Acta Biomed 2016; Vol. 87, Supplement 3: 45-50

Review

Imaging of congenital pulmonary malformations

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Summary. Congenital pulmonary malformations represent a broad spectrum of anomalies that may result in varied clinical and pathologic pictures, ranging from recurrent pulmonary infections and acute respiratory distress syndrome, which require timely drug therapy, up to large space-occupying lesions needing surgical treatment. This classification includes three distinct anatomical and pathological entities, represented by Congenital Cystic Adenomatoid Malformation, Bronchopulmonary Sequestration and Congenital Lobar Emphysema. The final result in terms of embryological and fetal development of these alterations is a Congenital Lung Hypoplasia. Since even Bronchial Atresia, Pulmonary Bronchogenic Cysts and Congenital Diaphragmatic Hernias are due to Pulmonary Hypoplasia, these diseases will be discussed in this review (1, 2). (www.actabiomedica.it)

Key words: congenital, lung, malformation

Congenital cystic adenomatoid malformation

It is a congenital anomaly that occurs in 70% of cases in the first week of life, and only 10% are diagnosed after the first year of life. It consists of a mass of lung tissue, mainly derived from the bronchioles, disorganized and multicystic. It occurs more frequently in the upper lobes without, however, excluding the possible involvement of any lung segment; it is contiguous to the bronchial tree and it is supplied by a branch of the pulmonary artery. Cystic Adenomatoid Malformations are classically classified into three types, according to their characteristics and the type of cysts contained; type I, the most frequent, contains one or more cysts greater than 2 cm in diameter, air and sometimes an air-fluid level, and are able to occupy the entire hemithorax (1-11). Although it is sometimes possible a prenatal ultrasound diagnosis, in newborns XR the cystic adenomatoid malformations show themselves as space-occupying lesions, hypo or hyper-diaphanous, according to their content (Fig. 1-A). CT may identify the bronchial and arterial branches supplying the lesion (Fig. 1-B) (1, 6, 11-15).

Bronchopulmonary sequestration

It is a congenital anomaly characterized by a disorganized area of the lung parenchyma, devoid of pulmonary arteries and bronchial branches; however, the sequestration is supplied by branches of the thoracic or abdominal aorta. This malformation results from an abnormal development of the cephalic portion of the primitive gut. There are two forms of bronchopulmonary sequestration: intra-lobar and extra-lobar. In the intra-lobar form, the most common, the lung sequestered is located inside of the pleura of one of the lobes; it is more often found in the postero-basal segment of the left lower lobe and in most cases is vascularized by a branch of the descending thoracic aorta. According to symptoms, it is usually diagnosed

