



Pulmonary Artery Sarcomas Presenting as Pulmonary Hypertension: A review and discussion of further treatment.

Hatcher HM, Patel N², Gounnaris I, Pepkeza J², Jenkins D², Sherbourne K, Horan G, Earl HM.

Sarcoma Team, Cambridge University Hospitals Trust, Cambridge, CB2 0QQ and ²Pulmonary Hypertension Service, Papworth University Hospitals Trust, Papworth, CB23 3RE

Background:

Pulmonary hypertension is a rare disorder which leads to progressive right heart failure and death without treatment. Pulmonary thromboendarterectomy (PTE) has improved survival for patients with thromboembolic pulmonary hypertension. However during the process of investigation patients with possible thromboembolic pulmonary hypertension several patients were found to have primary pulmonary artery sarcomas. Due to their rarity there is no consensus on treatment but we report the findings of the largest case series.

Aims:

To understand the scale of primary pulmonary artery sarcomas, the presentation and review treatment options.

Methods:

Review treatment and outcomes of all pulmonary artery sarcomas referred to the pulmonary hypertension service or the local sarcoma team from 1999-2009.

Results:

Over the 10 years over 500 PTE surgeries were performed. 15 patients (9 male, 6 female) were identified with pulmonary artery sarcoma, of whom 10 had surgery. 13 of these were identified by the pulmonary hypertension service and 2 referred directly to oncology with a large mass arising from the pulmonary artery detected on CT. Of the 10 patients that had surgery 3 are alive a median of 28 months (range 13-44 months), 2 of whom had chemotherapy and radiotherapy despite residual right heart failure after surgery. 3 patients died shortly after surgery and 1 patient died of unrelated disease 18 months after surgery. Of the 5 not having surgery 3 had other treatment (2 had chemotherapy and radiotherapy and 1 had radiotherapy alone). 4 of the patients had anthracyclines and 3 had ifosfamide.

Conclusions:

Postoperative treatment with chemotherapy (+/- radiotherapy) was associated with improved outcome despite ongoing right heart failure in some patients or concerns about pulmonary artery rupture. Chemotherapy and /or radiotherapy also improved symptoms for those with inoperable disease.