early afterdepolarizations. In cellular preparations, as well as with monophasic action potential recordings, magnesium administration has been reported to suppress such activity.6

In conclusion, proarrhythmic actions may occur with use of amiodarone. Should polymorphous ventricular tachycardia develop in association with its use, the drug should be discontinued, electrolyte imbalances should be corrected, and pacing should be considered if necessary. Concomitantly, an infusion of intravenous magnesium sulfate should be administered until an adequate period of time has passed.

REFERENCES

Recurrent Alveolar Proteinosis Following Double Lung Transplantation*

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We present a case of recurrent alveolar proteinosis following double lung transplantation.

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Key words: alveolar proteinosis; lung transplantation

Abbreviations: PAP=pulmonary alveolar proteinosis

Pulmonary alveolar proteinosis (PAP) is a disorder characterized by excessive accumulation of surfactant-like material within the alveoli. Traditional management has been with bronchopulmonary lavage as needed for symptomatic relief. Progression to pulmonary fibrosis is rare.1 We present a case of a patient with PAP who progressed to pulmonary fibrosis (end-stage lung disease) treated with double lung transplantation who developed recurrent PAP in the transplanted lungs within 3 years of transplantation.

CASE REPORT

A 41-year-old woman was evaluated for lung transplantation because of end-stage lung secondary to PAP (Fig 1). Her disease had been diagnosed at age 27 years, and she had been treated with bronchopulmonary lavage (12 episodes) with initial good results but had a slowly declining course. Then, 8 years after diagnosis, she was referred to a major transplantation center for possible heart-lung transplantation. However, she experienced a spontaneous regression of symptoms and was not evaluated further. Approximately 6 years later, she was referred to our institution with worsening symptoms. On presentation, her FVC was 30% of predicted, her FEV1 was 35% of predicted, and her arterial blood oxygen saturation was 81% while breathing room air. During the ensuing 7 months, her condition steadily deteriorated and she underwent double lung transplantation. Postoperatively she did well. Routine follow-up was with chest radiography, pulmonary function tests, and surveillance bronchoscopic evaluation with transbronchial biopsy. Over the next 2 years, she experienced periodic episodes of bronchitis and developed mild obliterator bronchiolitis with concomitant decrease in pulmonary performance.

A little over 3 years after transplantation, she had a transbronchial biopsy of the right upper lobe to evaluate a new infiltrate seen on the chest radiograph (Fig 2). Specimens were negative for rejection or infection with fungus or a mycobacterial species. She returned in 2 months complaining of increasing fatigue and dyspnea on exertion. Another BAL and transbronchial biopsy were performed.

Histologic sections from the transbronchial lung biopsy re-
diagnostically, it is characterized by a symmetric pattern of alveolar filling bilaterally that is similar in appearance to pulmonary edema. Early in the disease process, discrete acinar shadows may be apparent that become more confluent and eventually lead to diffuse areas of parenchymal consolidation. High-resolution computed tomography has been used to demonstrate the interstitial involvement that is usually not apparent on the chest radiograph. Rarely, the process may progress to radiographically apparent interstitial involvement. However, progression to severe pulmonary fibrosis, or “end-stage lung disease,” is distinctly uncommon and is one of the interesting features of this case.

The cause of alveolar proteinosis has been attributed to various factors including inhaled agents, altered immunity, or locally produced toxic agents. Overproduction of surfactant-like material by the type 2 pneumocytes was considered to be the source of the lipoproteinaceous material filling the alveoli. Recent work has shown that it is more likely to be the result of defective clearance of the material by alveolar macrophages and this can be shown experimentally in laboratory animals. This case is of interest not only because of the unusual course of disease leading to pulmonary fibrosis, but also because of the recurrence of the disease in the transplanted lung in the absence of exposure to a toxic agent supports the theory of reduced alveolar clearance secondary to diminished macrophage activity. It may well be that the underlying defect exists in circulating monocytes. Experimentally, macrophage dysfunction can be induced in normal human monocytes after exposure to the lipoproteinaceous material that fills the alveoli. More work needs to be done to elucidate the fundamental cellular alteration responsible. Radiologists and pulmonologists need to be aware that this entity may rarely progress to pulmonary fibrosis. Physicians involved in lung transplantation programs should be informed that PAP can recur in the transplanted lungs and may be heralded by the appearance of small rounded opacities seen on the chest radiograph (Fig 3).

REFERENCES

FIGURE 2. Diffuse involvement of both lungs with small rounded opacities representing airspace filling with lipoproteinaceous material.

FIGURE 3. Histologic section reveals an alveolus filled with granular and flocculent material (large arrows) and lined by reactive type 2 pneumocytes (small arrows) (hematoxylin-eosin, original X400).