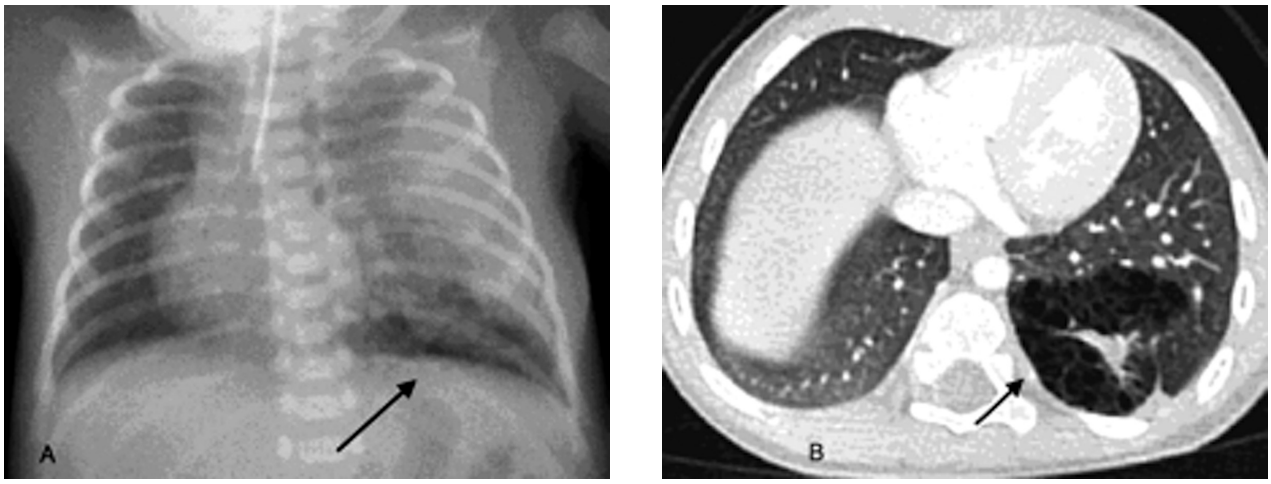

Figure 1. (A) XR shows accentuated left basal retrocardiac bronchovascular markings. (B) CT detects a malformation characterized by multiple parenchymal cysts with thin walls and thickened band of dysplastic parenchyma

in adulthood (1, 2, 7). At XR the intra-lobar sequestration appears as a space-occupying lesion with well defined margins or as a hyperdiaphanous formation, sometimes with air and septa inside (Fig. 2-A). With CT it is possible to observe it as a consolidation with its vascularization arising directly from the aorta or as a multicystic lesion with a mucous, fluid or air-fluid content (Fig. 2-B) (1, 7). The extra-lobar sequestration is instead surrounded by its own pleural coating, it is more frequent in the left pulmonary basis and it is replenished from a branch of the abdominal aorta. It is often accidentally diagnosed in childhood, frequently as a space-occupying lesion, rarely infected (1, 2, 7). At XR and CT examinations the extra-lobar sequestration appears as a well defined and homogeneous space-occupying lesion, which does not contain air, but may contain cystic areas. The CT scan also may identify the tributary arterial branch (1, 7).

