INTRODUCTION

In the past, the term known as bronchopulmonary dysplasia (BPD) has been a difficult disease to prevent and treat among premature infants and neonates. Bronchopulmonary dysplasia is characterized pathologically by acute, reparative, and chronic stages of lung injury caused by oxygen toxicity and barotrauma for respiratory distress. Clinically, the degree of BPD is based on gestational age and graded into mild, moderate, or severe. Changes in the science of assisted ventilation and medical treatment have reduced the morbidity and mortality of bronchopulmonary dysplasia. The current autopsy report suggests that BPD is still a pathologic entity that can present challenges in clinical management.

STUDY METHODS

Autopsy case report and a review of the English literature concerning the epidemiology and pathology of bronchopulmonary dysplasia.

CLINICAL PRESENTATION

A 21 year old Caucasian woman, who had died the current pregnancy in the local emergency department with previous rupture of membranes and delivered at 31 5/7 weeks female infant by spontaneous vaginal delivery. The mother's previous labors were normal and uncomplicated. She had a chronic asymptomatic infection in the chest and a history of a thyroidectomy at age 17. The infant was delivered by C-section on G4P3 at 36 weeks and 4 days. The initial APGARs were 7 and 8 at one and five minutes respectively. The infant was treated with five days of phototherapy and has received a gradual wean. An echocardiogram at 3 weeks of age showed normal cardiac chambers, vessels, a thickened mitral valve, and a patent ductus arteriosus which later closed. The patient remained hospitalized on O2 for chronic lung disease and was discharged at age 3 months. Oxygen supplementation and ventilation was initiated and were positive for Methicillin sensitive Staphylococcus aureus and Klebsiella pneumoniae which were treated with intravenous antibiotics. The infants bronchopulmonary dysplasia and increased airway secretions and bronchitis. A pneumotachograph developed secondary to gastric perforation and an exploratory celiotomy was performed on day 81. The patient experienced respiratory arrest and died in the immediate post-operative period despite resuscitative efforts. An autopsy was requested.

Autopsy findings included an intact surgical gastrostomy site and pale skin. The lungs were normally lobulated and had multiple dilated airways, measuring up to 3.9 mm (Figure 1). Areas of normal appearing lung tissue had fibrosis consolidation and thickening of the septal interstitium (Figure 3) and thickening of pulmonary arterial walls (Figure 4). These findings are consistent with bronchopulmonary dysplasia. There was fibrosis of the airways, the alveoli, the capillaries, and the lungs. The lungs had microscopic findings of focal hemorrhage, fibrosis, and dystrophic changes in the periosteal and medullary oblongata. The cause of death was attributed to multi-system failure secondary to bronchopulmonary dysplasia.

DISCUSSION

Bronchopulmonary dysplasia, also known as chronic lung disease, is a pathologic disease entity that has considerable, constant morbidity and mortality despite improvements in mechanical ventilation and surfactant treatment. A recent definition of BPD, provided by the National Institute of Child Health and Human Development on BPD defines it as the need for oxygen supplementation at 28 days of life, yet separates this into different grades depending on the need for oxygen at 28 weeks PMA (postmenstrual age).

In 1969, Wilson and Minkly described a new form of respiratory disease of unknown cause, later known as Wilson-Minkly Syndrome, in prematurely infants occurring between weeks 5 and 50 of birth, not receiving mechanical ventilation, and having radiographic changes of severe fibrosis with areas of emphysema. Subsequently, in 1967, Northway and colleagues introduced the term bronchopulmonary dysplasia to describe a chronic lung disease occurring in premature infants receiving mechanical ventilation and supplemental oxygen who developed radiographic abnormalities indistinguishable from Wilson-Minkly Syndrome. The clinical presentation of prematurity-related bronchopulmonary dysplasia includes symptoms of airway obstruction or dysfunction, difficulty in oxygenation and the need for assisted ventilation due to respiratory distress. The symptoms are typically seen from the first few days of life and persist for more than 30 post-natal months. Newborns with bronchopulmonary dysplasia may have increased airway secretions and airway hyperreactivity.

The pathogenesis of BPD is multifactorial with inflammation playing a key role, along with surfactant deficiency of prematurity, hypoxygenia, hamartoma, and nutritional deficiencies. It has been noted, due to changes in treatment modalities and the differences in the blood oxygen pressure between "new" BPD compared with "old," specifically, reductions in the size and number of blood vessels. The pathology has been described in these stages consisting of an early, chronic stage which last from 3 to 30 days consists of bronchial and bronchomascular damage and airway developing a fibroproliferative process, and the interstitium being edematous with fibrosis and smooth muscle proliferation. The second stage, a chronic, fibrotic stage, BPD is defined as lasting at the end of the first week. The infant may demonstrate signs of interstitial fibrosis at the most severe third stage. Chronic BPD has a long-standing airway and vascular damage that is such as hyperplasia of the medial smooth muscle layer and an increase in adventitial fibrous tissue, which is seen grossly and microscopically in the current case. The epidemicology varies from institution to institution and increases with prematurity. The role of mortality is associated with the duration of ventilation. In a report spanning 1985-1992 at a single institution, of 47 infants on MV for > 27 days, 20 died (43 mortality). In a study from the national institute of child health, 10% of the infants ventilated for at least 2 months or more than 4 months, respectively. An 8 year cohort study of BPD at six centers, from 1994 to 2002, demonstrated an overall incidence ranging from 12.6% to 21.7%. The multi-center was 14.8% of the population. This study found an overall incidence of BPD at 23.3% among 14.4% very low birth weight infants and this incidence increased with decreasing birth weight. Most infants with BPD will improve over several months and are weaned to CPAP and then supplemental oxygen. Infants with severe BPD may have ventilator dependence and may develop pulmonary hypertension and cor pulmonale. A recent development is that severe BPD can lead to neurodevelopmental risk concerns. The development of ventilator treatment and surfactant therapy, very low birth weight infants are surviving and the prevalence of BPD has remained fairly constant through the clinical picture as evolved.

CONCLUSION

Morbidity and mortality due to bronchopulmonary dysplasia has improved with mechanical ventilation and surfactant therapy, yet the current case report demonstrates the challenges in caring for these patients.