



Clinical-Diagnostic and Morphopathological Correlations in Congenital Pulmonary Malformation of the Respiratory Tract Type I in Newborn with Progressive Respiratory Distress

Clinical Case Presentation and Literature Review

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Summary

The author's reported experiencing a case of progressive acute respiratory distress caused by MCCA type I in a newborn with successful surgical resection. The presented case reflects characteristic clinical symptomatology, the preoperative image of diagnosis being of vital importance in the management of the malformation.

The histological examination made it possible to establish some characteristic structural aspects for MPCCR type 1 which presents as a vicious bronchopulmonary abnormality, which, simultaneously with cystic lung lesions, are present signs of immaturity and hypoplasia, cystic bronchiectasis that mimic perifocal adenomatoid aspects associated with primary atelectasis, focal distelectasis concurrent with arterial hypertrophic-stenotic angiopathy.

The authors conclude that early surgical intervention with economical resection of the congenital cystic formation and preservation of the remaining adjacent lung parenchyma in cases

of acute respiratory distress in the neonate with adenomatoid cystic malformation type 1 can be considered a potentially effective option, which can be performed in safe conditions with acceptable results.

Congenital cystic malformations of the lungs in children are serious lesions with similar embryological and clinical features that result from compromised interaction between mesodermal and ectodermal lung elements during embryological development. Included in this group were: bronchogenic cysts, pulmonary sequestration, congenital cystic adenomatoid malformation, and congenital lobar emphysema^[30,46]. Currently, the terminology is being revised, the spectrum of these congenital lesions includes: bronchogenic cysts, congenital airway malformation, and peripheral cystic alveolar growth anomalies. The differential diagnosis of pleuropulmonary blastoma, which is a malignant neoplasm characteristic of the child's age, should also be considered^[21].

Congenital pulmonary malformation of the airways (MPCCR), previously called congenital cystic adenomatoid malformation, is a hamartomatous lesion that includes a diverse group of cystic lung lesions that are characterized by a disordered architecture of lung tissue, determined by hyperproliferation and dilatation terminal respiratory bronchioles and lack of normal alveoli, grouped into types 0 to 4. The nomenclature of this nosological entity has been changed due to the fact that types 1, 2, and 4 are cystic lesions and only type 3 is adenomatoid^[11,21].

The incidence of MPCCR is estimated at 1:10000 – 1:35000 pregnancies^[22,37], noting that in some bibliographic sources a rather high incidence of 1:2500 live newborns is presented^[33]. Mostly, this malformation is a unilateral process (95%), limited in 80-85% of cases to a single lobe^[47].

MPCCR occurs sporadically, although in 15-20% of cases they are associated with other malformations, with type 2 of this malformation being an exception, where more than 60% of cases are associated with other malformations, including heart defects, renal, gastrointestinal and skeletal abnormalities^[40]

Case Presentation

Newborn M., male, was transferred 12 hours after birth to the neonatal surgery ward of our institution with signs of respiratory distress, rib cage bulging, subcostal draft, cyanotic integuments.

The child was born at term, weight being 3870 g, Apgar score 6/7. From heredocollateral antecedents we could mention that the mother is 26 years old. The pregnancy was monitored, at the antenatal ultrasound at the age of 36-37 weeks, macrocysts were observed in the left chest, one of which was large, developing on a hyperechoic area, the diagnosis being suggestive of congenital cystic adenomatoid malformation. No other associated abnormalities were found.

On the lung x-ray, performed on the first day of life, uneven pneumatization of the lungs was

found, with the lung volume on the right side reduced, the mediastinum being shifted to the right. In the projection of the lower lobe of the left lung, a round radio transparent cystic formation was detected, with a clear outline, without vascular and bronchial pattern, with a diameter of 5 cm x 5 cm, which was compressing the upper lung lobe; pleural sinuses – free. It was concluded that the radiological picture is characteristic of cystic adenomatoid malformation type 1.



Fig. 1 Patient M., aged 1 day. Anteroposterior lung radiograph performed on the first day of life. Explanations in the text.

The CT scan of the chest, carried out later, showed a conglomerate of oval lesions with thin walls, with air content, in the left hemithorax, the largest measuring 65 x 42 x 37 mm, being located in lung segments S6, S8, S9, S10, without post-contrast enhancement, with the presence of a hydro-aerial level with a thickness of 19 mm, liquid content having an increased protein level, possibly mucinous (native density +23UH). The formation creates a mass effect, collapsing the outstanding lung segments and moving the heart and mediastinal structures to the right. Descending aorta - with dextroposition. Adjacent to the macrosist, 2 air cysts with dimensions up to 14 mm, lateral dimensions up to 7 mm, were observed. At the same time, moderate air accumulation was found in the region of the anterior mediastinum on the right with a moderate mass effect on the thymus. Pulmonary cystic

lesions of the left lung were suggestive of congenital cystic adenomatoid malformation (fig. 2).

At the echocardiographic examination: heart cavities not dilated; the pump function of the myocardium of the left ventricle within normal limits. FOP CAP 4.0 mm (systolic-diastolic flow in the A.P. lumen); tricuspid valve insufficiency

gr.I. Moderate pulmonary hypertension (PSAP 54 mmHg.).

Blood gas evaluation revealed pH = 7.23; pCO₂ = 30 mmHg; pO₂ = 54.3 mmHg; HCO₃ = 14.1 mmol/l; BE = -13.8 mmol/l. The rest of the paraclinical evaluations were within normal limits except for AST – 82 U/L, Br. total – 29.0 mcmol/l, Br. conjugate – 12.0 mcmol/l.

