

Recurrent Mediastinal Sarcoma in the Aortic Arch

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As mediastinal sarcomas commonly present as large tumors invading adjacent vital structures, complete resection is frequently challenging. For such tumors, aggressive surgical strategies, such as the resection and reconstruction of the invaded vital structures under cardiopulmonary bypass, may be required to achieve complete resection and to improve survival. Herein, we report a case of recurrent mediastinal sarcoma invading the aortic arch and arch vessels that was successfully removed by total arch replacement.

Key words: 1. Mediastinal neoplasms
2. Recurrence
3. Aortic arch
4. Resection
5. Reconstruction

Case report

A 52-year-old man presented to Asan Medical Center with a recurrent mediastinal mass incidentally found on follow-up chest computed tomography (CT) (Fig. 1). The patient had undergone the excision of an anterior mediastinal sarcoma 6 years ago at another hospital. That mass measured 150 mm×100 mm, and was completely removed through a median sternotomy combined with a left thoracotomy. His past medical history revealed total thyroidectomy due to thyroid cancer and paroxysmal atrial fibrillation that had persisted for the previous 6 years.

Preoperative transthoracic echocardiography (TTE) revealed severe left ventricular (LV) dysfunction with an ejection fraction (EF) of 22% accompanied by mild mitral and tricuspid insufficiency. In addition, a newly developed anterior mediastinal mass measuring 35 mm×30 mm abutting the aortic arch was observed. The mass, which had a broad base attached

to the right brachiocephalic vein, encircled both the right brachiocephalic and left common carotid arteries.

We planned complete en bloc excision of the invaded tissue, followed by reconstruction of the aortic arch and superior vena cava (SVC). After a redo median sternotomy, cardiopulmonary bypass (CPB) was established by cannulating the ascending aorta and right atrium. The aorta was cross-clamped, and then 1 L of cold del Nido cardioplegic solution was administered via the aortic root. A left-sided Maze procedure with cryoablation was performed through a left atriotomy. After closure of the left atriotomy, the entire tumor, including the invaded SVC, right brachiocephalic vein, and aortic arch, was resected en bloc under total circulatory arrest at a nasopharyngeal temperature of 21°C (Fig. 2). First, the right brachiocephalic artery was anastomosed to a 4-branched vascular graft (Hemashield Platinum Woven Double Velour Vascular Grafts 24 mm; Maquet Holding BV & Co. KG, Rastatt, Germany) to secure immediate

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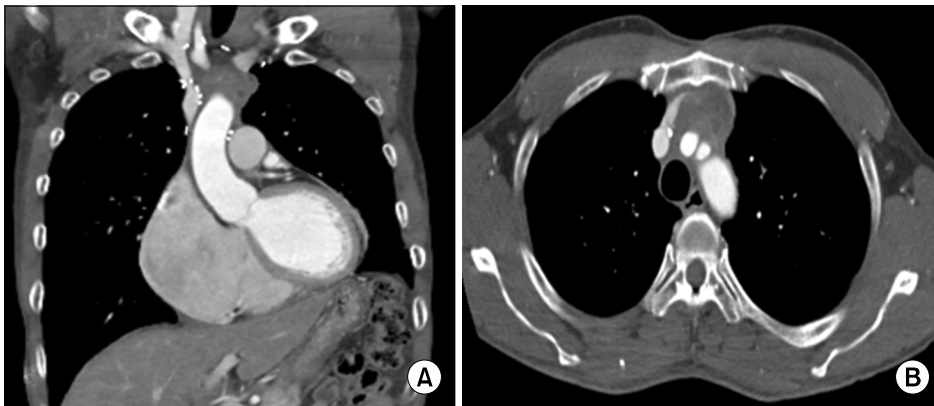


Fig. 1. Preoperative chest computed tomography showing an anterior mediastinal mass (35 mm×30 mm) abutting the aortic arch and encircling the right brachiocephalic artery and left common carotid artery. (A) Coronal view. (B) axial view.

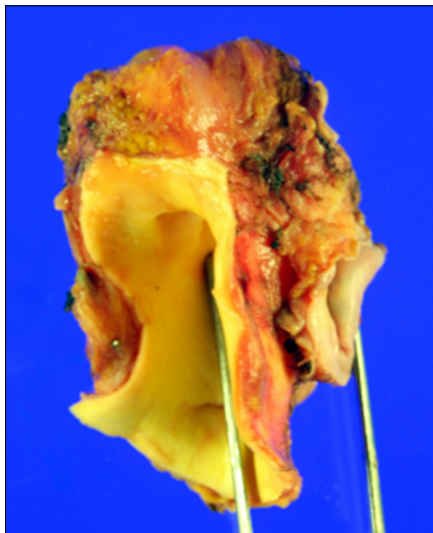


Fig. 2. Gross specimen: mediastinal tumor and partially resected superior vena cava, right brachiocephalic vein, and aortic arch.

unilateral antegrade cerebral perfusion. Then, anastomoses of the descending thoracic aorta, left common carotid artery, and proximal ascending aorta were performed, in order. The left subclavian artery was anastomosed under beating-heart CPB after the release of aortic cross-clamping (ACC). The SVC was then reconstructed using a bovine pericardial patch tailored in a tubular shape. Weaning from CPB was smooth, and normal sinus rhythm was restored. The intraoperative frozen biopsy revealed a negative resection margin, free of tumor cells.

