THORACIC SURGERY

SURGICAL TREATMENT OF AN ADVANCED STAGE THYMOMA IN A GOOD'S SYNDROME PATIENT – CASE REPORT

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Abstract

We report a rare case of an advanced stage thymoma with right superior pulmonary lobe, superior vena cava, innominate vein and pericardium invasion in a patient with Good's syndrome. In a multidisciplinary discussion, surgical resection was deemed the best initial approach, since invaded structures could be safely managed.

The tumor was fully resected and included partial resection of the superior pulmonary lobe, superior vena cava and innominate vein. The encircled right phrenic nerve was dissected from the tumor and preserved. The superior vena cava and innominate vein were reconstructed using autologous pericardium patch. Immunoglobulin replacement and radiotherapy were initiated afterwards. No signs of relapse at 6 months follow-up. In such advanced cases, aggressive surgical intervention should be considered as first line of treatment, as long as full resection can be anticipated, since complete resection is the leading factor for long-term prognosis.

INTRODUCTION

The association of thymoma (the most common tumor of the anterior mediastinum in the adult) with an immunodeficiency state, known as Good's syndrome, is rare. The cause and pathogenesis remains unknown and the initial presentation of this often-asymptomatic syndrome can result from local progression and compression by the tumor and/or recurrent bacterial infection due to higher susceptibility. One of the most consistent immunological abnormalities is hypogammaglobulinaemia.[1]

Complete surgical resection of the tumor is the basis of the treatment and the most important aspect related to long-term prognosis. As in other advanced tumors, the strategy dependents of resectability.[2] When not possible, multidisciplinary approach with induction chemotherapy is an option, as an attempt to achieve complete or partial resection. Although new protocols, in selected groups, have shown favorable results,[3] in general, neoadjuvant therapy is associated with lower response rates, higher surgical complexity and worse prognosis.[4,5]

Here we report a case of a patient with Good Syndrome and Masaoka stage III thymoma treated initially with a radical resection followed by radiotherapy.

CASE REPORT

A 39-year-old woman with no relevant medical history was admitted with urinary symptoms, diarrhea, fever, and facial and upper limb swelling. Urinary and gastrointestinal bacterial infections were diagnosed. Laboratory tests revealed a hypogammaglobulinemia and the Chest X-ray identified a mediastinal mass, posteriorly characterized by CT scan as a massive anterior mediastinum tumor (7.9x6.7x6.2cm), compressing the superior vena cava (SVC) and innominate vein (IV) (Fig. 1A and 1B). A video-assisted surgical biopsy was performed and a B1Thymoma was diagnosed. The presence of hypogammaglobulinemia, infections and thymoma suggested the diagnose of Good´s syndrome.

Chest MRI (Fig. 1C and 1D) revealed possible invasion of the IV and pericardium (Masaoka stage III), but no signs of SVC or ascending aorta involvement. In multidisciplinary discussion, the decision was to perform surgical resection as first line and best treatment option. After median sternotomy, intraoperatively, an anterior mediastinal mass was found surrounding the right phrenic nerve and invading the superior pulmonary lobe, the proximal SVC, the IV and the underlying pericardium (the ascending