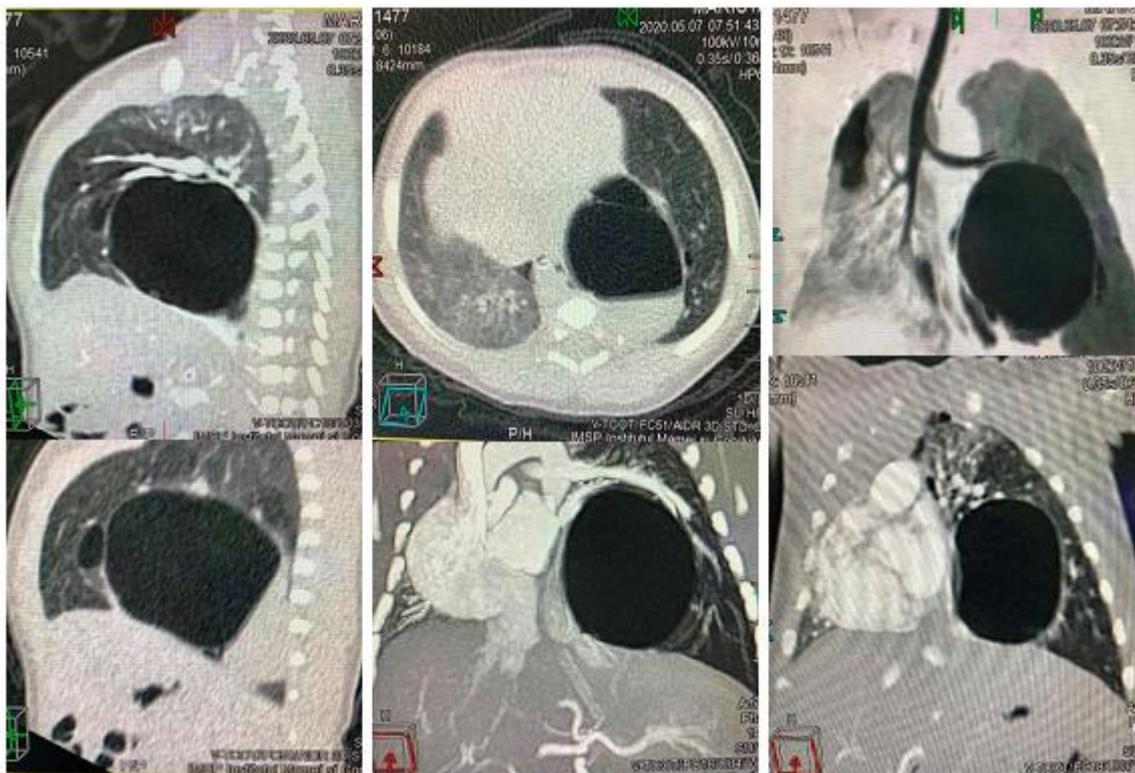


Fig. 2. Patient M. Preoperative CT scan. Explanations in the text.

Later, the child's condition continued to worsen, with respiratory depression, episodes of desaturation, which conditioned the indication for surgical intervention, resorting to lateroposterior thoracotomy on the left. Intraoperatively, the presence of the macrocyst with mucinous content, located in segmental S6, S8, S9, as well as multiple small adjacent cysts was found (fig. 3). Excision of the cystic formations was carried out with the preservation of the adjacent lung segments, and a wide respiratory wave and full expansion were observed. Pleurization of the remaining area after excision with pleural drainage according to Bulau was resorted to.

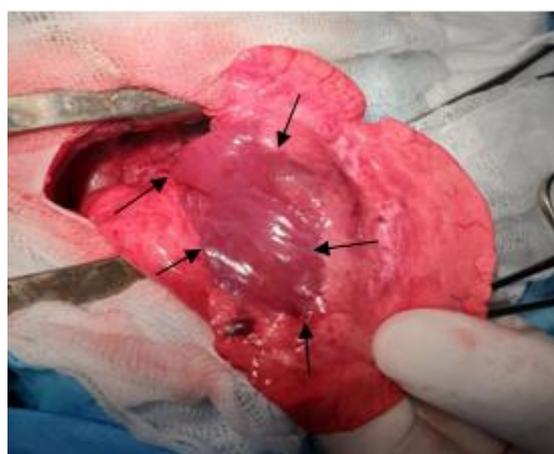


Fig. 3. Intraoperative aspect of the sector with cystic changes, located in the lower lobe of the left lung.

Postoperatively, the child was transferred to the intensive care unit, where he had been for 2 days. Later spontaneous breathing was restored, the child having a minimal need for oxygen. On the 6th postoperative day, a compressive pneumothorax developed, which required repeated reinsertion of the pleural drain, the complication being successfully resolved within a few days. The child was discharged on the 34th day in satisfactory condition.

The macroscopic examination of the resection piece found a multi-chamber appearance of the cystic formation with the predominance of two major cystic cavities delimited by a septum, but communicating with each other through an opening of ≈ 1.0 cm. The internal surface of the cysts had a pearly-rosacea color, shiny, without erosions but with small hemorrhagic extravasations. The lacunar appearance could be observed, and in some areas the appearance of a nodular pavement was observed. Inside the cyst of smaller sizes, the presence of some holes has been attested, which communicated with the lumen of the adjacent bronchi (fig. 4).

