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PATHOMORPHOLOGICAL FEATURES OF BRONCHIOLOECTATIC EMPHYSEMA, TRACHEOESOPHAGEAL FISTULAS, AND STENOSIS OF THE BRONCHI AND TRACHEA

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Abstract
Bronchopulmonary malformations in congenital and early developmental anomalies often manifest through structural defects in the bronchiolar wall, particularly involving the muscular layer. Aplasia and hypoplasia of the smooth muscle components lead to functional and morphological disturbances, including ectatic dilatation of the bronchioles. This process contributes to the development of centrilobular emphysema, particularly in the form known as Leschke emphysema. The underdevelopment of the mucosal lining and the muscular layer results in poor airway stability, facilitating emphysematous transformation of the respiratory bronchioles. Such anatomical and functional defects predispose patients to recurrent respiratory infections. Over time, secondary inflammatory processes dominate the clinical picture, often evolving into bilateral, polysegmental bronchopneumonia. Postmortem examination commonly reveals pathological changes in lung segments 2, 6, 7, 8, 9, and 10 near the pulmonary hilum, where dense foci of mixed exudative inflammation, areas of atelectasis, and emphysema are identified. These morphological changes are crucial for understanding the pathogenesis of post-bronchopneumonic complications and are essential for accurate diagnosis and treatment planning.
The current study aims to investigate the pathomorphological features of bronchioloectatic emphysema, tracheoesophageal fistulas, and bronchial and tracheal stenosis, emphasizing their clinical relevance and diagnostic implications.

Keywords: Bronchiolectasis, Leschke emphysema, bronchial stenosis, tracheoesophageal fistula, pathomorphology, deformation, respiratory bronchioles, bronchopneumonia.

Introduction

Globally, congenital developmental anomalies are diagnosed in approximately 4–6% of all live births per 1,000 newborns, with bronchopulmonary anomalies accounting for 4–7% of these cases [1]. In 2022, an estimated 34 million newborns were born worldwide with bronchopulmonary malformations, of which approximately 20–25% died within the first 7 days of life [2]. In the

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United States and European countries, this figure is around 4.5 million annually, while in the Russian Federation and other CIS countries, it reaches about 7 million newborns each year [3]. In Uzbekistan, approximately 2,842 infants are born annually with bronchopulmonary anomalies, and among them, nearly 720 die during the early neonatal period. An additional 25–35% of these infants die in the late neonatal period [4]. These statistics highlight the urgency of this issue and justify the relevance of the dissertation topic. While maternal and infant mortality rates remain low in developed countries, Uzbekistan ranks 15th among 174 countries in terms of infant deaths due to congenital anomalies, indicating significant gaps in maternal and child health protection efforts in the country [5].

Despite numerous studies conducted in CIS countries focusing on bronchopulmonary anomalies, the mortality rate due to congenital anomalies among infants over the past five years has declined inconsistently and remains on average at about 1.2% annually [6]. Following the COVID-19 pandemic in 2020, this rate has not significantly improved. In Uzbekistan, between 2018 and 2023, the average mortality rate remained between 1.2% and 1.7%, with the highest rate observed in 2022. That year, 27 infant deaths per 1,000 live births were recorded due to secondary infections related to congenital anomalies (a total of 15,660 neonatal deaths) [7].

Common types of bronchial developmental anomalies in neonates include dysontogenetic bronchiectasis, bronchial atresia, congenital tracheobronchoesophageal fistulas, cystic bronchial hypoplasia, and solitary pulmonary cysts [8]. These conditions are not only congenital but also highly prone to complications and high mortality. Therefore, studying their pathomorphological features is of critical importance in modern pediatric pathology and neonatal care [9].

Research Objective

To study the pathomorphological features of bronchioloectatic Leschke emphysema, tracheoesophageal fistulas, and stenosis of the bronchi and trachea in newborns.

Materials and Methods

Data and autopsy materials from 113 cases confirmed with congenital anomalies, referred to the Republican Pathological Anatomy Center, were used to obtain bronchial and lung tissue samples. Morphological methods included hematoxylin and eosin staining, and the obtained data on tissue structural components were analyzed.