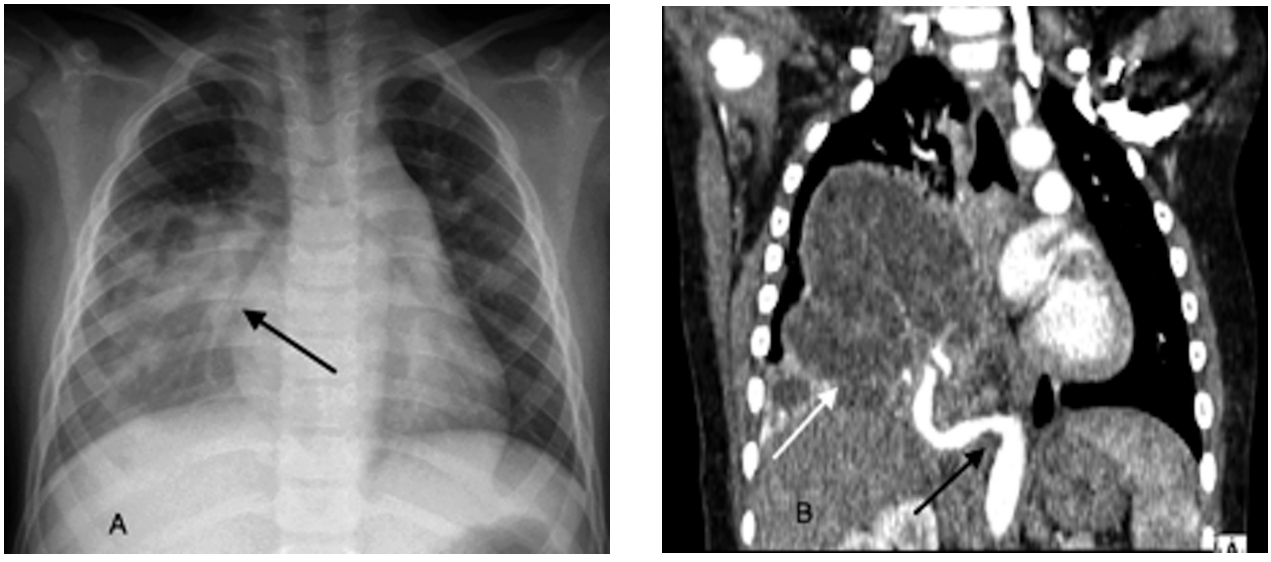

Figure 2. (A) XR shows parenchymal densification in the middle and lower right lung field. (B) CT displays a complex expansive formation characterized by multiple septa taking contrast (white arrow), associated to a dorso-basal area of pulmonary consolidation replenished by an abnormal blood vessel originating from the aorta near the thoraco-abdominal transition (black arrow)

Congenital lobar emphysema

In most cases this disease is recognized during the first month of life and rarely after the second month. It is an anomaly characterized by a marked hyperinflation of a lung. This defect is often associated with congenital bronchial atresia which may be secondary to intraluminal mucosal abnormalities, ab-extrinsic compression or cartilage deficiency. In some cases, however, it does not have any association with bronchial obstructions. It is more frequent in the left upper lobe and manifests itself with the typical symptoms of the respiratory distress syndrome (1, 2, 3, 5). Radiographic exams show marked hyperinsufflation and air trapping, but the involved lobe may appear radiopaque if filled with fetal lung liquid (XR: Fig. 3-A; CT: Fig. 3-B). Often loss of healthy lobes volume may be associated (1, 3, 5).

Bronchial atresia

It is a developmental anomaly incidentally discovered in adulthood, inevitably associated with congenital lobar emphysema. It is a defect characterized by focal lumen reduction or obliteration of a lobar or segmental bronchus. In most cases it is found in the upper lobes, less frequently in the middle lobe and in the lower lobes. In almost all patients a reduced perfusion of the lung distal to the obstruction coexists due to air trapping. Patients are often asymptomatic but infections of the pulmonary parenchima downstream obstructions are not uncommon (1, 2, 3, 5, 9). XR and CT examinations demonstrate hypertranslucency and hypovascularization of the lung affected, frequently associated with increased volume, contralateral dislocation of the mediastinum and mucus plugs within bronchial lumen (CT: Fig. 4-A, 4-B) (1, 3, 5, 9).

Pulmonary bronchogenic cysts

It is a cystic malformation caused by duplication of the cephalic portion of the primitive gut, resulting in abnormal development of the lung bud. Bronchogenic cysts can be localized to the mediastinum or the lung, present a thin wall with typical cellular structure of the bronchi and are filled with serous, bloody or proteinaceous fluid. They are more frequent in the middle and lower lobe (1, 2, 4, 10). The XR examination highlights a round opacity with regular margins, while CT shows the presence of a round nodular opacity with thin wall and content with density between 0 and 20 HU (in case of bloody or proteinaceous content density can be higher than 80 HU) (Fig. 5-A). The presence of air inside the cysts indicates a present or previous infectious process (1, 4, 10). On MRI cysts appear hypoin-

