Brief Communications

Malignant peripheral nerve sheath tumor of the mediastinum: A temporary aortic transection approach

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We report a case of mediastinal malignant peripheral nerve sheath tumor (MPNST) in a patient with neurofibromatosis type 1 (NF-1, von Recklinghausen disease). The tumor had a complex involvement with mediastinal structures and was resected by a temporary aortic transection approach under cardiopulmonary bypass (CPB).

Clinical Summary

A 40-year-old man with NF-1 was referred to our hospital because of hoarseness and enlargement of an abnormal shadow on his chest radiograph. On admission to our hospital, chest computed tomographic (CT) scan showed multiple masses in the mediastinum and chest wall. The largest one, measuring 12 × 9 × 8 cm, was centered in the aortopulmonary window, encasing the ascending aorta and compressing the right pulmonary artery, left atrium, and left main bronchus (Figure 1). Compared with his chest CT 4 years previously, this mediastinal lesion showed marked growth, whereas the other masses had remained unchanged in size. A CT-guided needle biopsy revealed MPNST immunohistochemically positive for S-100 protein. We initially chose systemic chemotherapy with cisplatin and doxorubicin to reduce tumor size, but chemotherapy proved ineffective. We then planned as a lifesaving measure to surgically resect the tumor to relieve its compression of vital structures. To best gain access to this complex lesion, we elected to temporarily transect the ascending aorta under CPB.

A median sternotomy and a left anteroaxillary thoracotomy through the fourth intercostal space were performed. An elastic, firm tumor was found densely attached to the ascending aorta, right pulmonary artery, and trachea. We exfoliated the tumor adhesion meticulously but could not proceed around the central right pulmonary artery because of tumor invasion into the vessel wall. At this time we observed the patient's blood pressure, heart rate, and arterial blood oxygenation dropping drastically in response to tumor manipulation. CPB was therefore started with femoral artery and inferior caval cannulation. After induction of cardiac arrest, the ascending aorta was doubly clamped and transected. Excellent tumor exposure was now achieved through these cut ends of the aorta (Figure 2). On closer examination, it was suspected that the tumor also had invaded the descending aorta. A combined vessel wall resection was thus necessary at these sites, but only the pulmonary artery was resected. If a complete resection of the tumor had been the aim, selective cerebral perfusion and replacement of the descending aorta would also have been required. Because we did not wish to take on such a surgical challenge, we were able to remove only a portion of the tumor. The pulmonary artery defect was reconstructed with a polytetrafluoroethylene tube graft. The unresectable part of the tumor remained in place around the descending aorta. The ends of the transected ascending aorta were anastomosed, and CPB was terminated without difficulty. The patient's postoperative course was uneventful, and he received postoperative radiotherapy to the mediastinum totaling 70 Gy. Twelve months after the operation, the patient is doing well with no mediastinal recurrence.

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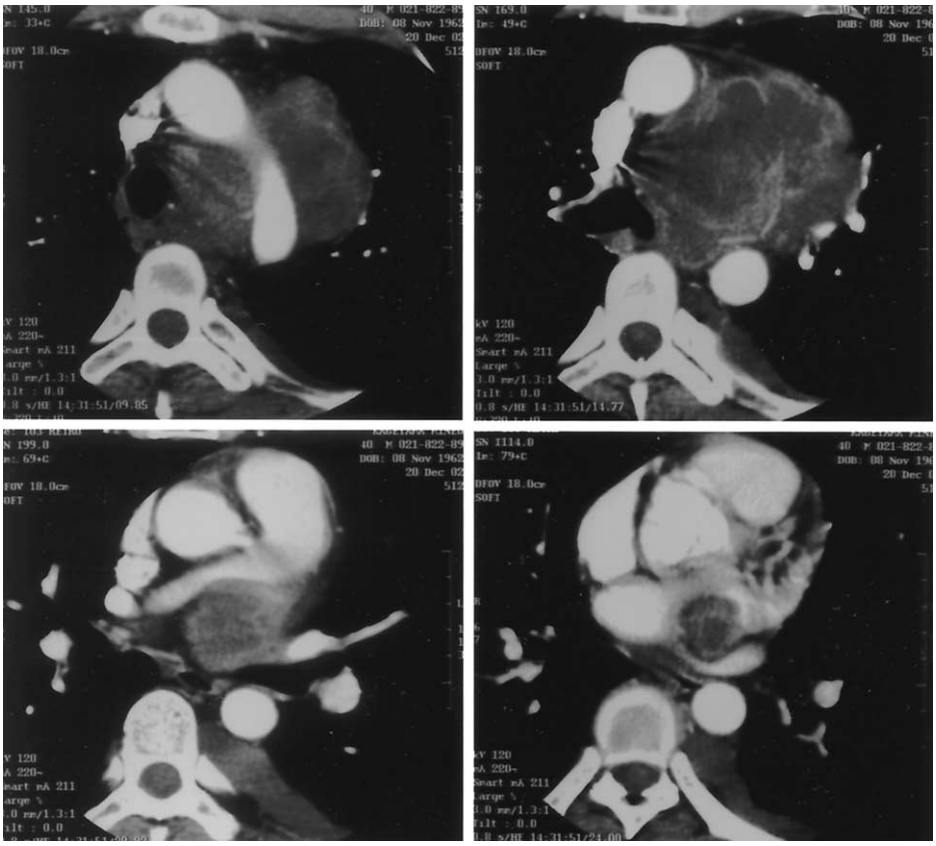

Figure 1. Preoperative chest CT scan showing tumor's complex involvement with mediastinal structures.