Results and Discussion

Aplasia and hypoplastic changes in the muscular system of the bronchiolar walls manifest as centrilobular pulmonary emphysema. The main factor is the underdevelopment of the myocyte population within the muscular layer of the bronchiolar walls, accompanied by a reduced development of the mucosal layer. As a result, the bronchial walls undergo ectatic expansion, leading to the formation of emphysematous dilated respiratory bronchioles. This condition, in turn, predisposes patients to secondary infectious factors, primarily developing into bilateral polysegmental bronchopneumonia. At autopsy, mixed exudative inflammatory foci, atelectasis, and emphysema were identified in lung segments 2, 6, 7, 8, 9, and 10 around the lung root. These findings correspond clinically and morphologically to bronchopneumonia following emphysema.

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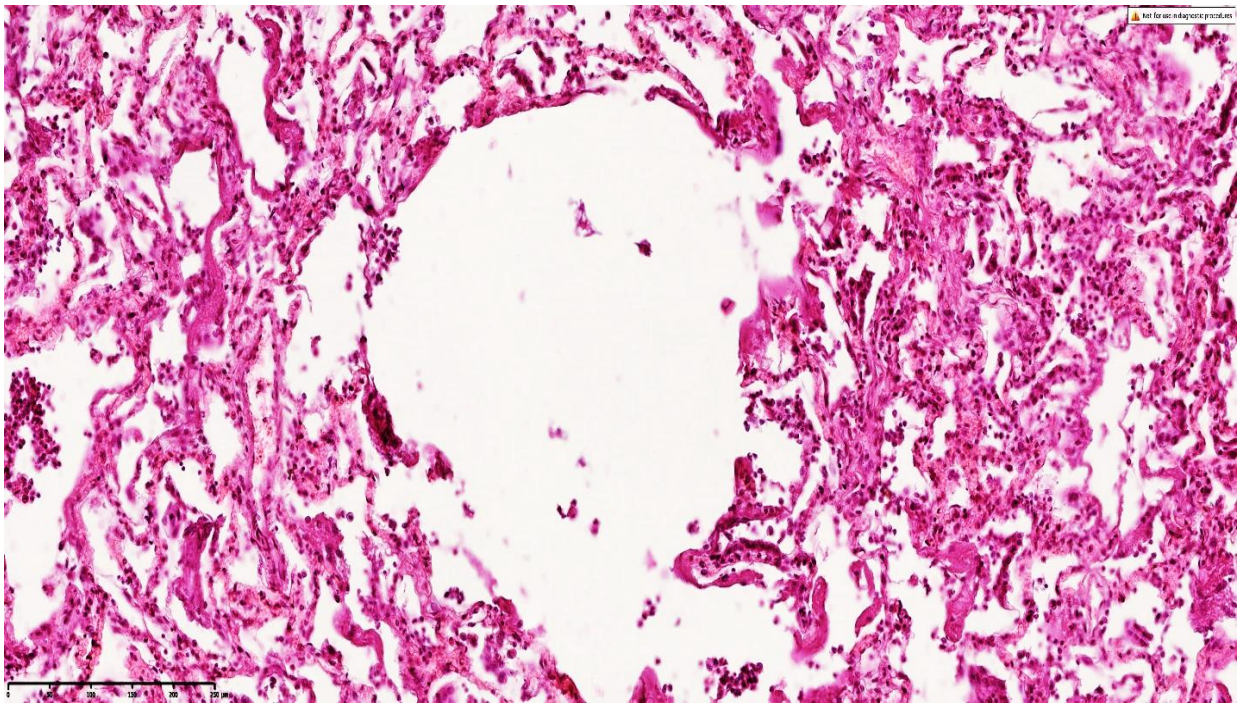
In the cystic hypoplasia type, dilated foci replace terminal bronchioles. These foci have a mucosal layer with a thickness similar to that of an indistinct alveolar wall, and the vessels appear irregular and tortuous. Alveolar cells are not identifiable in these areas. Desquamation of the mucosal epithelium of bronchioles is observed, with vascular dilatation, leukocyte diapedesis, macrophages, and a variable mixture of other tissue components in the inflammatory exudate. Bronchiolar obstruction due to these processes is characteristic. Therefore, to prevent the development of aspiration pneumonia in patients, morphological criteria necessitate surgical interventions such as lobar resections and pneumonectomy (see Figure 1).



