CASE REPORT

Status postpneumonectomy for pulmonary artery sarcoma

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SUMMARY

Primary pulmonary artery sarcoma is a rare disease that has a poor survival prognosis due to misdiagnosis with pulmonary thromboembolism or metastatic embolisation, detailed image findings and complicated surgical procedures. Surgical procedures established for treatment include pneumonectomy and pulmonary endarterectomy. Survival after surgery still remains at 1 year survival of 50%. The following case report demonstrates a patient's status postpneumonectomy for pulmonary artery sarcoma that presented with no complications after 1.5-year follow-up.

BACKGROUND

Pulmonary artery sarcoma is a rare type of tumour that arises from the tunica media of the pulmonary artery.¹ Only about 300 cases have been reported since its first recognition in 1923.^{2,3} Presentation is similar to that of chronic pulmonary thromboembolism, presenting particularly with dyspnoea, chest pain, cough and haemoptysis. Clinical findings include pulmonary artery systolic flow murmur, hepatomegaly, oedema, cyanosis and digital clubbing.^{4,5} Imaging that has aided in diagnosis for pulmonary artery sarcomas includes CT and MRI with gadolinium.^{2,5}

Positron emission tomography with CT (PET-CT) is also established as a well-suited approach to evaluate pulmonary artery masses.⁶ Published treatment for pulmonary artery sarcoma includes pneumonectomy for curative treatment and pulmonary endarterectomy for a palliative approach.² Adjuvant chemotherapy has also been issued as a means of prolonging survival.⁷ Despite treatment measurements, 1 year prognosis for patients with primary pulmonary artery sarcoma is 50%.^{2,5} The following case describes a patient who had undergone pneumonectomy and reported no complications after 1 year.

CASE PRESENTATION

A 59-year-old man presented to his primary care physician clinic on December 2012 with coughing and occasional haemoptysis. Evaluation and work up by the team concluded that the patient had a possible pneumonia; he was treated with a 10-day course of moxifloxacin. However, on February 2013, his symptoms began to recur along with night-sweats, fevers reaching 102°F and unintentional 40-pound weight-loss. Another course of moxifloxacin was performed with no improvement in symptoms.

INVESTIGATIONS

CT scan showed a cavitary lesion on the right upper lobe of the lung. A transbronchial biopsy showed chronic active inflammation without signs of infection or metastatic cells. When evaluated again on May 2013, the patient reported localised right chest pain in addition to his previous symptoms. CT scan showed a mass in the right pulmonary artery suspicious for embolus versus neoplastic lesion. A PET-CT scan showed a hyperactive mass in the right pulmonary artery suggestive of an active neoplasm (figure 1). Owing to the concern of malignancy and worsening of symptoms, surgery was performed on May 2013.

TREATMENT

The surgical approach included a median sternotomy to reveal the heart. Circulatory arrest and use of cardiopulmonary bypass was performed in traditional manner. After exposing the right pulmonary artery, a longitudinal incision revealed a mass that extended toward the left pulmonary artery branch. Frozen section pathology revealed high-grade malignant intimal sarcoma with positive margins reaching the left pulmonary artery (T2b G3 N0 M0). Excision of the right pulmonary artery and Dacron graft replacement of the left pulmonary artery was performed. Later, a right pneumonectomy was performed. After the removal of the lung, the operating fields were closed and the patient was sent to the surgical intensive care unit.

OUTCOME AND FOLLOW-UP

The hospital course was complicated by atrial fibrillation with rapid ventricular response and an episode

Figure 1 Focal uptake correlates to the mass along the right pulmonary artery (white arrow), extending along the vessels towards the right hilum (SUVm=8.3). There is a small focus in the right hilar region. Hypermetabolic activity is suggestive of a neoplastic lesion rather than an embolus.

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of anaemia. The patient was brought back to the operating room due to suspicion of haemothorax. In the operating room, the patient was cardioverted electrically. Surgery was performed and 4 L of clotted blood was evacuated from the empty right chest. No other complications occurred throughout his hospital stay and the patient was discharged on postoperative day 9.

The patient was followed for 1.5 years. He underwent radiation and chemotherapy to ensure proper treatment. He currently does not report any deterioration in his health except a chronic, dry cough and occasional dyspnoea on exertion. He reports regaining normal weight, no fevers or chills and no chest pain or haemoptysis. CT scan of the chest shows no evidence suggestive of return of malignancy.