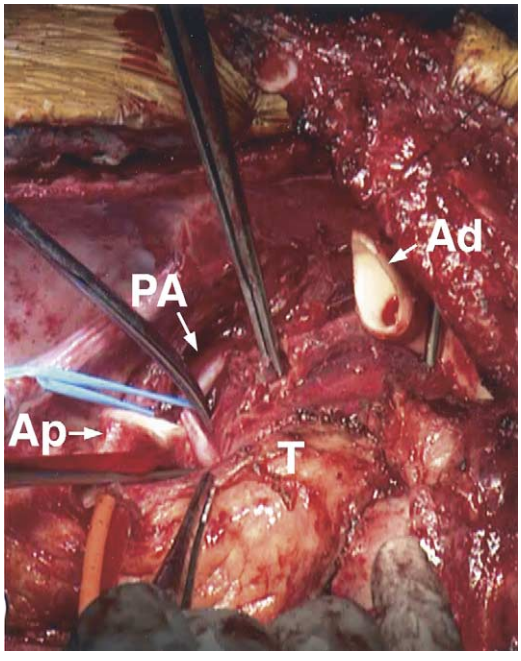

Figure 2. Intraoperative view after transecting ascending aorta. *Ad,* Cut end of ascending aorta (distal side); *PA,* right pulmonary artery; *Ap,* cut end of ascending aorta (proximal side); *T,* tumor.

Discussion

MPNST is a rare kind of tumor, usually associated with NF-1, that typically arises from a simple or plexiform neurofibroma. The tumor in this case was considered to have arisen from a neurofibroma of the left vagus and recurrent laryngeal nerve. In patients with NF-1, malignant transformation of neurofibromas, which occurs in 2% to 16% of cases, is considered to be a poor prognostic factor.[1,2] Because MPNST is associated with frequent local recurrence and metastasis, especially in patients with NF-1, surgical intervention has been the best form of treatment for these tumors. In this case the tumor was large, had a complex involvement with mediastinal structures, and was in a life-threatening location. Because the patient was relatively young, in good general condition, and free of other cardiopulmonary disorders, we chose an aggressive surgical approach.

Surgical techniques requiring CPB for advanced thoracic malignancies have been discussed and are considered beneficial in appropriate patients.[3] Temporarily dividing great vessels, such as the ascending aorta, to remove a tumor constitutes the last resort. To our knowledge this approach has not been previously reported. Under CPB, resection of this tumor by mobilizing the ascending aorta might have been possible, even without aortic transection, but this approach would have encountered enormous technical

difficulties. We decided that a temporary aortic transection approach would provide better surgical exposure and enable manipulation of the tumor, which we hoped would shorten the cardiac arrest time. Although our procedure resulted in incomplete excision, we achieved our primary purpose of preventing cardiopulmonary collapse as a result of tumor compression. This case demonstrates that a temporary aortic transection approach may be justified for appropriate patients who otherwise would have no viable treatment option.

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Twisting of pedicled left internal thoracic artery graft three hundred sixty degrees clockwise: Does it change the outcome?

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(left to right)

Twisting of aorta-coronary venous conduits has been reported previously with fatal outcome and is an extremely uncommon occurrence.[1] However, twisting of the pedicled left internal thoracic artery (LITA) graft was not found to be reported in a search of the English-language literature (MEDLINE and PUBMED), and its outcome is unknown. We report a case of an inadvertent clockwise 360° twisting of a LITA graft sequentially anastomosed to the diagonal and left anterior descending coronary arteries (LAD) with an angiographically documented patency with good distal runoff.

Clinical Summary

A 54-year-old man had a history of class III unstable angina. Echocardiographic evaluation was unremarkable. The treadmill test result was positive for inducible ischemia. Coronary angiography showed a normal left main coronary artery and total occlusion of the proximal LAD. The diagonal artery was arising from the diseased segment of the LAD. The left circumflex artery was nondominant and normal. The right coronary artery was totally occluded past the right ventricular branch. A left ventricular angiogram showed good function, and there was no regional wall motion abnormality or mitral regurgitation.

Coronary artery bypass grafting was performed by sequential grafting of the LITA to the diagonal artery and the LAD and a reversed saphenous vein graft to the distal right coronary artery under conventional cardiopulmonary bypass. After the distal anastomoses were completed, just before removal of the vascular clip from the LITA, we realized that the pedicle was twisted 360° clockwise proximal to the LITA–diagonal artery anastomosis. Although distal runoff in the diagonal and LAD was good, we were concerned about the patency of the graft.

The following options were contemplated: (1) to leave it as it was, (2) to transect the LITA at the origin and make it a free graft after untwisting, and (3) to revise the distal anastomoses. The third option was not considered further because of the involved technical difficulties. We selected the first option. The patient was weaned from cardiopulmonary bypass on the first attempt without any inotropic support and was shifted to the intensive care unit for elective ventilation. The next morning, after extubation, coronary angiography was repeated and showed a patent LITA conduit with good flow to the diagonal artery and the LAD (Figure 1). Postoperative electrocardiography revealed a normal pattern with no ST-T changes. Predischarge echocardiography did not show any regional wall motion abnormality. The patient was discharged on the sixth postoperative day. At 3 months of follow-up, he has resumed all his physical activities and is free from angina. Coronary angiography repeated at 6 months showed a patent LITA graft with good flow to the LAD and the diagonal artery (Figure 2).

Discussion

The frequency of twisting of coronary bypass conduit is unknown. Roberts and colleagues[1] reported twisting of venous bypass conduits. All 3 affected patients died, 2 patients on the operating table

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