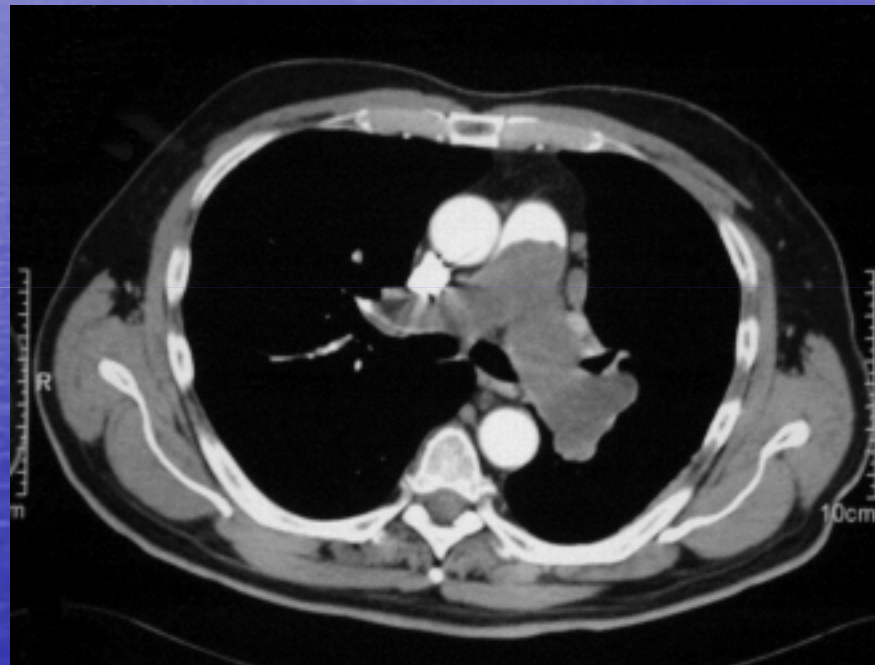


Pulmonary artery sarcomas presenting as Pulmonary hypertension



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Pulmonary Hypertension (PH)

- Rare (2-3 per million per year)
- Increased pulmonary artery pressures
- Presents with progressive breathlessness and signs of right heart failure
- Thrombo-embolic disease most common cause
- Delays in diagnosis common (median 1 year delay)
- Treatment is with anticoagulation and surgery (PTE)



Surgery for Thromboembolic PH

- Pulmonary thrombo-endarterectomy (PTE)
- Cooling of body to 18 degrees Celsius
- Cardiopulmonary bypass
- Segmental dissection of laminated thrombus



Pulmonary artery sarcomas

- Rare (< 250 in literature)
- Similar presentation to pulmonary hypertension
- May be discovered at PTE
- No treatment consensus



Methods

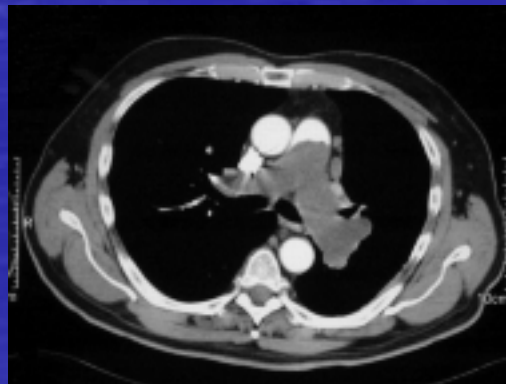
- Retrospective review of patients referred for PTE or oncology with pulmonary artery sarcoma from 1999-2009
- Information was obtained on
 - Patient characteristics
 - Surgery
 - Chemotherapy
 - Radiotherapy
 - Survival

Results

- Patients with PH referred who had PTE = 500
- Patients with pulmonary artery sarcoma = 15
- Male = 9
- Female = 6
- Median age = 48 years (26-76)
- Metastases at presentation = 4

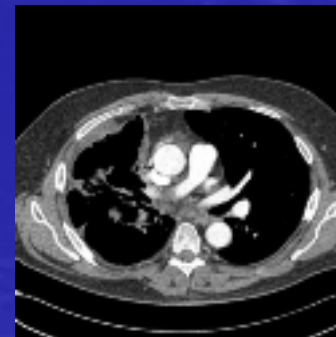
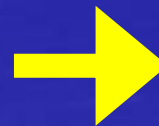
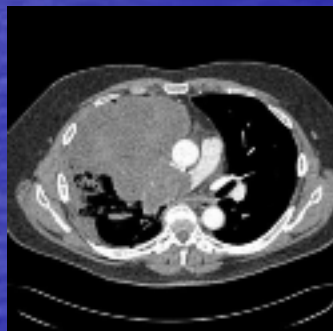
Presenting features

- Breathlessness 14
- Heart failure NYHC II-IV 12
- Duration of symptoms 6-48 months
- Suspected malignancy 7



Treatment

- Surgery N = 10
 - PTE
- Radiotherapy N = 5
 - Radical = 2
 - Palliative = 3
- Chemotherapy N = 6
 - Adjuvant = 3
 - Palliative = 3

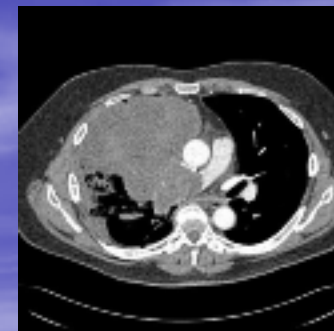


Survival

- Median overall survival (surgery) 28 months (0-46)
- Median overall survival (no surgery) 8 months (0-16)
- Suggestion that surgery + chemotherapy + radiotherapy had improved survival but too few patients
- Generally treatment with 2+ modalities better than 1



Summary



- Pulmonary artery sarcomas are rare
- Presentation is similar to pulmonary hypertension
- Some features suggest sarcoma rather than emboli
 - Rapid progression of symptoms
 - Central occlusion on CT
 - Failure of resolution after anti-coagulation
- Surgery and probably multi modality treatment improves survival