

CONGENITAL LOBAR EMPHYSEMA (CLE) RADIOLOGIC AND IMAGISTIC (CT) DIAGNOSTIC

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Abstract

A new born child, age: one month, sex: masculin, with acute respiratory distress syndrome, perioronasal cyanosis and hypoxemia has been diagnosed (radiological and imagical – CT) with congenital lobar emphysema (CLE) at the level of the right superior lobe.

One month after, the new born child has been diagnosed with the aforesaid syndrome, and, now two months old, he has been subjected to a surgical intervention consisting in lobectomy at the level of the right superior lobe.

His postsurgical evolution has proved satisfactory with pulmonary reexpansion.

Key words: congenital pathology, pneumopathology, congenital lobar emphysema

Introduction

Congenital malformations of the lung are rare and vary widely in their presentation and severity(4).

Congenital lobar Emphysema (CLE) is infrequent and usually present at birth(8).

The superior lobes are affected (5) and a single lobe usually is involved; however, patients can show multiple lobar involvement (8). The rates of occurrence are: left upper lobe – 41%, right middle lobe – 34%; right upper lobe - 21%(5,8).

Frequently, cartilage plates in the bronchi are absent at the level where cartilage is expected (5,8). The abnormality is related to intrinsic bronchial narrowing. In these cases, there is weakened or absent bronchial cartilage, so that there is inspiratory air entry but collapse of the narrow bronchial lumen during expiration. This bronchial defect result in lobar air trapping (7,8).

Pulmonary arteries are normal in patients with CLE (8). Angiography shows slow and poor arterial filling (5).

Hypoxemia (in severely affected patients) may occur (8).

CASE PRESENTATION

Anamnesis and clinic findings:

1 (one) month new – born child, male has been brought to Louis Turcanu Children Emergency Hospital from Timisoara on April 21, 2009 and has been hospitalized in the Pediatric Department.

Upon hospitalization, he presented a severe general state, perioronasal cyanosis, aerated secretions at the mouth level, mixed dispnea, continuous exhausting cough and disseminated subcrepitan rhonchi.

Paraclinic findings:

- Gases into the blood: ph=7,431; pCO₂ (carbon dioxide pressure) = 44,2mmHg; reduced pO₂ (oxygen pressure) = 38,1mmHg (normal: pO₂ = 75,0 – 100,0mmHg).
- Blood exam: HGB = 11,5g/dL (normal: 12,0 – 16,5g/dL), lymphocytes (11,25x10³/uL), monocytes (1,87x10³/uL).
- Cardiac echographic examination (conclusion): normal structured cord, moved to the left and down side, inclusive the aortic arch is moved 2 (two) inter – rib space down.
- ECG: RS – regulated, 160 beats per minute, QRS + 60⁰ axis, left branch minor block, the rest presents a normal electric path.

Radiological and imagical aspects:

Standard Radiography:

Marked hyperaeration and overdistention of the right upper lobe with mediastinal shift to the left. Aer ation and left lung reduction (hypoventilated by means of compression) (Fig. 1, Fig. 2).

Computer Tomography:

Pronounced distention of the right superior lobe with lobar hypovascularisation in comparison with the left superior lobe. The hyperinflated LSD directs the medistine structures towards the left side, having a compressive effect on the LMD and LIED (Fig. 3, Fig. 4).

Virtual Bronchoscopy:

At the level of the right superior lobar bronchus, an anterior and inferior oriented tract can be noticed, with a permeable lumen extending up to the bifurcation level, where, due to the extremely reduced dimensions of the segmentary bronchi diameter, the endobronchial navigation can no longer continue. The diameter of the right superior lobar bronchus has diminished in comparison with the left superior lobar bronchus (1,4 mm at emergency, in comparison with a 1,8 mm diameter).



Fig. 1 Postero-anterior view.
 Marked hyperaeration and overdistention
 and mediastinal shift to the left.



Fig. 2 Lateral view. Marked
 hyperaeration and overdistention of the
 retrosternal space.

