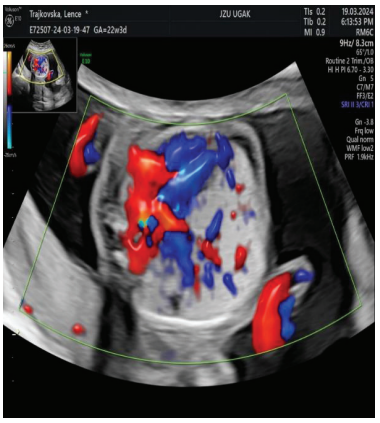
Conclusion: Data from large population registries suggest an incidence of congenital lung cysts ranging from



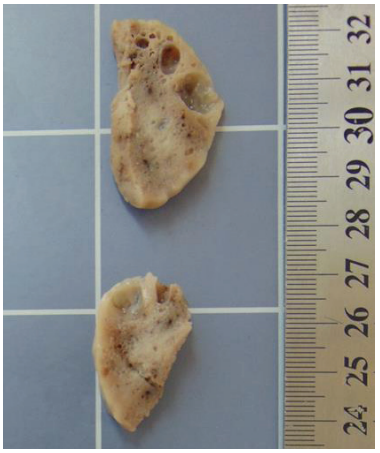
Picture 1: 2D US-sagittal plane od fetal lung.



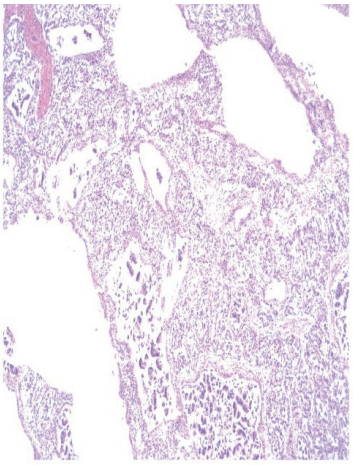
Picture 2: Color doppler-transverse view -thorax.



Picture 3: Color Doppler – fetal hearth with mediastinal shift on the right.



Picture 4: Macroscopic appearance.



Picture 5: Microscopic appearance of lung tissue of both fetal lungs (up-left, down-right).

1 in 8,300 to 1 in 35,000 live births. Large-cyst subtypes account for approximately 70% of CPAM cases or 2 to 8 per 100,000 live births. Type II lesions comprise 15 to 20% of CPAM cases. Although lesion puncture during pregnancy and postnatal surgical excision are recommended treatments, in developing countries, parents may, unfortunately, choose to terminate the pregnancy due to limited diagnostic and treatment options [1-6].

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