1 Figure: In bronchioloectatic Leschke emphysema, a centrilobular cystic dilated focus is identified (1), with atelectatic foci in the alveolar spaces at its periphery (2). Sclerotic and atrophic changes are observed in the walls of small bronchioles (3). At the perimeter of the cystic dilated focus, developing inflammatory exudate and signs of bronchopneumonia are present. Staining: H&E. Magnification: 10×10.

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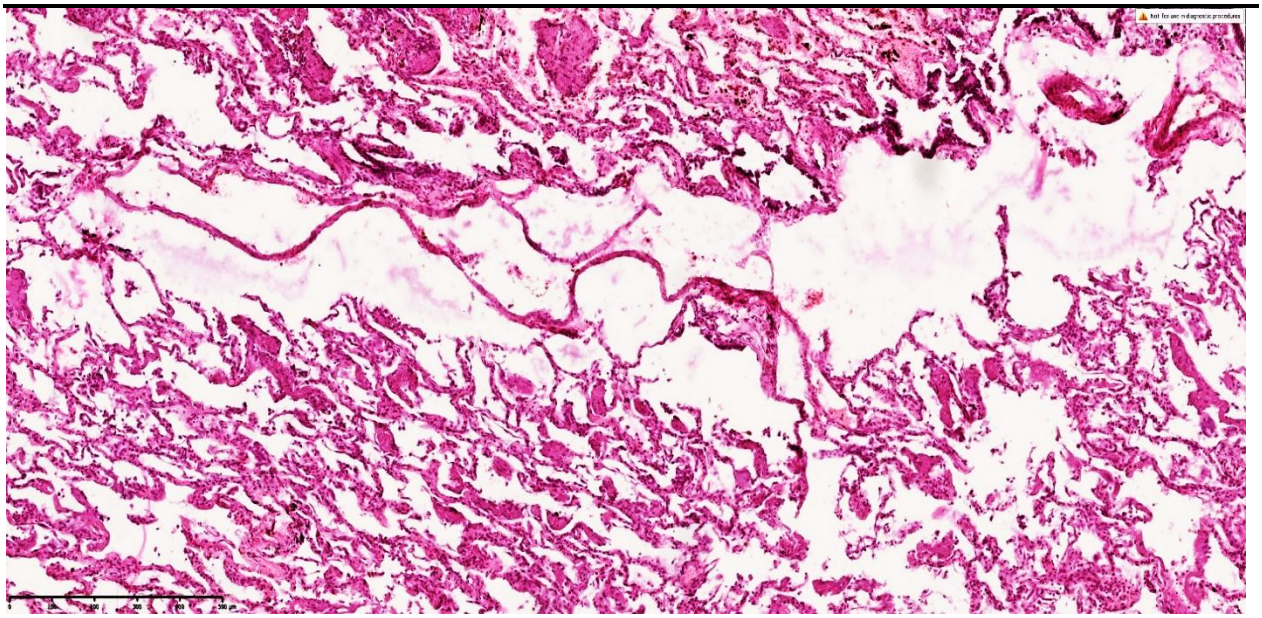

Figure 2: In bronchioloectatic Leschke emphysema, segmental bronchiolar dilatation is distinctly depicted, with a developing centrilobular cystic focus identified (1). Atelectatic foci are present in the alveolar spaces at the periphery (2). Sclerotic and atrophic changes are observed in the walls of small bronchioles (3). At the perimeter of the cystic dilated foci, developing inflammatory exudate and signs of bronchopneumonia are evident. Staining: H&E. Magnification: 10×10.

Certainly, in bronchioloectatic Leschke emphysema, besides the cystically dilated centrilobular areas within bronchiolar dilatation foci, markedly dilated bronchiolar segments and severe capillary congestion in the alveolar wall vessels are consistently observed as components of inflammatory foci resembling bronchopneumonia. Additionally, atelectasis is constantly found around the cystically dilated alveolar spaces. In our study, 22.1% of cases exhibited hyaline membrane formation, indicating clinical and morphological signs of respiratory distress syndrome (RDS) (see Figure 3).

