RETROSPECTIVE INSTITUTIONAL STUDY OF 38 PATIENTS TREATED FOR PULMONARY ARTERY SARCOMA

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ABSTRACT

Objective:

To determine the optimal surgical procedure to treat pulmonary artery sarcomas responsible for pulmonary hypertension.

Methods:

Between 1997 and 2014, 38 patients were surgically treated for pulmonary artery sarcomas. Twenty-one patients were male; the mean age was 56 years (range, 26 to 78). Common symptoms were the ones of acute or chronic pulmonary thromboembolic disease. Also, 21 patients experienced mild to severe pulmonary hypertension with a mean TPR of 473 Dyne/sec/cm^{-5}. Clinical presentation and preoperative work-up confirmed the suspicion of pulmonary artery sarcoma in 18 patients. The required surgical procedures included: pulmonary endarterectomy in 32 patients (combined with a right pneumonectomy in 5 and with a replacement of the main pulmonary artery by a homograft reconstruction in 1), pneumonectomy only in 5 (3 right and 2 left), with the use of cardiopulmonary bypass in 3 cases. In one patient, the right pulmonary artery only was replaced on cardiopulmonary bypass.

Results

Final pathology showed 33 high-grade and 5 intermediate grade sarcomas. Thirty day mortality was 13% (5 patients). Repeat pulmonary resection was required in 2 patients due to recurrent disease. Moreover, 18 patients received adjuvant therapy. Mean follow-up was 19

months (range, 1 to 99); of the 11 patients alive at follow-up, 4 were noted to have recurrent disease. 1, 3 and 5-year survival was 63%, 29% and 22% respectively.

Conclusion:

The prognosis of this orphan disease remains poor. Bilateral pulmonary endarterectomy may yield significant survival rates because it warrants completeness of resection without sacrificing the pulmonary vascular bed. Heart-lung transplantation has to be discussed in young patients with neither extrathoracic nor pleural disease.

KEYWORDS

Sarcoma -pulmonary arterial hypertension - pulmonary thromboendarterectomy- cancer