DISCUSSION

Pulmonary artery sarcoma presents similarly to a chronic thromboembolism of the pulmonary artery. Symptoms include dyspnoea, chest pain and haemoptysis.^{4 5} Signs are similar to that of cor pulmonale, including increased pressures in the right heart chambers.

Differentiating pulmonary artery sarcoma from a thromboembolism may be performed with contrast-enhanced CT or more precisely using MRI with gadolinium.^{2 4 6} Tumours will brighten with gadolinium while thromboembolisms will remain with no change in brightness. PET-CT scans also serve as another imaging modality that may distinguish tumours from thromboembolism.⁶ Echocardiography seldom detects sarcoma; however, it is capable of demonstrating pulmonary hypertension.²

Other clinical suspicions for an occluding mass rather than an embolus include a trial of anticoagulation and no medical history significant for deep vein thrombosis. With a poor response to anticoagulation therapy, sarcoma becomes highly suspected. Deep vein thrombosis is frequently involved with massive pulmonary embolisms; suggested by Well's criteria, absence of deep vein thrombosis lowers the probability of a pulmonary embolism. The rarity of a sarcoma increases as a more probable diagnosis.⁴

Currently, the literature disputes these presentations between thromboembolism versus primary pulmonary artery sarcoma. However, metastases or metastatic embolisation can also mimic these cases. Use of imaging may help determine whether the tumour mass is primary or of metastatic origin, but only surgical resection and histology provides a definitive diagnosis.⁸

CT scan findings that are suggestive of pulmonary artery sarcoma include filling defects occupying the entire luminal diameter of the proximal or main pulmonary artery, usually accompanied by extraluminal extension of the tumour; heterogeneous histological appearance, characterising the combination of necrosis, haemorrhage and ossification in the tumour; and unilaterally, as pulmonary thromboembolisms typically occur bilaterally.^{4 9} Two surgical approaches to treat primary pulmonary artery sarcoma include pulmonary endarterectomy and pneumonectomy. The case report presented demonstrates survival from pneumonectomy. Ahmed *et al*⁴ describe a case involving the pulmonary endarterectomy approach. Grazioli *et al* compared the two modalities based on their experience with patients. The study showed that although pneumonectomies tend to acquire more hospital complications compared to pulmonary endarterectomy patients, the survival after pneumonectomy averaged 26.8 months compared to pulmonary endarterectomy with 6.6 months; 12-month survival status postpneumonectomy versus pulmonary endarterectomy was listed at 67% and 37%, respectively. Therefore, they suggest

that pneumonectomies portray a survival benefit while pulmonary endarterectomies are favoured in high-risk patients requiring a shift to palliative treatment.² However, recognition of the extent of invasion of the primary sarcoma may determine the effectiveness of a pneumonectomy procedure; one must ascertain that the tumour is confined in one lung.⁹

Debate exists on whether adjuvant chemotherapy and radiotherapy should be utilised on afflicted patients. Mussot *et al*⁷ suggest that adjuvant therapy should be based on an individualised basis. Grazioli *et al*² further demonstrate that chemotherapy and radiation should be given to those who have adequate reserve; compromised clinical conditions contraindicate the use due to risk of premature death. Jamieson argues that adjuvant therapy should be employed in every patient for terms of palliation rather than a curative means. Future studies in DNA analysis for direct chemotherapy may improve the prognosis of pulmonary artery sarcoma.¹⁰

Learning points

- ▶ Primary pulmonary artery sarcoma is better treated with pneumonectomy rather than pulmonary endarterectomy. Pulmonary endarterectomy is better suited for palliative treatment.
- ▶ Pulmonary artery sarcoma presentation can be confused with pulmonary embolism. A Positron emission tomography with CT scan is a reliable imaging tool to differentiate between the two diseases.
- ▶ Despite treatment, pulmonary artery sarcoma tends to occur with 1 year mortality. However, as presented with this case, prolonged survival is still possible.
- ▶ Treatment with pneumonectomy plus postoperative chemotherapy presents a possible increase in survivability.

Contributors The following authors contributed as listed: JAS was primary author; AG was abstract author; and HT was tertiary author.

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