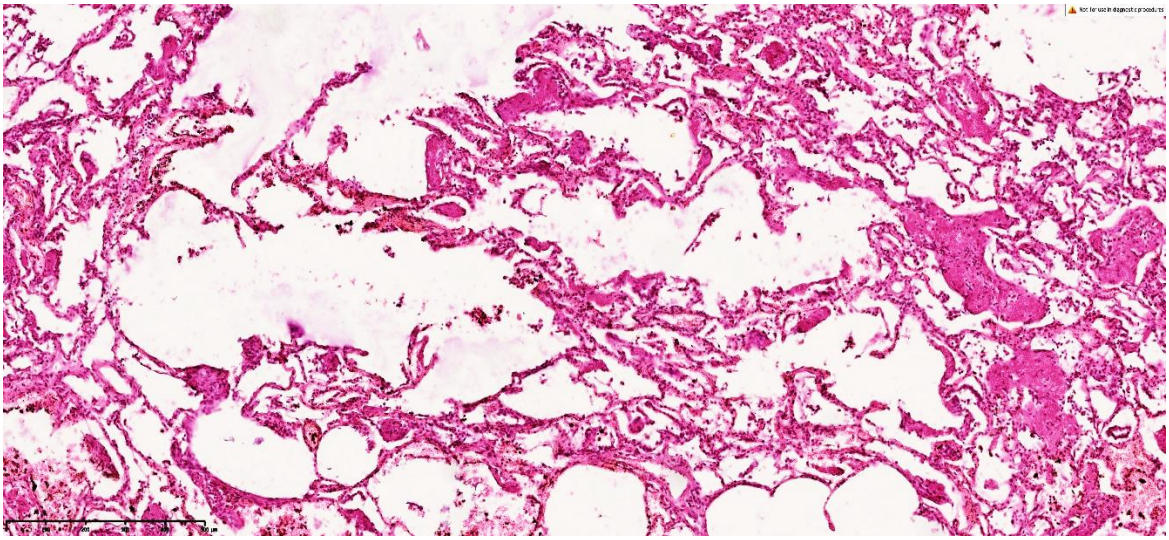

Figure 3: In bronchioloectatic Leschke emphysema, multicentrilobular cystically dilated foci are identified (1), with atelectatic foci present in the alveolar spaces at the periphery (2).

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Sclerotic areas are observed in the walls of small bronchioles (3). At the perimeter of the cystic dilated foci, developing inflammatory exudate and signs of bronchopneumonia are evident.

Staining: Hematoxylin-Eosin. Magnification: 10×10.