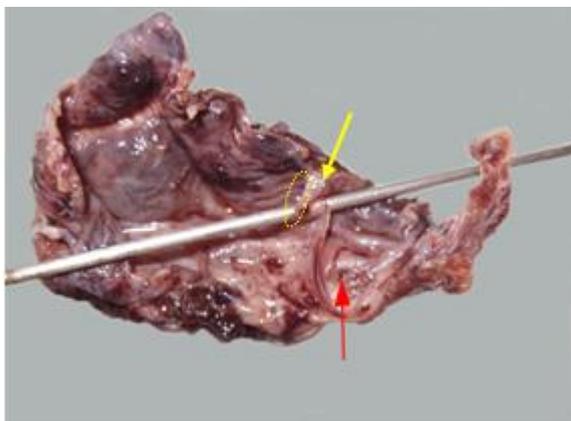


Fig. 4. The appearance of the internal surface of the multichamber cystic formation delimited by a septum, but which communicated through a hole (yellow arrow); the presence of amici holes is indicated by the red arrow

Histological examination established that the wall of the multi-chamber cyst was presented by a differentiated fibro-muscular-epithelial plaque with deficient muscular per circumscription. The

connective tissue had a frequently loose fibrillar appearance, of variable density and thickness with a mixed vascular network of various caliber. The epithelium lining the internal surface of the cystic formations was of the ciliated bronchogenic type in a non-proliferative pseudostratified focus (fig. 5). In the area of the conjunctive area, small solitary groups or chaotically dispersed acinar structures of peribronchial glands (black arrow) and immature broncho-alveolar elements (white arrow) (fig. 6) were found (fig. 6), single bronchi of medium caliber with a normal appearance (fig. 7). Groups of bronchi of various caliber were frequently found that had cystic-dilated aspects with a deficient fibro-musculo-epithelial plate, mimicking the adenomatoid appearance, while at the same time immature or hypertrophied arterial vessels were determined in various proportions with the reduction of the bronchiolo-alveolar component and alveolar in that area (fig. 8).

In some areas the parenchyma between the cystic parietal plate and the pleura was much more diminished, being characterized by a discrepancy between the bronchial and vascular caliber, the adventitia of the vessels being sclerotized the alveoli large but reduced in number, and in some foci in the perivascular areas the interstitium was slightly thickened.

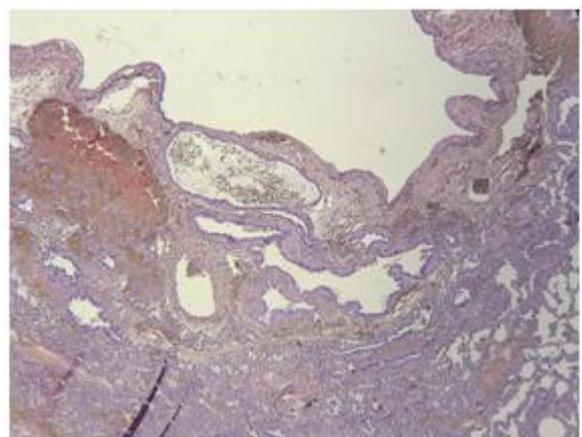


Fig.5. Fibromusculo-epithelial cystic parietal wall differentiated with abundance of bronchial

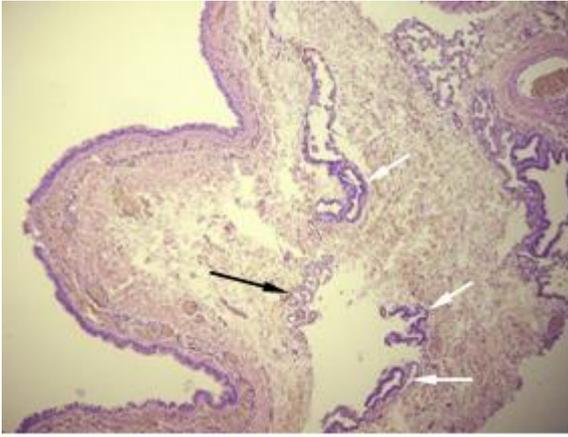


Fig.6. Deficient fibromusculo-epithelial cystic parietal wound, islands of glands and immature bronchio-alveolar elements in the lax fibrillary connective area

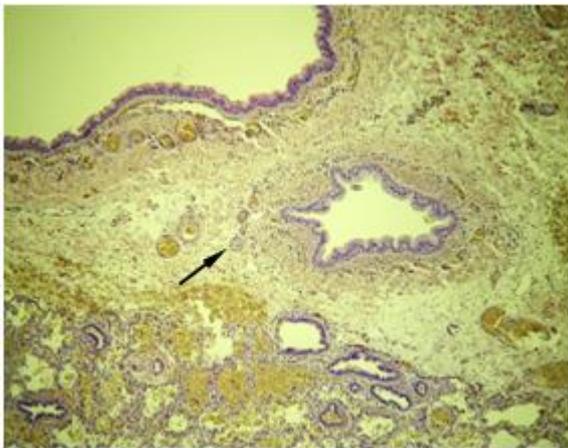


Fig.7. Cystic parietal wound with attestation of a medium caliber bronchus in the vascularized conjunctival area with single glandular acini, clusters of ectasis bronchioles

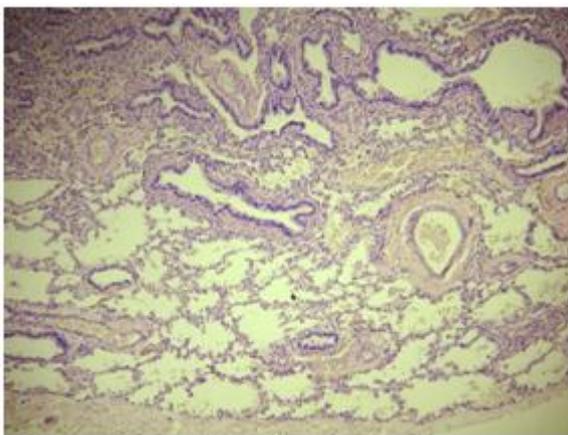


Fig.8. Bronchiectasis in the pericystic area discordance between the vascular caliber and the bronchial lumen, immaturity and hypoplasia of the component bronchiolo-alviolare terminale

The perichytic alveolar parenchyma on extensive areas was aatelectasis with a thickened interstitium, an insignificant hypercellularization was observed, with the accentuation of the adventitia of arterial vessels, some with aspects of an immature fetal type (fig. 9), and others - with stenotic hypertrophic changes. It was observed that the peribronchiolar vessels appeared in the form of bundles with angiomatosis, and an accentuation of the sclerosing processes of the adventitia was also observed. Simultaneously with these changes, distelected foci were determined with varied interalveolar interstitium, and single desquamated alveolocytocytes were observed intraalveolar (fig. 10).

