Total resection of the aortic arch intimal sarcoma using the L-incision technique

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Abstract

A 26-year-old male suffering from sudden right lower abdominal pain and lumbago was referred to our hospital. Enhanced computed tomography demonstrated bilateral kidneys and spleen infarctions, and a large tumour was found occupying the aortic arch and thoracic descending aorta. We suspected that these infarctions were due to tumour embolization. The aortic arch and thoracic descending aorta were resected with the tumour and then reconstructed using the L-incision technique. A microscopic examination revealed the presence of an intimal sarcoma. The patient was treated with adjuvant chemotherapy and showed a good postoperative course. Neither recurrence nor metastasis has been observed during the 3 years since the operation.

Keywords: Aortic operation • Chemotherapy • Tumour

CASE REPORT

A 26-year-old male Japanese patient was referred to our hospital with a sudden right lower abdominal pain and lumbago. The level of C-reactive protein was 11.8 mg/dl. Enhanced computed tomography and magnetic resonance imaging demonstrated a large, polypoid and intravascular tumour that occupied the aortic arch and thoracic descending aorta (Fig. 1A), and bilateral kidneys and spleen infarctions were seen (Fig. 1B). Based on these findings, we suspected that the aortic tumour was malignant. Because it was obvious that the tumour was the source of the embolisms that caused bilateral kidneys and spleen infarctions, we did not perform an endovascular biopsy in order to prevent the occurrence of any further embolic events. To improve his prognosis, we decided to perform an aortic arch replacement even though the surgical mortality rate seemed to be relatively high based on the EuroSCORE [1].

The patient’s chest was opened through an upper partial median sternotomy and a left anterolateral thoracotomy in the fifth intercostal space, using the L-incision technique [2] under deep hypothermic circulatory arrest using selective antegrade cerebral perfusion and the lower body arrest technique. We connected 8 mm artificial grafts to the left axillary artery and the left femoral artery for blood perfusion. We inserted a venous cannula into the inferior vena cava from the right atrial appendage. Systemic circulatory arrest was applied at a nasopharyngeal temperature of 20°C. We took special care to avoid any further embolization of the tumour to the main organs, because the tumour was thought to be fragile and mobile. The entire aortic arch and descending aorta were resected and reconstructed using a 22 mm artificial graft with four branches.

The macroscopic findings of the resected aorta showed that the tumour had originated from the intima, grown into the inner lumen and also spread along the intima (Fig. 2A). According to histopathological evaluations, there was no invasion to the media or adventitia (Fig. 2B). Immunohistochemically, the tumour cells were positive for vimentin, negative for alpha-smooth muscle actin, CD-31, CD-24, factor VIII-related antigen (RA), S-100 protein and desmin. These features indicated that the tumour was an intimal sarcoma. Although there were no tumour cells at the end of the great arteries, tumour cells were observed at the end of the brachiocephalic and the left subclavian arteries.

The patient was treated with adjuvant chemotherapy consisting of adriamycin and ifosfamide, and showed a good postoperative course. Neither recurrence nor metastasis has been observed during the 3-year postoperative follow-up.

DISCUSSION

Primary malignant tumours of the aorta are extremely rare. The male/female ratio is 9/5 and the mean age is 59.5 years (ranging from 3.5 months to 85 years) [3]. Although aortic sarcoma, the most common type of primary tumour of the aorta, frequently occurs in the thoracic descending aorta, tumours in the aortic arch and its branch vessels are less common. Aortic sarcoma has been categorized as intimal and mural types [4]. The intimal type may have a tendency to extend along the intimal surface and...
frequently forms intraluminal polyps that may cause embolisms. The mural type may arise in the media or adventitia, and usually has less-aggressive behaviour. Most of the reported aortic sarcomas are of the intimal type, similarly so in our case.

The most common symptoms of these tumours are due to embolic events of the peripheral or mesenteric artery [4]. Other clinical symptoms are intermittent claudication, abdominal pain, back pain, fatigue and metastatic complications. Since the intimal type develops in the vascular lumen, the tumour is likely to metastasize via the blood flow and cause symptoms due to embolic events. Because aortic intimal sarcoma is a rare disease, it is not always suspected preoperatively. The diagnosis may be elusive, and in many cases, has been made by a histological examination either at autopsy or at the time of surgery for an arterial embolism, arteriosclerosis disease or aneurysm. Magnetic resonance angiography is the most sensitive diagnostic tool for aortic tumours because it may distinguish them from these other diseases and thus reveal the extent of tumour involvement in the aortic wall and adjacent structures. Bone scintigraphy and positron emission tomography are useful to detect any other sources of the tumour or its metastatic lesions [4, 5]. The use of these examinations reduces the risk of embolization due to the mobile tumour and contrast-induced renal failure compared with endovascular biopsy.

The prognosis of this disease is generally poor, and the mean survival period after the diagnosis ranges from 8 to 14 months [3]. Although the prognosis is dismal, surgery remains the main therapeutic strategy for such aggressive tumours. In fact, most of the reported cases [3, 5] were treated surgically. Although the clinical course of this disease is generally rapid and associated with a poor outcome, we decided to perform an operation to prevent further embolic events to the major organs. We performed an entire aortic arch replacement, including both the ascending aorta and the middle of the descending aorta, to remove the tumour completely. The L-incision technique [2] was extremely useful for this operation. We were able to resect the whole aortic arch with the tumour, without any tumour embolization to the major organs. Clear surgical margins are important for survival after the surgical resection of aortic intimal sarcoma. In our case, however, positive tumour cells were observed at the end of the brachiocephalic and the left subclavian arteries. We thus performed adjuvant chemotherapy.

The effectiveness of adjuvant therapies, such as chemotherapy and radiotherapy, remains unknown. The European guidelines

Figure 1: (A) Preoperative enhanced computed tomography (sagittal reconstruction) showed an intraluminal tumour like a polypoid lesion in the distal arch and thoracic descending aorta. The tumour was seen at the left subclavian artery. (B) Multiple infarctions were seen in the bilateral kidneys and spleen.

Figure 2: (A) The gross appearance of the resected specimen showed that the soft and polypoid tumour was growing in and over the aortic intima. No tumour cells were observed at the end of the specimen in the perioperative frozen section diagnosis. (B) Haematoxylin-eosin stain showed that the tumour consisted of abundant myxoid stroma with a dense proliferation of oval, spindle or polygonal-shaped cells with hyperchromatic nuclei. Chondroid differentiation was focally seen.
and the short German guidelines recommend treatment based on doxorubicin and ifosfamide as chemotherapeutic agents [3]. The role of adjuvant radiotherapy is limited to residual disease after a surgical resection or in the presence of bone metastasis. We performed adjuvant chemotherapy using adriamycin and ifosfamide. Neither recurrence nor metastasis has been observed during the 3 years since the surgery. However, further follow-up is mandatory to confirm the patient’s prognosis.

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REFERENCES