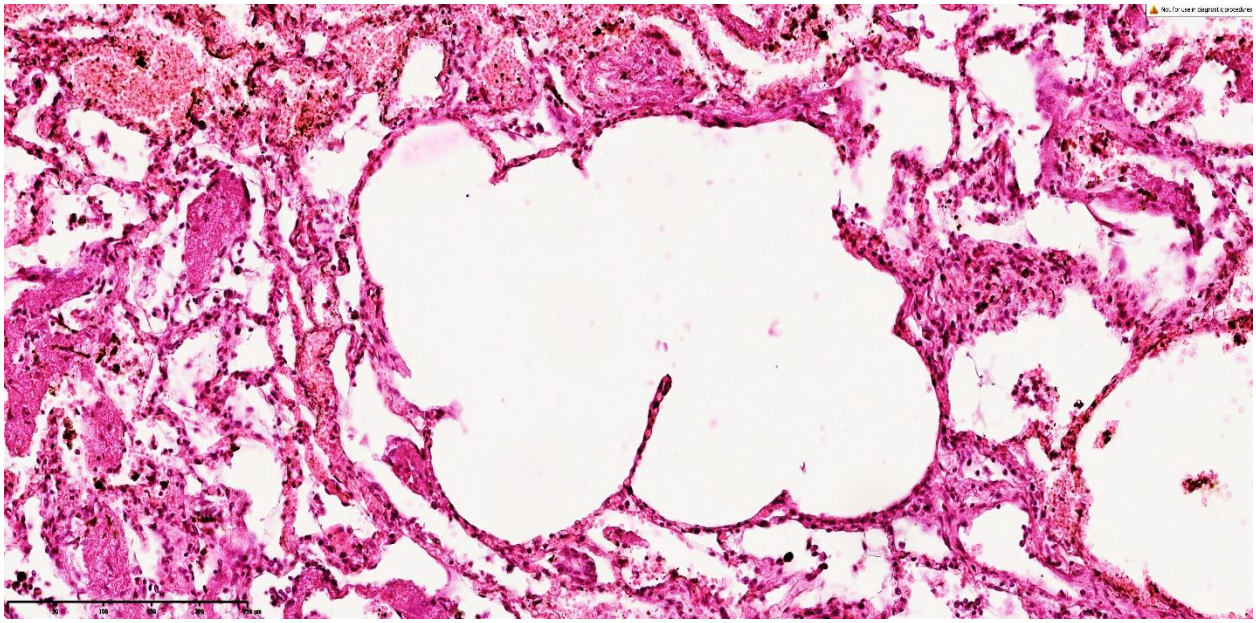

Figure 4: In bronchioloectatic Leschke emphysema, a centrilobular cystically dilated focus is identified (1), with atelectatic foci in the alveolar spaces at the periphery (2). Sclerotic and atrophic changes are observed in the walls of small bronchioles (3). At the perimeter of the cystic dilated focus, developing inflammatory exudate and signs of bronchopneumonia are evident.

Staining: H&E. Magnification: 10×10.

Thus, based on the above, the most characteristic morphological features of Leschke emphysema are primarily bronchial wall bronchiectasis. Clinically and morphologically, when patients develop polysegmental bronchopneumonia, usually between the ages of 12 to 15, complications such as cor pulmonale, pulmonary hypertension in the small circulation, and cardiac asthma occur. Microscopically, numerous respiratory bronchioles in the centrilobular areas undergo cystic emphysema. Due to severe congestion in the blood vessels, various degrees of developing polysegmental bronchopneumonia, atrophic and sclerotic changes in the bronchiolar walls, inflammatory exudates of varying severity, and desquamation foci in the mucous membrane of small-caliber bronchioles are observed. Meanwhile, no significant pathological changes are seen in medium and large caliber bronchi.

This indicates, from the perspective of its specificity, that the muscle layer hypoplasia and aplasia of the small-caliber bronchiolar walls occur, which is confirmed by the presence of most centrilobular cystically dilated foci. The aim of our study is to investigate the morphological features of each bronchial developmental anomaly, and based on the discussion of the above morphological examination results, to develop practical recommendations.

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Tracheoesophageal fistula, bronchial and tracheal stenosis mainly occur due to various influencing factors during the 9th to 12th weeks of intrauterine development of the esophagus and trachea. During the separation of these tubes, the integrity may be preserved or interrupted at varying degrees, resulting in fistulas or atresias forming proximally, medially, or distally. Clinically, newborns manifest various signs within the first minutes of birth, which pose a threat to life. Mortality rates are approximately 34% within 24 hours, 61.3% after 24 hours, and exceed 75% by the 3rd day, with rates of 90% or higher by days 4-5. One of the main features is the development of aspiration pneumonia and aspiration syndrome.

Morphologically, the boundaries of tracheoesophageal fistulas are characterized by areas where stratified and simple prismatic cuboidal epithelia meet, with zones of metaplasia. The esophageal contents may include gastric fluid and maternal milk residues. Residual aspirated material appears in the tracheal wall's inner surface, bronchial and bronchiolar lumens as a homogeneous foreign substance, demonstrating strong eosinophilic staining. The mucous membrane of the trachea shows varying degrees of hypoplasia of the ciliated epithelium, with underdevelopment of the submucosal layer and stromal tissue, including variably sized small glandular alveoli, indicating developmental delays and morphological anomalies (see Figure 5).