Compared to the pericystic areas, towards the periphery, the alveolar parenchyma showed a much more differentiated structure, with varied morpho-functional aspects, characterized by ectasized bronchiolar segments lined with partially vacuolated serous and pseudo-mucous content. The interalveolar interstitium was thin, with varying degrees of maturation. The small-caliber arterial and segmental capillary vascular network was characterized by mild stenotic hypertrophy (Fig. 11). At the same time, segments with reduced aeration, slightly sclerogenized interstitium, unevenly hypertrophied and stenosed arteries and arterioles were detected (fig. 12).

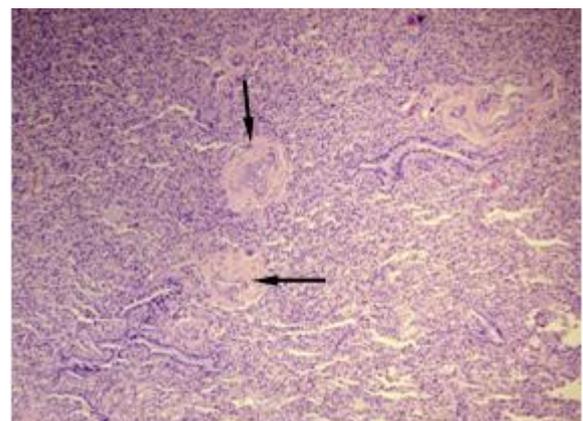


Fig.9. Primary atelectasis, fetal type vessels, interstitial immaturity with mild interstitial thickening.

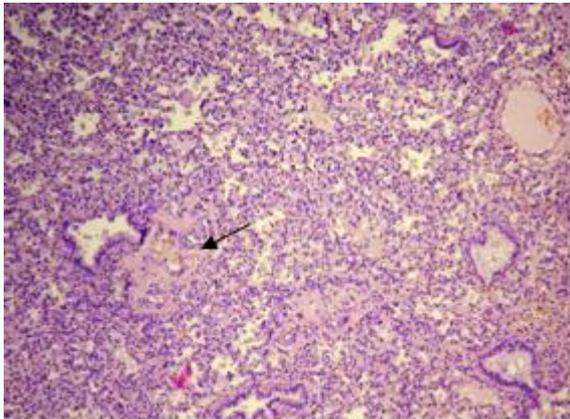


Fig.10. Moderate dystelectasis, mild bronchiole ectasia, vascular devices in peribronchial bundles with angiomatosis

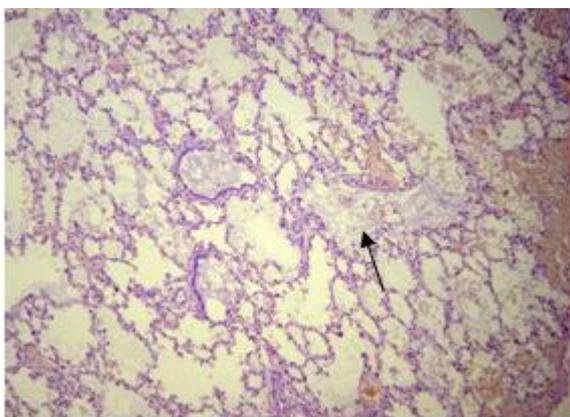


Fig.11. Respiratory bronchioloectasis lined with sero-pseudomucous content.

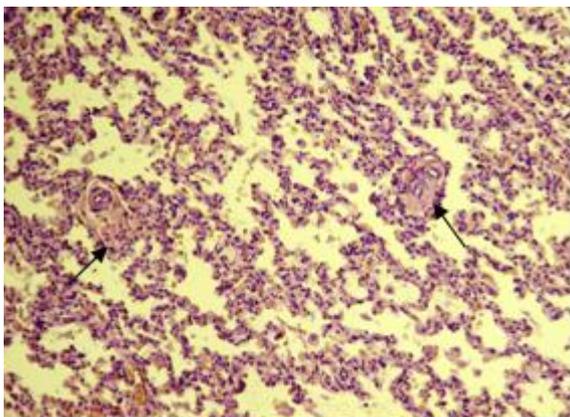


Fig. 12. Respiratory bronchioloectasis lined with sero-pseudomucous content

Similar changes were also witnessed at the level of the mucilaginous chamber cyst, but compared to the larger one in the parietal fibro-musculo-epithelial plate, a deficient musculature was determined, sometimes manifested by dispersed myocytes. In the connective area, cystic bronchial devices with a small communication, with immature cartilaginous plates could be observed (fig. 13). We note that the presence of a chronic or acute process, including sclero-cicatricial changes in the examined lung tissue as well as stromal or muscle proliferative changes were not found.

