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Case Report

Extensive Thoracic Organ Resection for a Patient Who Harbors Spindle Cell Sarcoma

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Because of its rarity, there has been no standard treatment protocol for spindle cell sarcoma of the pericardium. Benefits of chemotherapy and radiation remain elusive. As a result of this, surgery is the mainstay of treatments. However, there was a phenomenal report of combination of chemotherapy, radiation and surgery treating a single patient who had survived at least 14 years after diagnosis[1]. Here, we reported a patient with non-metastatic spindle cell sarcoma of the pericardium who underwent aggressive thoracic surgery.

A 27-year-old female patient presented with pericardial effusion at the local hospital. Pericardial biopsy demonstrated spindle cell sarcoma. She received no further treatment and was then referred to the Cardiothoracic Surgical Unit, Department of Surgery, Ramathibodi Hospital due to increased shortness of breath. At the first visit, she was severely troubled by discomfort. Physical examination demonstrated diastolic rumbling murmur at the apex with increase S2 sound over the left upper parasternal border. Figures 1 and 2 showed a huge lobulated mass engulfing the pulmonary veins, the left bronchus and the descending thoracic aorta. It also externally compressed the left atrium. By knowing the natural history and her worsening symptoms, she decided to have palliative surgery with informed consent.

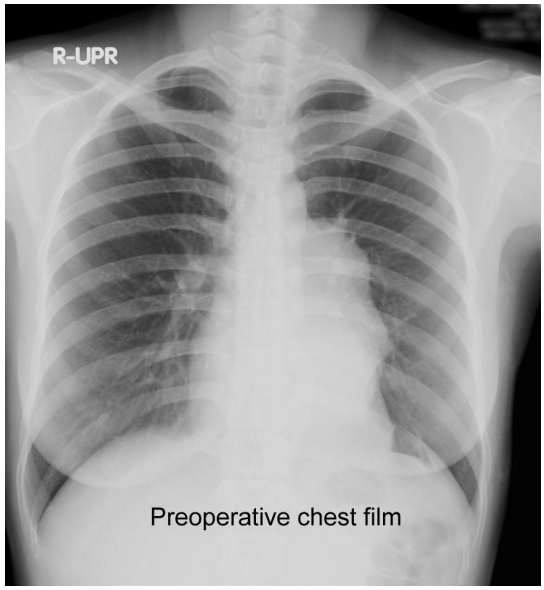

Figure 1 Preoperative chest x-ray showed a mediastinal mass without abnormal lung field.

Operative Procedure

The patient was in a supine position with a bump under her chest. With the cardiopulmonary bypass, she underwent left pneumonectomy first with details as follows: resection of left pulmonary artery, on cardiopulmonary bypass at 25 ˚C, cut the left main

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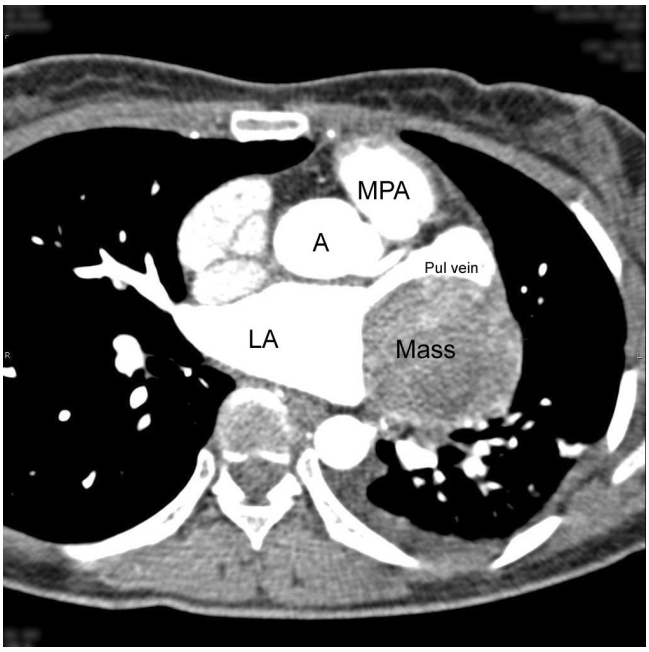

Figure 2 CT scan revealed the mediastinal mass. It adhered to the left atrium and compressed the left pulmonary vein.