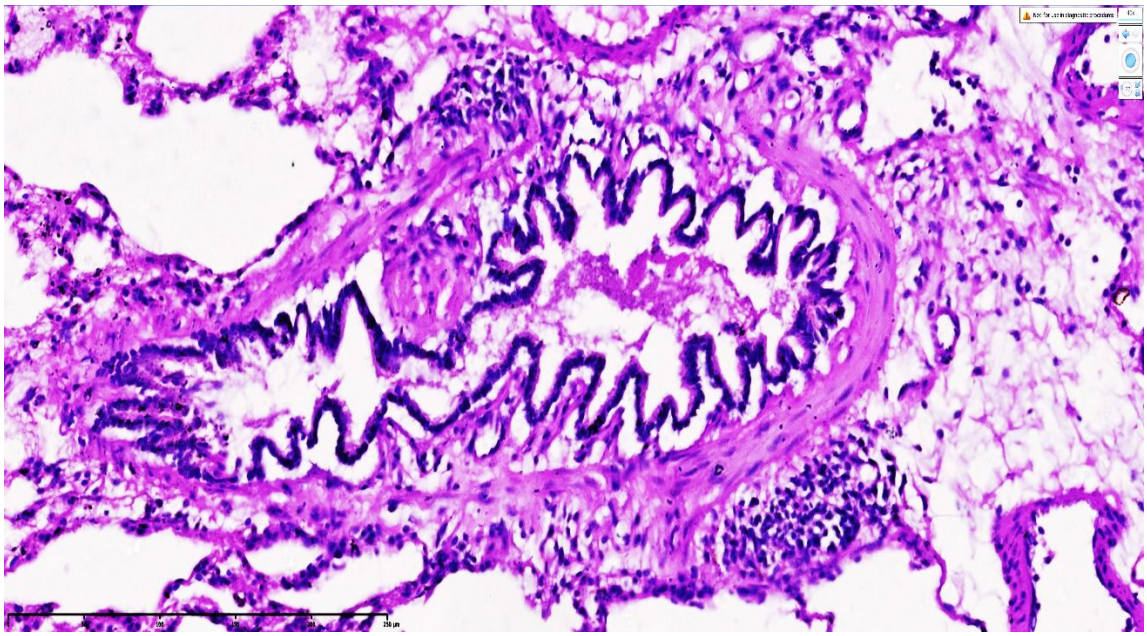

Figure 5. Hypoplasia of the tracheobronchial tree (TBT) structure, with numerous sclerosis and fibrosis areas in the peribronchial regions, and the bronchial wall exhibiting a homogeneous uniform structure. Staining: H&E. Magnification: 10x10..

Specifically, the ciliated epithelium within the mucous membrane appears as low cuboidal in shape, with very poorly developed cilia; there is vascular congestion and perivascular edema present. The hypoplasia of the tracheobronchial tree (TBT) structure in the mucous membrane, along with the absence of germinative centers, is observed. The connective tissue in the mucosa is not composed of sparse fibers but rather consists of coarse, mixed fibers arranged chaotically, which characterizes the deformity of the bronchi from a clinicomorphological perspective.

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In cases of tracheoesophageal fistulas, the mucosal layer of the bronchial walls shows delayed development, with epithelium that is smaller in height and volume, and a reduced number of intraepithelial lymphocytes of various types. Persistent vascular congestion is observed in small blood vessels within the interstitium, along with interstitial edema, an increase in sparse and coarse fibrous structures, and proliferative foci of fibroblasts and histiocytes. The submucosal layer is thickened beyond normal, and the glandular alveolar structure appears diffusely expanded. Massive foci of epithelial metaplasia and focal erosive lesions in areas of epithelial migration are also present (see Figure 6).