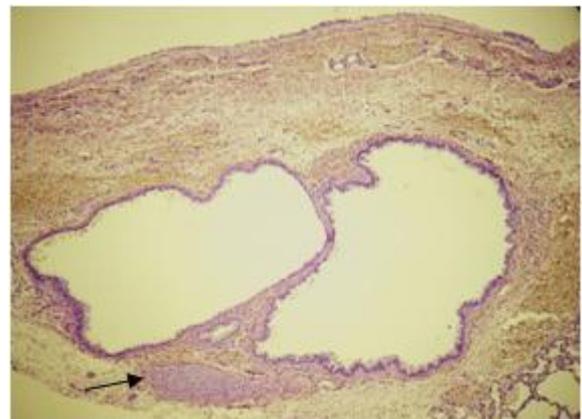


Fig.13. Cystic bronchiectasis with fibro musculo cartilaginous plate dysplasia (arrow).

At 3 months postoperatively, complete re-expansion of the left lung was observed, with the presence of air mucrocysts located in the lower lobe of the left lung and adjacent pneumofibrotic changes (fig. 14).

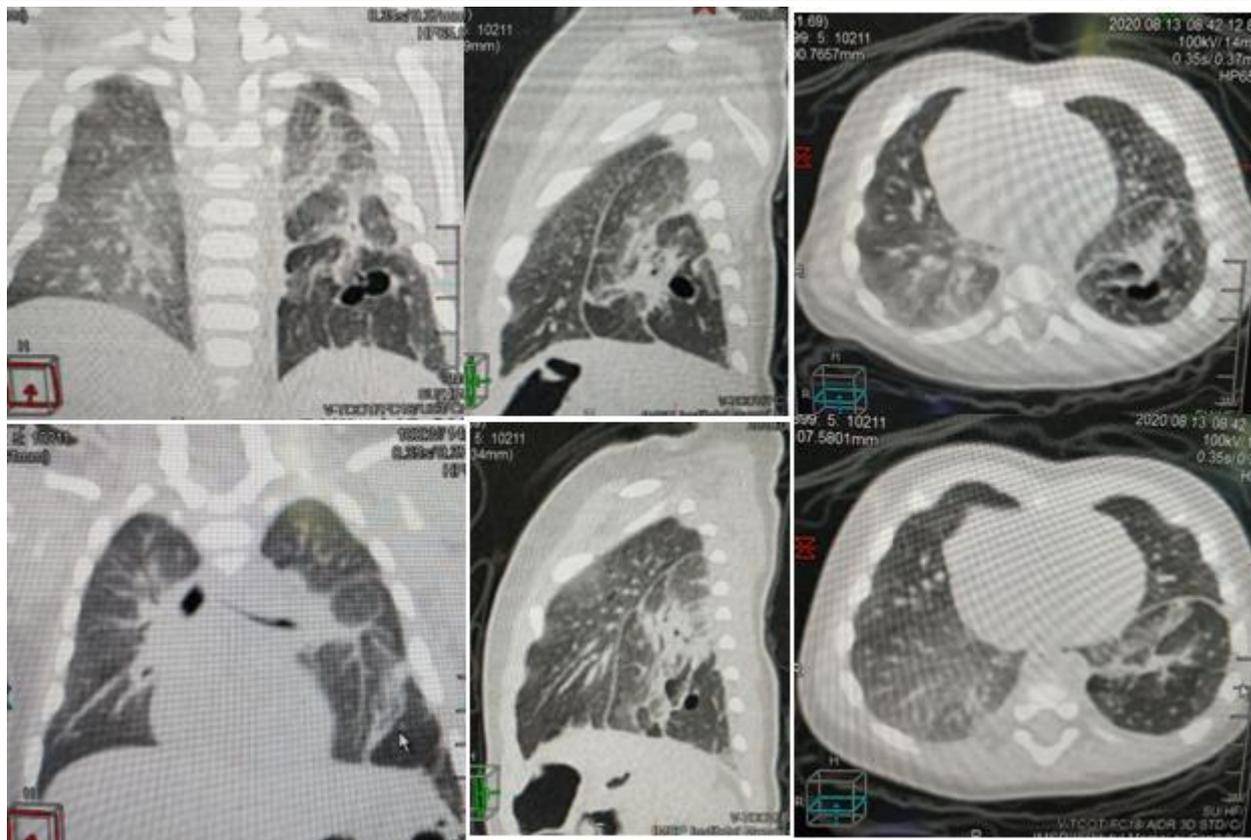


Fig. 14.The patient MEffective postoperative CT 3 months after surgery. Explanations in the text.

At lung scintigraphy, a diffuse decrease in pulmonary perfusion was found in the left lung, predominantly in the remaining segments of the lower lobe (fig. 15)

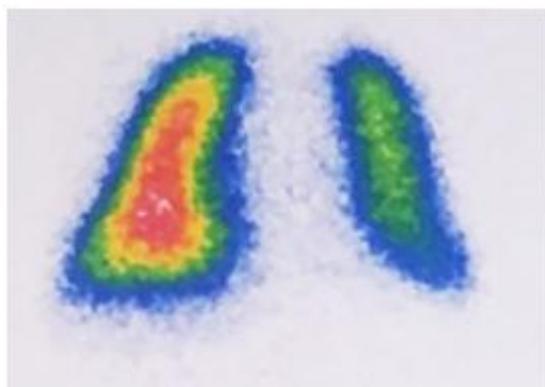


Fig. 15.Lung scintigraphy performed 90 days postoperatively. Decreased pulmonary perfusion in the left lung, especially in the basal segments.

Discussions

Congenital pulmonary malformation of the airways is the most common congenital lesion detected in the neonatal lung, which contributes to the development of respiratory distress, infection,

and pneumothorax, with the pathogenetic mechanisms remaining unknown^[20].

