Case report

Extended operation for invasive thymoma with intracaval and intracardiac extension

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Abstract

Two cases of invasive thymoma with intracaval and intracardiac extension into the right atrium are reported. Radical excisions and reconstructions of the superior vena cava (SVC) requiring extracorporeal circulation were performed. Invasive thymoma with this growth pattern is extremely rare, and patients with SVC obstruction should be evaluated for the resection of tumors and reconstruction of the SVC.

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1. Introduction

Radical excision and reconstruction of the superior vena cava (SVC) requiring extracorporeal circulation (ECC) were mandatory for advanced thymoma [1], although ECC is rarely required in the excision of thymoma. We report two cases of invasive thymoma with intracaval and intracardiac extension into the right atrium (RA).

2. Case 1

A 77-year-old man, complaining of swelling of the face and the left upper extremity, was admitted. Computed tomography revealed a mass in the anterior mediastinum infiltrating the SVC and the RA (Fig. 1A, B). Echocardiography revealed the tumor thrombus in the RA. The diagnosis of thymoma with tumor thrombus extending into the SVC and the RA was made. An urgent surgery was performed instead of preoperative chemo- or radiotherapy, because the tricuspid orifice was likely to be obstructed. A median sternotomy was performed. A firm and nodular tumor was found in the anterior mediastinum. ECC was instituted by cannulating the right femoral artery and vein and the right jugular vein. After both brachiocephalic veins and inferior vena cava were cross-clamped, the SVC and the RA were opened widely without cardiac arrest, showing the complete occlusion of the lumen by the encapsulated tumor thrombus measuring 19 $\times$ 8 $\times$ 6 cm (Fig. 2). The tumor thrombus that adhered only to a small portion of the orifice of SVC and extended across the tricuspid orifice into the right ventricle was removed, and the SVC was reconstructed using two 10 mm polytetrafluoroethylene grafts, one from the orifice of the SVC to the right brachiocephalic vein and the other from the atrial appendage to the left brachiocephalic vein in an end-to-end fashion, respectively. Pretracheal lymph nodes could not be dissected completely because of severe adhesion (Masaoka’s Stage IVb). Operation time was 384 min and ECC time was 81 min.

Pathologic examination revealed that the tumor was type A thymoma (WHO classification) that had directly invaded the junction of the bilateral brachiocephalic veins, from where the tumor thrombus extended into the SVC and the RA. Postoperatively the patient received radiotherapy (40 Gy), and he was still well 21 months after surgery.

3. Case 2

A 27-year-old female complaining of dyspnea was admitted. Computed tomography revealed an anterior mediastinal mass, and magnetic resonance imaging showed the continuous mass extending into the SVC and the RA.
(Fig. 1C, D). Superior vena cavaography showed the complete obstruction with multiple collaterals. Preoperative transthoracic TruCut needle biopsy was consistent with thymoma. She was initially treated by chemotherapy consisting of cisplatin (80 mg m\(^{-2}\) on day 1), vindesine sulfate (3 mg m\(^{-2}\) on days 1 and 8) and mitomycin (3 mg m\(^{-2}\) on day 1). Radiotherapy (16 Gy) could not be completed due to the bone marrow suppression. After two courses of chemotherapy, the tumor size was reduced by about 40%. Then the resection of tumors and reconstruction of the SVC using two 8 mm polytetrafluoroethylene grafts were performed under ECC using the same fashion as of case 1. After removal of the tumor, acute circulatory failure occurred secondary to an inadequate venous drainage of ECC. In this case, operation time was 649 min and ECC time was 150 min. The following day, she died due to multiple organ failure. Postoperative pathologic examination revealed no viable cell.

4. Discussion

SVC syndrome is usually associated with advanced malignancy and has a dismal prognosis. The causes of SVC syndrome include advanced lung cancer (57%), mediastinal tumors (20%) and metastatic solid malignancy (5%) [2]. SVC syndrome associated with thymoma is rare (4%) [2], and the most common cause is extrinsic compression rather than intracaval growth in the SVC and the RA [3]. Thymoma with this growth pattern is extremely rare, and only seven cases have been reported [3,4]. It has been suggested that small thymoma in the anterior mediastinum infiltrated the brachiocephalic vein and grew along the venous stream into the SVC and down to the RA in polyp fashion [3].

Most patients with malignant SVC syndrome are treated with radiotherapy and/or chemotherapy for palliation with a mean survival of 3 months [2]. Radiologic stenting is a safe technique, which offers rapid palliation. However, radical excision for neoplasms with SVC obstruction, infiltrating the SVC, bilateral brachiocephalic veins, and the RA is rarely performed, such as in this case.

In the treatment of thymoma, surgery is considered as the main therapy. Recently, multimodal therapy, including surgery, radiotherapy and chemotherapy, is thought to be effective for advanced invasive thymoma [5]. Among the
seven reported cases, preoperative chemotherapy was performed for one case because the tumor was considered unresectable. Since case 1 involved an impending cardiac obstruction, surgical intervention, followed by radiotherapy, was selected. In case 2 with multiple collaterals, preoperative chemoradiotherapy followed by radical excision was planned. Case 1 had been well for 21 months after surgery. Minato et al. [3] also reported his patient who received three courses of postoperative chemotherapy and was doing well 29 months after surgery. The 1-year survival rate of these nine cases was 71.4%. Case 2 who was treated by preoperative chemotherapy, unfortunately died due to multiple organ failure. In case 2, we should established ECC by the ascending aortic cannulation and venous cannulation inserted at inferior vena cava with cardiac arrest. However, we can expect an improved survival by combining radical surgery with chemoradiation therapy for invasive thymoma with intracaval and intracardiac extension, and therefore patients with thymoma should be evaluated for SVC reconstruction with ECC.

To summarize, we report two cases of invasive thymoma with intracaval and intracardiac extension. Radical excisions and reconstructions of the SVC requiring ECC were performed.

References

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