Secondary diaphragmatic eventration after resection of extralobar pulmonary sequestration

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ABSTRACT
Phrenic nerve preservation is an important factor in operations involving the resection of an extralobar pulmonary sequestration. We present a case of secondary diaphragmatic eventration due to damage to the phrenic nerve after resection of an extralobar pulmonary sequestration.

KEY WORDS: Diaphragmatic eventration, extralobar pulmonary sequestration, plication

INTRODUCTION
Pulmonary sequestration covers 10-30% of all cystic congenital malformations of the lung.[1,2] It is classified as intralobar or extralobar according to whether the sequestration resides inside the normal visceral pleura of the lung or if it is covered by its own visceral pleura respectively. In both types, no communication between the lesion and the tracheobronchial tree exists and its blood supply comes from aberrant systemic arterial vessels.

CASE REPORT
Our patient was the first child of two healthy parents. No medication was taken by the mother during pregnancy. A prenatal ultrasound (U/S) examination revealed the extralobar pulmonary sequestration of the left lung and its blood supply from the descending aorta. The pregnancy was concluded without any problems and the child was born at a gestational age of 37 weeks with a birth body weight of 3.7 kg. On the second postnatal day, the infant showed clinical signs of respiratory distress and was transferred to the neonatal intensive care unit (NICU) for stabilization and further investigation. The diagnosis was set and confirmed with plain chest radiographs that showed a sequestrum in the lower lobe of the left lung just above the diaphragm.

No other systemic abnormalities were observed. Preoperative evaluation was conducted with U/S and Doppler examination and magnetic resonance imaging (MRI). The patient was operated for resection of the lesion when he reached the age of 10 days old at a private pediatric cardiothoracic clinic. Complete resection of the lesion was successful. However, the operation was described by the surgeons to be difficult and not uneventful as careful dissection of the left phrenic nerve was needed and performed. After surgery, the infant was sent to the NICU for careful monitoring.

On the second postoperative day, a routine plain chest radiograph revealed elevation of the left hemi-diaphragm which led to the suspicion that there was paresis of the left diaphragm [Figure 1]. On the 10th postoperative day, a second radiograph was performed with a contrast...
medium but there was no change in the findings and the diagnosis of diaphragm eventration was confirmed [Figure 2]. The patient was observed for the following six weeks. All appropriate examinations for evaluation of the condition did not show any improvement. Consequently, the patient was reoperated on 1.5 months after the first operation for left diaphragm plication. The operation was successful and the patient was released from the hospital ten days later. After 12 months, he is doing very well and no operation-related symptoms have been reported.

**DISCUSSION**

Extralobar pulmonary sequestration is a rare congenital malformation of the lungs and is characterized by the fact that it is completely separated from the normal lung and covered by its own visceral pleura. These lesions have an incidence of approximately 90% on the left side with a 4:1 male to female ratio. There is no communication between these lesions and the bronchial tree. Their blood supply is usually by the descending aorta while their venous drainage is to the azygos or hemiazygos veins. An association as high as 30% between extralobar sequestration and CDH has been reported while there is a lower association between other conditions such as arterial-venous shunt and congestive heart failure and this lesion. Primary association with diaphragmatic eventration has also been reported.

Diagnosis of this malformation can be made in the prenatal period by U/S and Doppler imaging which demonstrates the lesion and its blood supply. After birth, due to the elevated possibility of association to other medical conditions, further imaging with plain chest X-rays, U/S, computed tomography (CT) or MRI is appropriate. Excision of the abnormal tissue is the indicated therapy. Although it may be asymptomatic, its relevance to infections, arterial-venous shunt and late malignancy are the main reasons that make surgical treatment necessary. Resection is performed via thoracotomy. The main principles of this operation are the identification and control of the blood supply as long as the phrenic nerve, usually adjacent to the lesion, is identified and carefully managed and preserved. Damage of the phrenic nerve can cause dramatic complications such as paresis or even paralysis of the diaphragm leading to secondary diaphragm eventration as it happened in our patient. If this is the case and no other problems from the prior operation are to be solved, the protocol for diaphragm eventration is followed. Observation and waiting for 4-6 weeks is recommended in order to allow the paresis of the phrenic nerve to be resolved. If there are any signs of improvement, further waiting will usually lead to full recovery. If no change is noted after the six week period, evaluation of the phrenic nerve is done by specific examination such as the application of faradic voltage and a negative result obliges the therapist to move to surgical treatment.

Diaphragm plication is the method of choice that is usually applied with extremely satisfactory results. Extralobar pulmonary sequestration, a rare congenital malformation of the lungs, can be prenatally diagnosed and treated surgically in the neonatal period with very good results. However, if great care is not taken during surgery for the identification, careful dissection, management and preservation of the phrenic nerve, secondary eventration of the diaphragm will complicate the outcome and a second operation will be needed for the patient to recover completely.

**REFERENCES**


**Figure 2:** X-ray prior to the release of the patient showing complete treatment

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