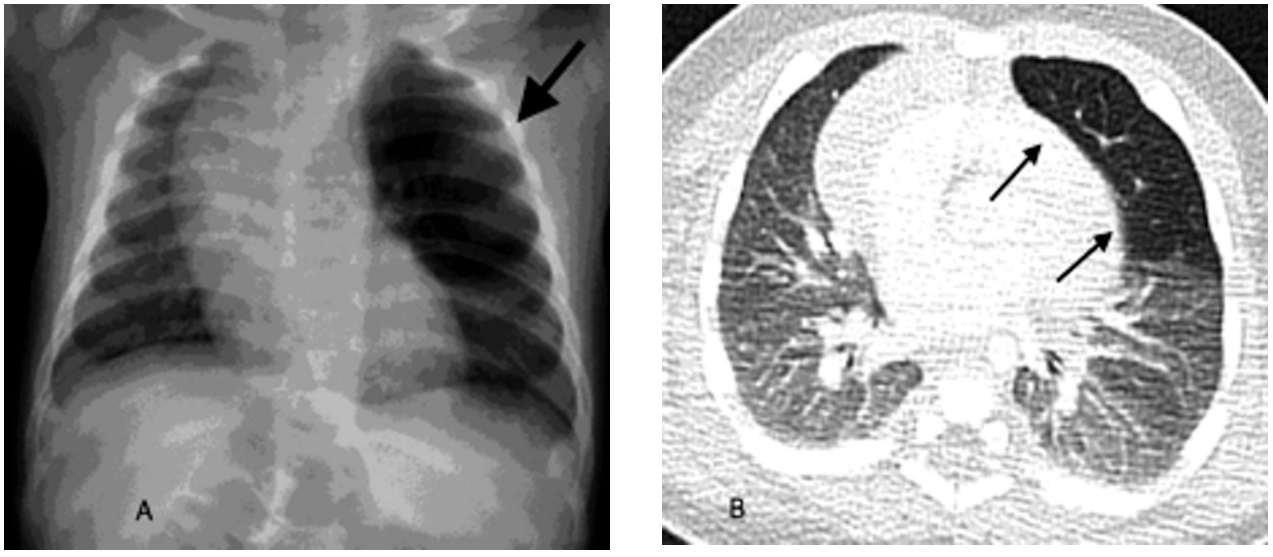

Figure 3. (A) XR shows an hyperlucency in the left upper lobe which displaces the contralateral upper mediastinum. (B) CT detects an increased radiolucency and distension of the left upper lobe; it is associated contralateral dislocation of the mediastinum

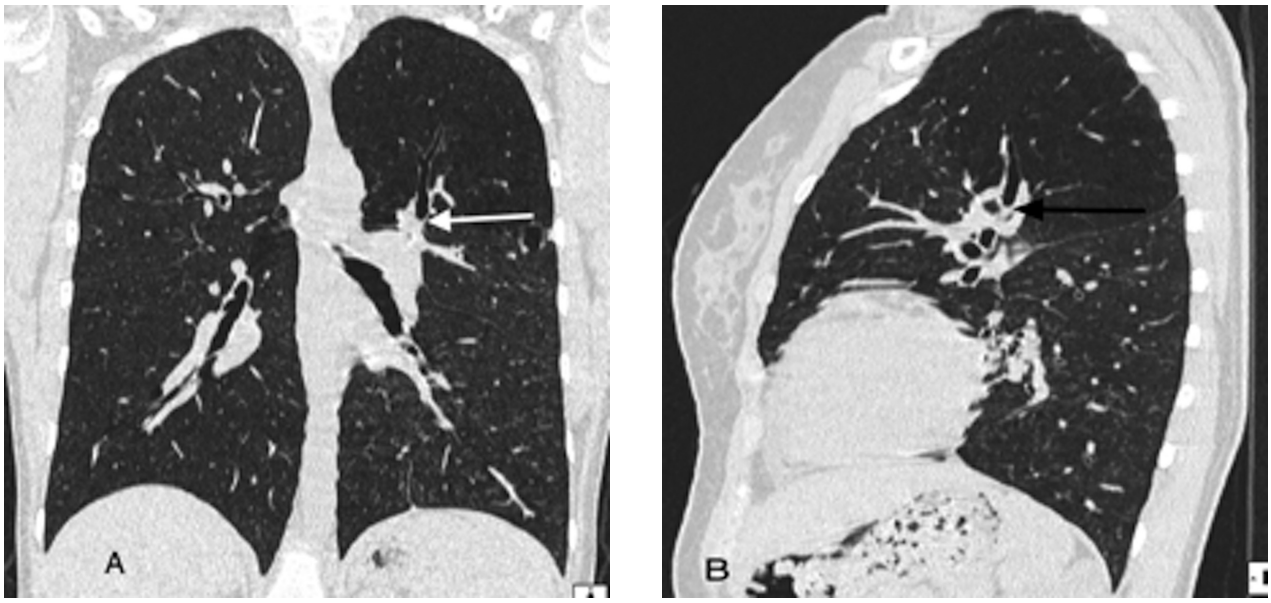

Figure 4. Stenosis of the afferent bronchus for the dorsal segment of the left upper lobe: (A) CT coronal projection (B) CT sagittal projection

tense on T1W (Fig. 5-B) and hyperintense on T2W sequences (1, 2, 4).