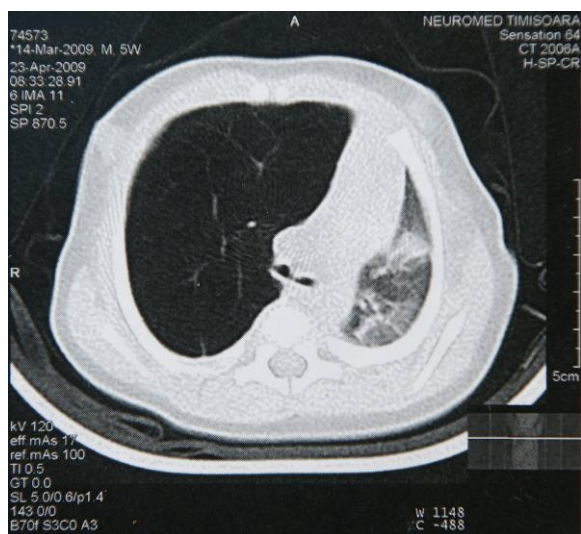


Fig. 3 Uper thorax section. Marked
 hyperaeration and overdistention of the right
 superior pulmonary lobe with mediastinal shift
 and compression with hypoventilation of the
 left superior pulmonary lobe.

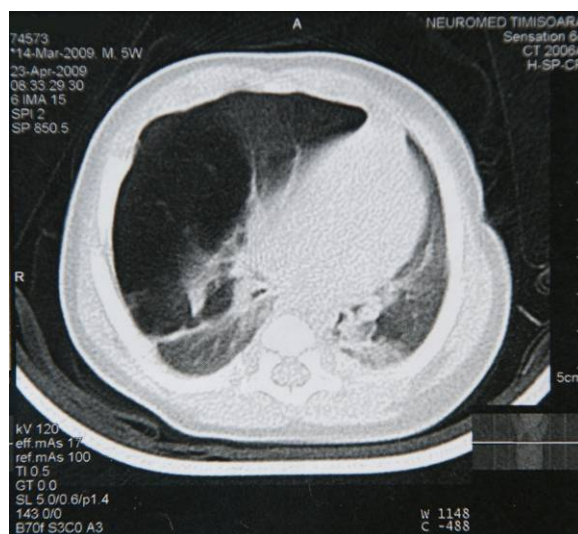


Fig.4 Midle thorax. Marked hyperaeration and
 overdistention of the right superior pulmonary
 lobe with mediastinal shift and compression
 with hypoventilation of the left superior
 pulmonary lobe.

Discussions

The congenital lobar emphysema can be detected even from the prenatal period in the presence of a hyperechogenic pulmonary zone (2). The echographic semiology is not very clear, and that's why, we shall resort to other medical imaging techniques, such as prenatal nuclear MRI and postnatal tomodensitometry (CT)(2).

As regards our medical investigation, the diagnosis established by us has been revealed by the postnatal tomodensitometry (CT).

The symptomatic form of the congenital lobar emphysema (CLE) must be immediately detected and operated because the subsequent diagnosis shall be conditioned by the patient's age at the moment when the surgical intervention is performed (3).

The surgical intervention can be avoided only in case of incipient forms of the syndrome, with negligible distension and without apparent symptomatology (3,6).

Unfortunately, the patient into question has been diagnosed with a quite serious distension and apparent symptomatology so, in this case, the surgical intervention

was absolutely compulsory. The surgery (right superior lobectomy) has been performed on May 18, 2009, more specifically, one month after the diagnosis has been established. The postsurgical evolution has proved satisfactory with the reexpansion of the left pulmonary and the segments left from the left lung (Fig. 5).



Fig. 5 Postoperative aspect. Re-expansion of the middle and inferior lobe of the right lung and re-expansion of the left lung.

As regard to the differential diagnosis (1) analysed by us after the standard cardiopulmonary radiography has been performed, the possibility of the left pulmonary hypoplasia with right pulmonary hyperinflation has been brought into discussion and analyzed.

Conclusions

1. The cardiopulmonary radiologic exam (standard radiography: front and profile) and the imagistic exam (CT or MRI) represents diagnosis elements in the cases of congenital lobar emphysema (CLE).
2. The surgery (lobectomy) represents the treatment indicated by the apparent symptomatic forms of the congenital lobar emphysema (CLE).

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