The total circulatory arrest time, lower body ischemic time, ACC time, and CPB time were 13 minutes, 23 minutes, 78 minutes, and 132 minutes, respectively. The patient was transferred to the intensive care unit and extubated after 12 hours. The patient

was transferred to the general ward on postoperative day (POD) 2 and was discharged on POD 9 without complications. Postoperative TTE showed no changes in overall cardiac function compared to the preoperative exam, except for slightly alleviated LV dysfunction, with an EF of 28%. Postoperative CT showed no evidence of a residual tumor, and confirmed the presence of a well-reconstructed arch, arch vessels, and SVC without architectural distortions (Fig. 3). The pathologic examination revealed a myxofibrosarcoma with resection margins free of tumor cells (pT1bN0M0, stage IA according to the American Joint Committee on Cancer seventh edition). The patient has been followed up at our outpatient clinic for the last 9 months, and no evidence of tumor recurrence has been found. Recent follow-up TTE revealed normalized LV systolic function, with an EF of 63% and sinus rhythm. The patient's resting pulmonary hypertension was relieved, and the extent of mitral and tricuspid insufficiency was reduced.

Discussion

Mediastinal sarcomas are uncommon tumors; they account for approximately 1% of all soft-tissue sarcomas and fewer than 10% of primary mediastinal tumors [1]. Mediastinal sarcomas are often asymptomatic until they grow to a considerable size, and the symptoms caused by the compression of surrounding structures are often nonspecific. Sarcomas originating from the mediastinum usually abut or invade vital structures in the early phases. Therefore, an early diagnosis before fatal local invasion can be difficult, and complete resection can be technically challenging

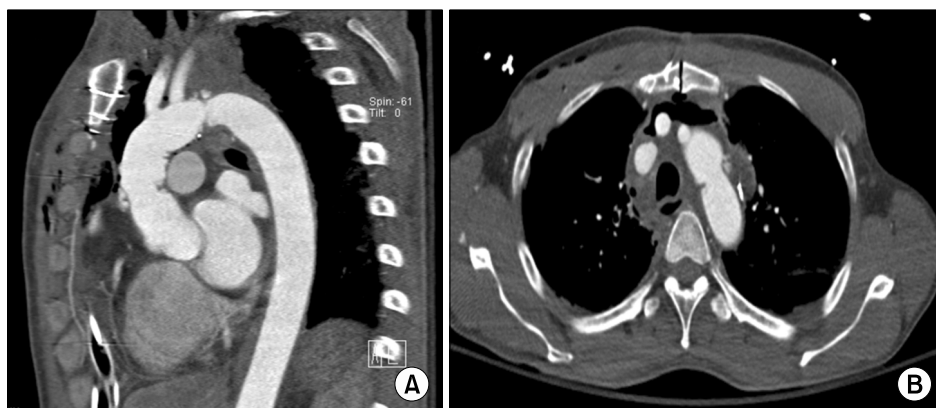


Fig. 3. Postoperative chest computed tomography. (A) Sagittal view. (B) axial view.

[2,3].

In the management of sarcomas arising in the mediastinum, the only treatment that has shown a significant survival benefit is complete surgical resection [4]. Although the predictors of survival after surgery have not been clearly identified, the size and grade of the tumor and the quality of the surgical margins are generally considered to be associated with survival [5], whereas the benefits of adjuvant therapies such as radiation therapy and chemotherapy have not been proven [4].

Several researchers have compared the survival outcomes of patients with mediastinal sarcoma based on surgical margin status after surgical resection (incomplete versus complete resection margin), showing complete resection to be associated with significantly improved overall and disease-free survival [1,6]. These results underscore that securing negative surgical margins is essential for prolonged survival, and that aggressive surgical strategies should therefore always be considered whenever needed and anatomically feasible.

In the present case, surgical management seemed extremely challenging, as the tumor had invaded the aortic arch and arch vessels. Nevertheless, we assumed that complete resection of the tumor would be feasible with a straightforward surgical strategy. Due to the location of the tumor, however, conventional selective antegrade cerebral perfusion for arch repair through the right axillary artery or innominate artery was not possible. For this reason, we sought to secure cerebral perfusion by the immediate restoration of right brachiocephalic artery flow with a minimal total circulatory arrest time by changing our routine sequence of arch anastomoses during total

arch replacement. Finally, we successfully completed the surgical procedure without complications using a modified surgical strategy with a reasonable procedural time despite the reported high surgical risks of total arch replacement in general.

The patient had atrial fibrillation with severe LV dysfunction. Loss of atrial contraction may markedly decrease cardiac output, and a persistently elevated ventricular rate may cause tachycardia-induced cardiomyopathy. A rapid ventricular rate may be associated with rate-related intraventricular conduction delays. Conduction disturbances compromise the synchrony of LV wall motion and reduce cardiac output, thereby exacerbating mitral valve insufficiency [7]. We decided to perform a Maze procedure to reverse those effects by controlling the ventricular rate.

In conclusion, concomitant resection and reconstruction of the great vessels using CPB and a modified arch reconstruction strategy may be a reasonable treatment option for recurrent mediastinal sarcoma invading these vital structures, because it may afford the best opportunity to cure the disease and to prolong survival.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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