Congenital diaphragmatic hernias

It is a congenital disease originating from an anatomical defect in the proper development of the diaphragmatic muscle that occurs in about 1 in 3000 newborns, with estimated 60% mortality rate. This malformation, often associated with chromosomal deletions as Turner syndrome or trisomy 21, can be part of a broader syndromic picture or be an isolated anomaly. The consequent herniation of abdominal contents into the chest comes at a critical time for lung development, altering the regular formation of bronchi and vessels; this condition results in a parenchymal hypoplasia with reduced surface area for gas exchange

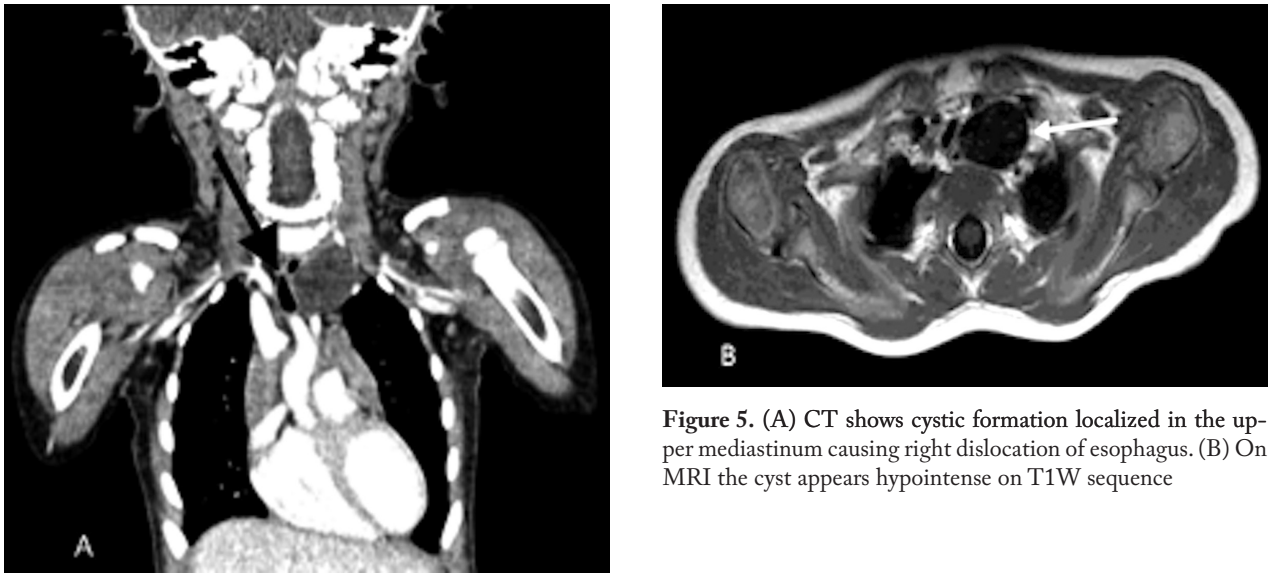

Figure 5. (A) CT shows cystic formation localized in the upper mediastinum causing right dislocation of esophagus. (B) On MRI the cyst appears hypointense on T1W sequence

