Dendriform pulmonary ossification (DPO) is a rare diffuse bone formation in lung parenchyma resulting from chronic lung pathology with underlying different etiology. It has been described mostly in older age groups, but this case report depicts DPO in a pediatric patient following complex cardiac surgery.

CASE PRESENTATION

A 2-year-old male patient was born with dextrocardia, double-outlet right ventricle, pulmonary atresia, malposed great arteries, atrioventricular and ventriculoarterial discordance, inlet ventricular septal defect, and Wolff–Parkinson–White syndrome. After systemic-pulmonary artery shunt during the neonatal period and then bidirectional Glenn operation at the fifth month of age, he underwent a double-switch operation to establish one-and-a-half ventricle physiology. This repair involved (1) a Rastelli-type intracardiac baffle to close his ventricular septal defect and reroute the left intraventricular flow to the aorta; (2) a hemi-Mustard–type atrial switch repair to redirect systemic venous flow to the tricuspid valve; and (3) a right ventricular–pulmonary artery conduit. Early postoperatively, his course was complicated with ventricular diastolic dysfunction and on postoperative day 1, and his worsening hemodynamics required initiation of extracorporeal life support (ECLS). After a week of ECLS, his lung compliance and function started declining. Serial radiographs of the chest demonstrated recurrent lung collapse and development of focal-to-diffuse radiopacity (Figure 1). Subsequently, copious endotracheal secretions and recurrent lung collapse required serial bronchoscopy, which demonstrated recurrent formation of thick mucus plugs and early formation of bronchial casts. During the third and fourth postoperative week, the patient underwent 2 consecutive computed tomography scans of the chest, which demonstrated worsening severe bilateral pulmonary consolidation and atelectasis with diffuse parenchymal high attenuation and high-density material (Figure 2). Even with these diagnostic tests and ongoing management, lung pathology was not fully understood, and lung function progressively worsened. He continued to require ECLS, and during the seventh postoperative week, he died due to a large intracranial hemorrhage.

An autopsy was conducted to explain the rapid progressive and irreversible lung pathology. It showed the intact cardiac repair and unobstructed intracardiac baffle pathways. Microscopically, cardiac tissues were well correlated with the defect and multiple surgeries, including ventricular hypertrophy, fibrinous pericarditis, and dystrophic calcification. Lung autopsy demonstrated diffuse DPO, without bone marrow.
elements, involving alveolar interstitial walls and ducts, bronchioles, and bronchi. Small pulmonary vessels of both lungs revealed focal osseous metaplasia (Figure 3).

DISCUSSION

Pulmonary calcification is a common finding seen incidentally on chest imaging in otherwise-asymptomatic patients; however, pulmonary ossification is a rare metaplastic process characterized by the histologic presence of mature bone in pulmonary parenchyma. DPO is a chronic lung disease with extensive parenchymal involvement. There are 2 varieties based on gross histologic pattern: nodular and dendriform ossification. DPO, the rarest type, is almost always an autopsy diagnosis. It is most seen in the setting of long-standing inflammation such as interstitial fibrosis, but sometimes it may be idiopathic in nature. In the literature, all reported cases of dendriform ossification are seen in deceased individuals ranging from 50 to 69 years of age. No cases have ever been reported in the pediatric population.

While the exact pathogenesis of DPO is unknown, the metaplastic process is presumed to occur due to chronic hypoxia resulting in a state of cellular acidosis. DPO usually demonstrates fat or marrow element with bony proliferation in the pulmonary parenchyma. Radiographically, DPO has a bilateral distribution and is predominantly seen with reticulonodular densities. High-resolution computed tomography demonstrates radiopaque densities in a peculiar branching tree pattern. Retrospectively, DPO can be a poor prognosticator if diagnosed in a patient who is alive, as most of these

FIGURE 1. Radiograph of the chest. A, Preoperatively, showing clear lung field; B, immediate postoperative, showing clear lung field and radiopaque mediastinal gauze pack; C, postoperative day 1, post-ECLS, showing clear lung field. D, Second week postoperatively, E, fourth week postoperatively, and F, seventh week postoperatively showing progressive increase in lung opacity. ECLS, Extracorporeal life support.
ossifications are resilient and do not improve with any treatment modality. However, DPO has diagnostic significance and should be ruled out to prevent confusion with other forms of abnormal calcium deposition or granulomatous processes. These abnormal calcium deposits are not as grave in prognosis as dendriform ossification.

FIGURE 2. Cross-sectional (A and B), coronal (C), and sagittal (D) CT scans showing diffuse parenchymal high attenuation and high-density material in bilateral lung fields.

Case Report

References