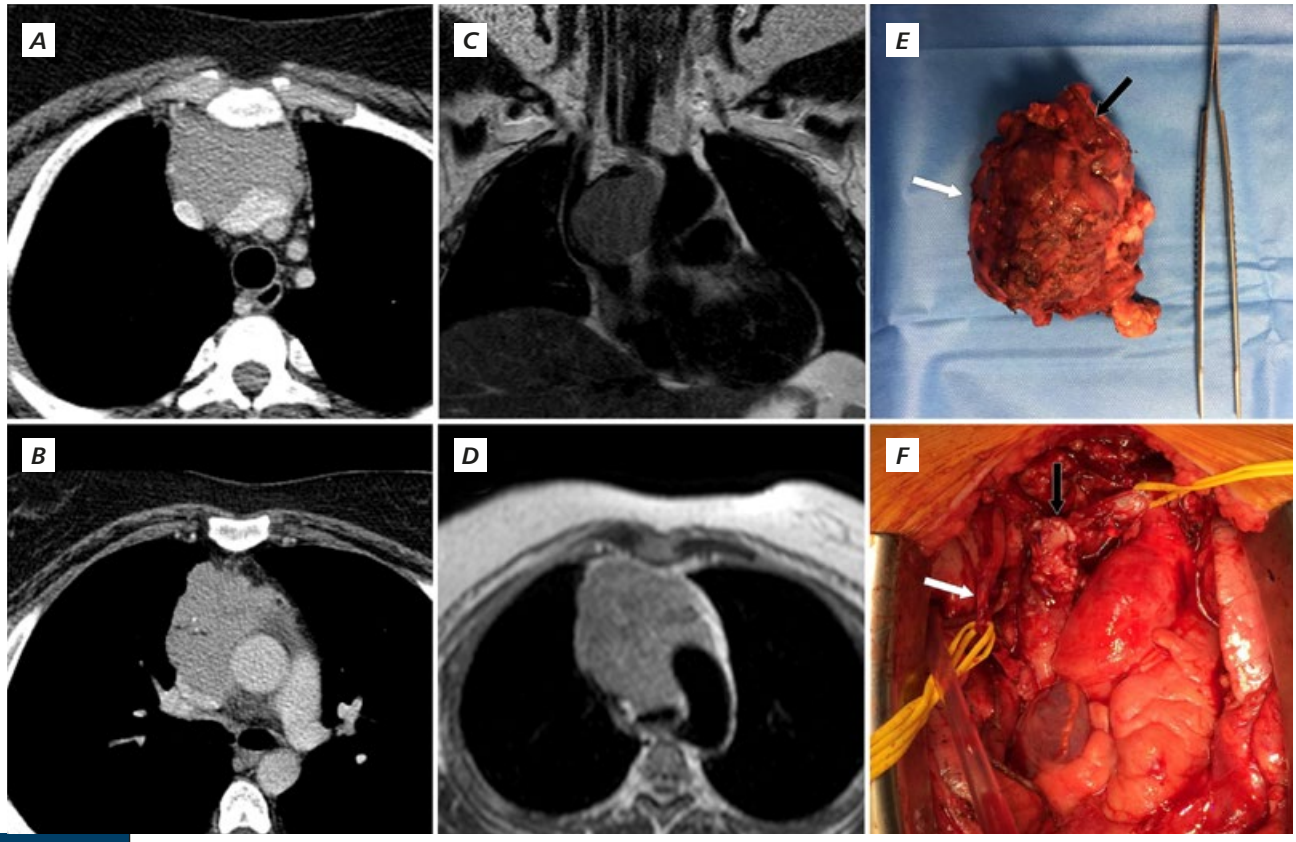

Figure 1

*Invasive thymoma in a patient with Good Syndrome. CT imagens showing the location and size of the tumor (**A** and **B**). MRI imagens showing the compression and invasion of the superior vena cava (**C**) and pericardium (**D**). Resected tumor (**E**, white arrow) with the invaded lung portion (**E**, black arrow). Venous repair with pericardium (**F**, black arrow) and preserved phrenic nerve (**F**, white arrow).*

aorta was not invaded). The right phrenic nerve was dissected from the tumor and preserved. The superior pulmonary lobe was partial resected with a mechanical surgical stapler (Fig 1E). The SVC and IV were partialy resected and reconstructed using autologous pericardium patch (Fig 1F) with partial clamping and without extracorporeal circulation. A complete macroscopic resection was achieved. The post operatory was uneventful.

Pathologic examination confirmed invasion of the lung and veins and focally positive surgical margins - pT3N0 / Masaoka III. Immunoglobulin replacement and radiotherapy were initiated. At 6 months follow-up, the patient was clinically well and there were no signs of metabolic active disease in the mediastinum (PET-CT with FDG).

DISCUSSION

The treatment of patients with Good's syndrome is not yet well established and the principles of management of thymoma are usually applied. Usually, removal of the thymoma does not reverse the immunological abnormalities and immunoglobulin replacement is required.[1]

Regarding the thymoma itself, for early stages (Masaoka I and II) surgical resection is enough, while for more advanced stages (III and IV) induction chemotherapy and/or radiotherapy may be required to decrease the tumor size and achieve resectability. In addition, the approach and extension of the resection depends on the involved structures and usually vascular involvement is not a contraindication.[6,7]

In the presented case, preoperative neoadjuvant treatment was not performed because it would delay the surgical resection without significant advantage, since the suspected invaded structures could safely be addressed. Despite intraoperatively the invasion was greater than suggested by the TC/MRI scans, complete resection was achieved.

While the invasion of the lung and pericardium was fairly straightforward to manage (with a broad lung and pericardium resection) the right phrenic nerve preservation was challenging. Resection of the nerve would mean paralysis of the homolateral diaphragm, and consequent associated comorbidity, so all efforts were done to preserve it. In the end it was possible because the nerve was enveloped and not invaded by the tumor. The resection and repair of the SVC and the IV was possible with partial clamping, avoiding extracorporeal circulation and its known side effects.[8] An autologous pericardium patch from an area distant to the tumor was used to decrease the risk of relapse. Arguably, a vein xenograft or synthetic patch could also be used.

Despite being a challenging procedure, the surgery proved to be a good first step in the treatment, as shown by the pathologic exam and the 6-month follow-up evaluation.

In conclusion, this case shows that in patients with

Good's syndrome, as in other patients with thymoma, the thymoma management is the critical step in the treatment. If possible, radical surgical resection should be initial step, even in the face of a complex procedure involving vascular reconstruction. Immunoglobulin replacement is required.

EDITOR COMMENT

I congratulate the authors on a job well done, as resectable thymomas should be approached primarily by surgery. However, a hypothesis for induction chemotherapy might have been considered, as the expected good response of a borderline resectable B1 thymoma to a cysplatin based regimen would have made their job easier, increasing the probability of a R0 resection, and is a good choice for downstaging in stage III/IV thymomas. Long term follow-up is of utmost importance as B1 Thymomas are slow growing tumors, and local recurences should be surgically managed whenever possible.[1]

Cristina Rodrigues
Associated Thoracic Surgery Editor

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