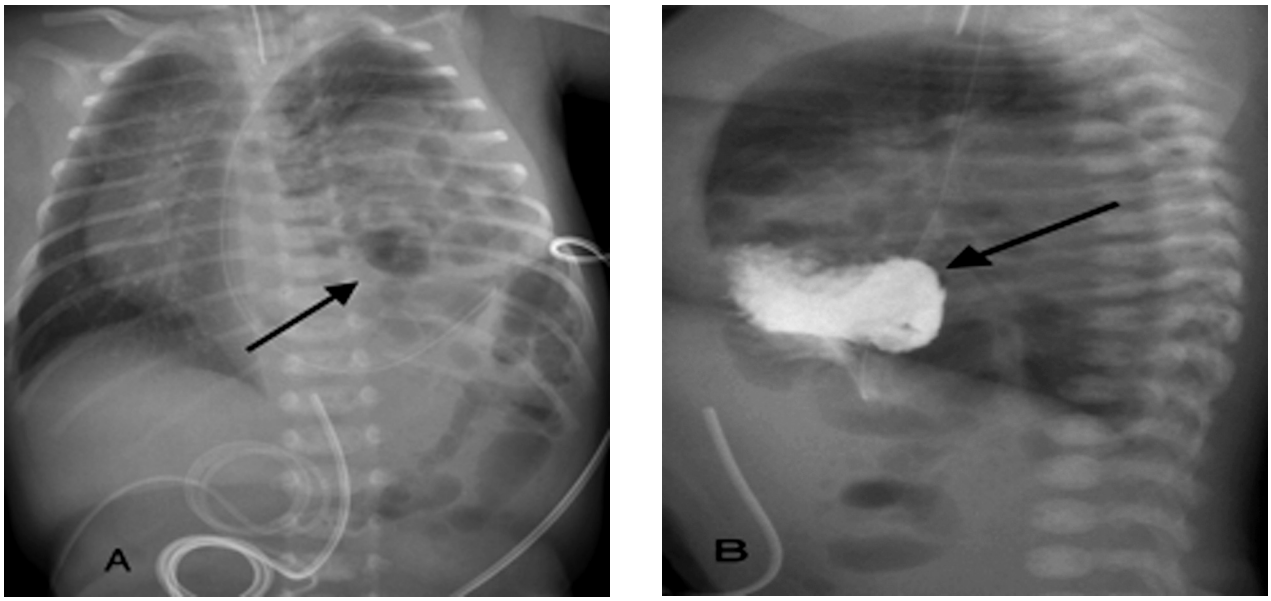

Figure 6. (A) Left hemithorax opacified by presence of a voluminous diaphragmatic hernia with protrusion of the stomach (XR). (B) By administration of Gastromiro (3 cc) via a nasogastric tube, stomach can be observed into the thorax, as well as bowel loops mainly of the small intestine (ileum) and colon (splenic flexure) (XR)

(1, 2, 8). XR shows the superelevation of the interested hemidiaphragm resulting in opacification of part of the ipsilateral hemithorax; in some cases, the colic shadow is evident in chest cavity below the involved diaphragm (Fig. 6-A, 6-B) (1, 8). CT and MRI discriminate organs and abdominal structures herniated in chest cavity, relationships with their blood supply and the volume of the residual lung. In particular, in the left diaphragmatic hernias, the entire small bowel and part of the large bowel (except left colon) can herniate into the thorax. The right diaphragmatic hernias are characterized in almost all cases by herniation, at various heights, of the right hepatic lobe into the anterior thorax. Although left hernias are more common (5:1), right hernias have the worst prognosis (estimated 57% mortality rate). Thanks to the high contrast resolution and the high definition for soft tissue, MRI can also detect presence of additional anomalies, such as adrenal glands and/or kidneys dislocation into the chest (1, 2, 8).

Conclusion

Integrated imaging is crucial for the diagnosis of congenital pulmonary malformations; in particular, XR has proven to be highly sensitive but unspecific. CT without contrast is highly specific for the diagnosis of Congenital Lobar Emphysema, Bronchial Atresia and Congenital Diaphragmatic Hernias. CT with contrast is very accurate for the diagnosis of Bronchopulmonary Sequestration, Congenital Cystic Adenomatoid Malformation and Pulmonary Bronchogenic Cysts (1). MRI has been useful only for certain specific cases, for example for the diagnosis of Pulmonary Bronchogenic Cysts (1, 2).

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