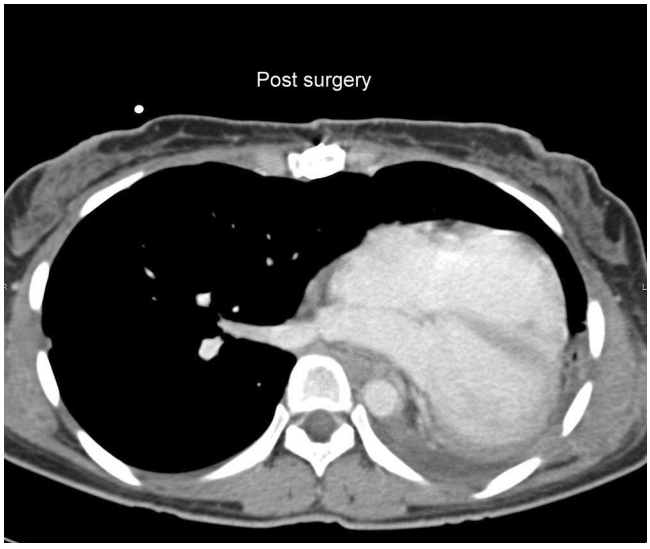

Figure 3 One week after surgery, CT scan showed thick tissue abutting the lateral border of the aorta. With some discussion, it was presumed to be a residual tumor.

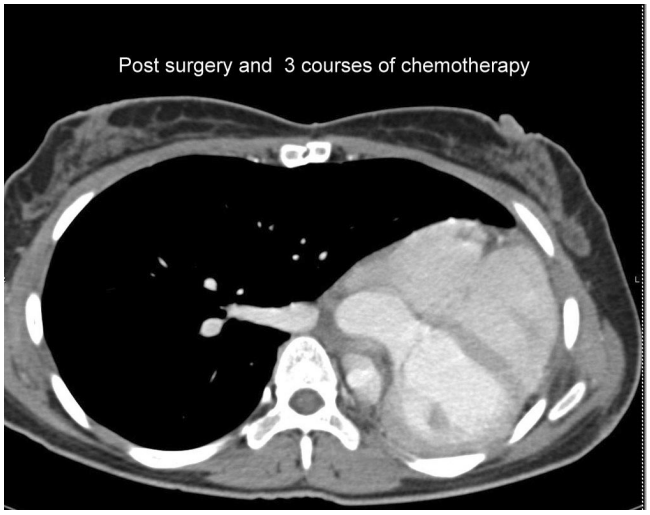

Figure 4 After 3 courses of chemotherapy, there was no previously identifiable mass along the descending aorta.

bronchus and then resection of left pulmonary veins *en bloc* with the posterior wall of the left atrium with cold blood antegrade cardioplegia. The left lung was then removed. The tumor adhering to the descending aorta was removed. Finally, the partially resected left atrium was reconstructed with a piece of bovine pericardium. Cardiopulmonary bypass time was 284 min and aortic clamp time was 145 min.

The patient came off from cardiopulmonary bypass without difficulty. However, there was a considerable amount of unidentifiable bleeding behind the heart. The mediastinum and the left chest were packed with swabs. Hemostasis was achieved only the first 4 hours. She was then transferred back to the operating room for re-sternotomy and left thoracotomy. Bleeding point was identified from the torn intercostals arteries. They were ligated and bleeding had stopped since then. Intraoperatively, the descending aorta was found free of tumor.

Hospital Course

She was transferred back to the ICU and was on mechanical ventilation for 24 hours. Extubation was successful; however, she developed junctional rhythm for 2 days and then returned to sinus rhythm. She also complained of palpitation when lying down and turning the left side down. This symptom disappeared after 2 weeks.

Follow Up

One week postoperatively, the CT chest was performed. Figure 3 demonstrated a 1.8 × 1.2 cm. mass behind the heart and lateral to the descending aorta. With intra-operative findings, the patient was presumed to have residual tumor; postoperative chemotherapy was planned. The final pathological

report revealed "poorly differentiated fibrosarcoma with pulmonary vein invasion".

Discussion

Sarcoma of the pericardium is a rare disease[1,2]. Moreover, response to chemotherapy is usually disappointing and unpredictable. Also, it is a radiation resistant tumor. Therefore, surgery seems to be the backbone of the treatment strategy. Most patients came to the hospital with compressive symptoms[3-9]: massive pericardial effusion, tumor tamponade or bulky tumor in the chest. If there is no metastasis, surgery is theoretically possible.

Our patient was young and previously healthy and most importantly non-metastatic case. With a curative intent, tumor involved organs that needed to be resected were left lung, pericardium, descending aorta and the left atrium. All of these organs were considered resectable or reconstructable i.e. the left atrium and the descending aorta could be reconstructed with pericardium and a piece of vascular graft respectively (although the descending aorta in this case was not resected as it was grossly free of tumor intraoperatively). More aggressive but possible method is resection of tumor ex vivo and autologous heart transplantation which has been reported with acceptable results[6-9].

Conclusion

Although removal of major thoracic organs is an extensive surgery, it is an alternative option for treatment of patients with large localized thoracic tumors that have poor response to chemotherapy and radiation.

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