Congenital cystic lung disease is considered to have been reported for the first time in 1687 by Bartholin^[17]. A variety of lung hamartomas, including congenital cystic lung disease, was described by Koontz (1925) based on a large case series^[41]. For the first time, the term "congenital cystic adenomatoid malformation" was described in the medical literature by Ch'In and Tang (1949), who considered that the first description of the pathology was made by Stoerk in 1897 as "bronchial cystic adenoma"^[2, 3, 28, 32].

MPCCR is usually diagnosed in newborns and young children (85% of cases)^[12], rarely been described in older children^[16,31] or adults^[14,39]. At the same time, some authors consider that more than half of diagnosed patients are asymptomatic, being occasionally detected radiologically in adulthood, and only 25% present clinical symptoms at birth^[25].

The congenital cystic adenomatoid malformation is characterized by the absence of normal alveoli

with excessive proliferation and cystic dilation of terminal respiratory bronchioles. Histologically, ciliated, cuboidal, or columnar cells can be seen lining the cystic lesions with a lack of architecture and the absence of cartilage. Bronchial atresia reflects the etiology of the lesion^[29]. Mucous cell clusters, visualized almost exclusively in type 1 of this anomaly, are considered as premalignant lesions with a risk of progression to mucinous adenocarcinoma^[36].

Hybrid lesions can also be encountered, consisting of cystic adenomatoid malformation and bronchopulmonary sequestration^[6,9]. Cases of coexistence of this malformation with bronchogenic cyst have been described^[34].

The persistence of these congenital lesions can cause recurrent respiratory infections, also having the potential for malignancy^[19,48]. Some studies found that 4%-10% of all pulmonary malignant neoplasias found in children and adolescents are associated with cystic malformations^[5,38]. Associations have been found between this malformation and some malignant tumors such as rhabdomyosarcoma^[10], pleuropulmonary blastoma^[4], bronchoalveolar carcinoma^[45] and mucinous adenocarcinoma^[18,23]. Cases of malignancy associated with congenital pulmonary malformation of the airways are described even in newborns^[25].

Stocker J.T. and I look (1977) classified the adenomatous cystic malformation into 3 predominant histopathological subtypes (types 1, 2, and 3)^[43], later 2 more subtypes (types 0 and 4) were described with a change in the nomenclature of the malformation^[42]. Type 0 (acinar dysplasia) is rare (<3%) and represents a pulmonary microcystic process with an unfavorable prognosis. Type 1 (60%) is characterized by the presence of multiple large cysts (> 2 cm) or a single dominant cyst, lined with pseudostratified ciliated epithelium, usually involving only part of the lung lobe with a favorable prognosis. Type 2 (20%) – microcystic, consists of multiple small cysts (0.5-2cm) unevenly arranged (sponge appearance), which appear at the terminal

bronchioles, this type frequently being associated with other serious congenital malformations. Type 3 (<10%) is a bulky solid mass (adenomatoid appearance), which consists of microcysts and solid tissue with a bronchiole-like structure lined with cuboidal ciliated epithelium and separated by areas of non-ciliated cuboidal epithelium. Type 4 (10%) has a macrocystic appearance with a lack of mucus-producing cells, having a major potential for malignancy (pleuropulmonary blastoma)^[16].

Prenatal ultrasound monitoring, most cases of MPCCP being diagnosed at 18-22 weeks of gestation, supplemented with magnetic resonance allows the diagnosis of congenital cystic adenomatoid malformation, with a reported accuracy of up to 91%^[8,15]. The ultrasound appearance of malformative cystic lesions is dependent on the type of anomaly and the particularities of evolution, including spontaneous regression and complete resolution of the lesions during the prenatal period or the presence of complications such as: pleural and/or pericardial effusions, hydrops, polyhydroamnios.

The radiological examination highlights the presence of cystic formations of various sizes, with thin walls, containing air. In some cases these cysts may contain fluid. Computed tomography aims at the differential diagnosis of cystic adenomatoid malformation with bronchogenic cysts, lobar emphysema or pulmonary sequestration^[19,35].

Prenatal treatment options for MPCCR include maternal steroid therapy (administration of betamethasone), minimally invasive procedures, or open fetal surgery. In macrocystic lesions with decompressive purpose, thoracoamniotic shunts can be used, in microcystic ones, open surgery is indicated^[8,13].

Postnatally, several authors opt for an early surgical treatment in all cases of bronchopulmonary malformations^[44], including in MPCCR^[7,24]. Some macrocystic forms of MCCR may require surgical resections even in the neonatal period to save viable lung parenchyma and resolve mediastinal displacement^[27]. Early

elective resections of asymptomatic forms are based on several arguments, including: avoidance of long observation periods with avoidance of repeated imaging investigations, no imaging method can confirm the morphopathological diagnosis, avoidance of malignancy and, potential for compensatory lung development^[26].

The management of asymptomatic forms remains controversial. Some reports indicate that only 10% of asymptomatic MPCCR develop complications when left untreated, suggesting that a non-surgical approach could be a reasonable alternative until symptoms appear^[1], with surgery reserved for symptomatic forms with the association of recurrent infections or pneumothorax. In favor of this strategy is also the potential for spontaneous resolution established by some studies^[8].

Conclusion

Therefore, the results of the histological examination allowed to establish some characteristic structural aspects for MPCCR type 1 which presents as a vicious bronchopulmonary abnormality, which, simultaneously with cystic lung lesions, are present signs of immaturity and hypoplasia, cystic bronchiectasis that mimic adenomatoid aspects perifocal associated with primary atelectasis, focal distelectasis concurrent with arterial hypertrophic-stenotic angiopathy. The presented case demonstrates that early surgical intervention with economical resection of the congenital cystic formation and preservation of the remaining adjacent lung parenchyma in cases of progressive acute respiratory distress in the newborn with adenomatoid cystic malformation type 1 can be considered as potentially effective option, which can be performed safely with acceptable results.