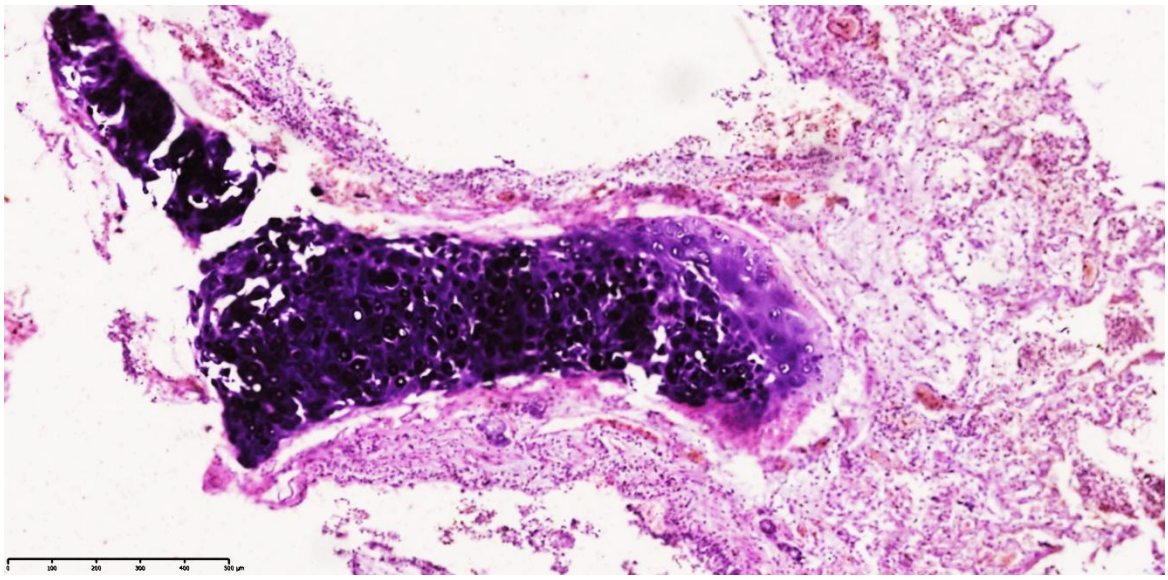

Figure 6. Fibromatous foci and areas of desquamation with scarring between all layers of the bronchial wall and the mucosal layer (1); inflammatory infiltrates and interstitial edema of varying degrees (2); erosive lesions on the epithelial surface of the mucous membrane (3). Staining: H&E. Magnification: 40x10.

As a result of mucoid degeneration in the submucosal layer, fibrillar structures become fragmented. There is also hypoplasia of the connective tissue in the walls of the trachea and bronchi, accompanied by a reduction in the size of chondrocytes from a morphofunctional perspective. The cytoplasm of these chondrocytes stains as clear vacuoles, with a deficiency of intercellular matrix around them and an increased amount of acidic mucopolysaccharides. In small-caliber bronchioles, a sharp increase in Clara cells indicates a significant proliferation of nonspecific inflammatory foci. The majority of changes are characterized by hypoplasia of the mucosa, submucosa, and muscular layers. Variability in the size of the smooth muscle cell population in the muscular layer reflects reduced contractility of the bronchial walls. Similarly, the connective tissue also appears as a sparsely cellular structure with persistent vascular congestion, formation of interstitial edema, and ongoing plasma exudation in the interstitial tissue. Regarding mortality, bronchospasm and bronchiolospasm are manifested by twisting of the bronchial mucosa, irregular trajectories of the respiratory bronchiolar mucosa, presence of various desquamated cells within the lumen, tissue fluid accumulation, and various homogeneous

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proteinaceous substrates. These changes contribute to the development of pneumopathy or secondary polysegmental bronchopneumonia in infants during the early neonatal period, often culminating in pulmonary edema. Therefore, recognizing the distinctive features of these bronchial developmental anomalies facilitates early neonatal diagnosis, prognosis, and reduction of mortality rates.

Conclusion

The study results highlight the significant morphological alterations in the development of bronchioles and bronchi, particularly in Leschke bronchioloectatic emphysema and tracheoesophageal anomalies. The hypoplasia and aplasia of the muscular layer of bronchiolar walls, cystic dilated centrilobular areas, desquamation in the bronchiolar mucosa, and inflammatory exudates contribute to the formation of polysegmental bronchopneumonia. Clinically and morphologically, these changes are associated with the development of pulmonary hypertension and cor pulmonale.

Developmental anomalies of the trachea and esophagus, including fistulas and stenoses, play a crucial role in upper airway pathologies, especially in the pathogenesis of aspiration syndrome and aspiration pneumonia. Morphological analysis revealed epithelial hypoplasia, metaplasia, lesions, as well as fibrosis and edema in the connective tissue of the lamina propria, indicating structural disruption of the bronchial and tracheal walls. These findings help explain respiratory dysfunction and the high neonatal mortality linked with these conditions.

Hypoplasia and aplasia of the muscular layer and connective tissue, along with capillary congestion and interstitial edema, contribute to bronchospasm and bronchiolospasm, resulting in significant impairment of pulmonary function. Additionally, the proliferation of Clara cells in small bronchioles reflects nonspecific inflammatory processes and microscopic pulmonary deterioration.

The results provide a vital basis for early diagnosis and morphologically guided preventive strategies targeting bronchial and bronchiolar developmental anomalies in clinical practice. Timely surgical interventions such as lobectomy and pneumonectomy are recommended to prevent polysegmental bronchopneumonia, pulmonary hypertension, and cor pulmonale, thereby reducing neonatal mortality rates.

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