Bibliografie

1. Aziz D., Langer J.C., Tuuha S.E., Ryan G., Ein S.H., Kim P.C. Perinatally diagnosed symptomatic congenital cystic adenomatoid malformation: to resect or not? *J. Pediatr. Surg.* 2004;39:329-34.

2. Bain G.O. Congenital adenomatoid malformation of the lung. *Chest J.* 1959; 36(4):430-3.
3. Belanger R., La Fleche, Picard J.L. Congenital cystic adenomatoid malformation of the lung. *Thorax.* 1964; 19:1-11.
4. Brcic L., Fakler F., Eidenhammer S., Thueringer A., Kashofer K. et al. Pleuropulmonary blastoma type I might arise in congenital pulmonary airway malformation type 4 by acquiring a Dicer 1 mutation. *Virchows Arch.* 2020; doi: 10.1007/s00428-020-02789-6.
5. Casagrande A., Pederiva F. Association between congenital lung malformations and lung tumors in children and adults: A systematic review. *J. Thor. Oncol.* 2016; 11(11):1837-45.
6. Chen H.W., Hsu W.M., Lu F.L., Chen P.C., Jeng S.F., et al. Management of congenital adenomatoid malformation and bronchopulmonary sequestration in newborns. *Pediatr. Neonatol.* 2020; 51(3):172-7.
7. Conforti A., Aloï I., Trucchi A., Morini F., Nahom A. et al. Asymptomatic congenital cystic adenomatoid malformation of the lung: Is it time to operate? *J. Thor. Cardiovasc. Surg.* 2009; 138(4):826-30.
8. David M., Lamas-Pinheiro R., Henriques-Coelho T. Prenatal and postnatal management of congenital pulmonary airway malformation. *Neonatol.* 2016; 110:101-15.
9. De Leon-Urena Z.A., Sadowinski-Pine S., Jamaica-Balderas L., Penchyna-Grub J. Pulmonary sequestration associated with congenital pulmonary airway malformation. *Bol. Med. Hosp. Infant Mex.* 2018;75:105-12.
10. Doladzas T., Arvelakis A., Karavokyros I.G., Gougoudi E., Pikoulis E., Patsouris E. Primary rhabdomyosarcoma of the lung arising over cystic pulmonary adenomatoid

- malformation. *Pediatr. Hematol. Oncol.* 2005; 22(6):525-9.
11. Durell J.,Lakhoo K. Congenital cystic lesions of the lung. *Early Human Develop.* 2014; 90: 935-39.
 12. El Amraoui W.,Bentalha A., Hamri H., El Kettani S.E., El Koraichi A. Congenital cystic adenomatoid malformation – dangers of misdiagnosis: a case report. *J. med. Case Rep.* 2017; 11:212. DOI 10.1186/s13256-017-1349-5.
 13. Fan D., Wu S., Wang R., Huang Y., Fu Y. et al. Successfully treated congenital cystic adenomatoid malformation by open fetal suegery. *Medicine.* 2017; 96:2.
 14. Feng A., Cai H., Sun Q., Zhang Y., Chen L., Meng F. Congenital cystic adenomatoid malformation of lung in adults: 2 rare cases report and review of the literature. *Diagn. Pathol.* 2012; 7:37. Doi:10.1186/1746-1596-7-37.
 15. Gallegos S.A., Sibaja M.J.R., Gaona J.M.G., Calvo J.A.R., Torrez B.V., et al. Congenital adenomatoid cystic malformation: expectant management outcomes. *J. Preg. Child Health.* 2018; 5: 387. doi:10.4172/2376-127X.1000387.
 16. Goyal J.P., Jindal S., Mishra M., Bhakhiri B.K. Congenital cyst adenoid malformation masquerading as bronchial asthma. *Int. J. App. Basic Med. Res.* 2020; 7:199-201.
 17. Holder T.M., Christy M.G. Cystic adenomatoid malformation of the lung. *J. Thor. Cardiovasc. Surg.* 1964; 47(5):590-7.
 18. Ishida M., Igarashi T., Teramoto T., Hanaoka J., Iwai M. et al. Mucinous bronchioloalveolar carcinoma with K-ras mutation arising in type 1 congenital cystic adenomatoid malformation: a case report with review of the literature. *Int. J. Clin. Exp. Pathol.* 2013; 6(11):2597-2602.
 19. Jain A.,Anand K., Singla S., Kumar A. Congenital Cystic Lung Diseases. *J. Clin. Imaging Sci.* 2013;3:5. <http://www.clinicalimagingscience.org/text.asp?2013/3/1/5/106620>.
 20. Jiang Y., Luo Y., Tang Y., Moats R., Warburton D. et al. Alteration of cystic airway mesenchyme in congenital pulmonary airway malformation. *Scientific Rep.* 2019; 9:5296. <https://doi.org/10.1038/s41598-019-41777-y>.
 21. Jones K.D. Pulmonary cystic disease and its mimics. *Surg. Pathol.* 2020; 13:141-63.
 22. Kanavi J.V., Thangaraj J., Thomas A. Congenital cystic adenomatoid malformation (CCAM): antenatal and postnatal management. *Int. J. reprod. Contracept. Obstet. Gynecol.* 2017; 6(6):2447-51.
 23. Kim M.Y., Kang C.H., Park S.H. Multifocal synchronous mucinous adenocarcinomas arising in congenital pulmonary airway malformation: a case report with molecular study. *Histopathology.* 2014;65: 926-32.
 24. Kim Y.T., Kim J.S., Park J.D., Kang C.H., Sung S.W., Kim J.H. Treatment of congenital cystic adenomatoid malformation – does resection in the early postnatal period increase surgical risk? *Eur. J. Cardio-thor. Surg.* 2005; 27:658-61.
 25. Koh J., Jung E., Jang S.J., Kim D.K., Lee B.S. et al. Case of mucinous adenocarcinoma of the lung associated with congenital pulmonary airway malformation in a neonate. *Korean J. Pediatr.* 2018; 61(1):30-4/
 26. Komori K.,Kamagata S., Hirobe S., Toma M., Okumura K et al. Radionuclide imaging study of long-term pulmonary function after lobectomy in children with congenital cystic lung disease. *J.Pediatr. Surg.* 2009;44:2096-2100.
 27. Kotecha S.,Barbato A., Bush A., Claus F., Davenport M. et al. Antenatal and postnatal management of congenital cystic

- adenomatoid malformation. Paediatr. Respir. Rev. 2012;13:162-71.
28. Kwittken J., Reiner L. Congenital cystic adenomatoid malformation of the lung. Pediatrics. 1962; 30:759-68.
29. Lakhoo K. Management of congenital cystic adenomatous malformations of the lung. Arch Dis. Child Fetal Neonatal Ed. 2009;94:F73-F76.
doi:10.1136/adc.2007.130542.
30. Lima M., Gargano T., Ruggeri G., Manuele R., Gentili A. et al. Clinical spectrum and management of congenital pulmonary cystic lesions. Pediatr. Med. Chir. 2008; 30(2):79-88.
31. Mathai A.M., Kini H., Pai M.R., Shetty A.B., Murali N. Congenital cystic adenomatoid malformation of lung type 1. J. Pediatr. Surg. 2010; 45:E25-E28.
32. Moncrief M.W., Cameron A.H., Astley R., Roberts K.D., Abrams L.D., Mann J.R. Congenital cystic adenomatoid malformation of the lung. Thorax. 1969; 24:476-87.
33. Morini F., Zani A., Conforti A., van Heurn E., Eaton S. et al. Current management of congenital pulmonary airway malformations: A “European Pediatric Surgeons Association” Survey. Eur. J. Pediatr. Surg. 2018; 28(1):1-5.
34. Oh M.H., Jung E.A., Lee J.H., Cho H.D., Seo K.H., et al. Coexistence of intrapulmonary bronchogenic cyst and congenital cystic adenomatoid malformation – A case report. Korean J. Pathol. 2011; 45:92-5.
35. Pardo L., Viveros J.M., Carrillo A., Polo A., Lopez A. et al. Radiological manifestations of congenital lung malformations. Experience of three hospitals in Bogota. Rev. Colomb. Radiol. 2019; 30(2):5117-25.
36. Pogoriler J., Swarr D., Kreiger P., Adzick N.S., Peranteau W. Congenital cystic lesions. Redefining the natural distribution of subtypes and assessing the risk of malignancy. Am. J. Surg. Pathol. 2019; 43(1):47-55.
37. Raman V.S., Agarwala S., Bhatnagar V., Panda S.S., Gupta A.K. Congenital cystic lesions of the lungs: The perils of misdiagnosis - A single-center experience. Lung India. 2015; 32:116-8.
38. Ramos S.G., Barbosa G.H., Tavora F.R., Jeudy J., Torres L.A., et al. Bronchioloalveolar carcinoma arising in a congenital pulmonary airway malformation in a child: case report with an update of this association. J. Pediatr. Surg. 2007;42:E1-4.
39. Scampolino A., Ambrosini A., Turini E., Morandi U., Stefani A. Congenital cystic adenomatoid malformation in adults, presenting as a single cyst. Asian Cardiovasc. Thorac. Ann. 2018; 26(5):407-9.
40. Sfakianaki A.K., Copel J.A. Congenital cystic lesions of the lung: congenital cystic adenomatoid malformation and bronchopulmonary sequestration. Rev. Obstet. Gynecol. 2012;5(2):85-93.
41. Spector R.G., Claireaux A.E., Williams E.R. Congenital adenomatoid malformation of lung with pneumothorax. Arch. Dis. Child. 1960; 35(183):475-80.
42. Stocker J.T. Congenital pulmonary airway malformation: a new name and expanded classification of congenital cystic adenomatoid malformation of the lung. Histopathology. 2002; 41(Suppl):424-31.
43. Stocker J.T., Madewell J.E., Drake R.M. Congenital cystic adenomatoid malformation of the lung: classification and morphologic spectrum. Hum. Pathol. 1977; 8:155-71.
44. Style C.C., Cass Darel L., Verla M.A., Cruz S.M., Lau P.E. et al. Early vs late resection of asymptomatic congenital lung malformation. J. Pediatr. Surg. 2019; 54(1):70-4.

45. Sudou M., Sugi K., Murakami T. Bronchioloalveolar carcinoma arising from a congenital cystic adenomatoid malformation in an adolescent: The first case report from the Orient. *J. Thorac. Cardiovasc. Surg.* 2003; 126:902-3.
46. Takeda S., Miyoshi S., Inoue M., Omori K., Okumura M. et al. Clinical spectrum of congenital cystic disease of the lung in children. *Eur. J. Cardio-thor. Surg.* 1999; 15:11-7.
47. Vladareanu R., Pelinescu-Onciul D., Zvanca M., Mihailescu D. Congenital cystic adenomatoid malformation. *Gynaecol. Perinatol.* 2007;16(2):68-72.
48. Yoo J., Lee S.M., Jung J.H., Ahn M.I., Cho D.G. et al. Adenocarcinoma arising in type 1 congenital cystic adenomatoid malformation: A case report and review of the literature. *Korean J. Pathol.* 2008; 42:396-400.