Case Report

Severe superior vena cava obstruction in thymoma - long term control and survival with combined modality treatment

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ABSTRACT

Thymoma is a tumor of thymus and is a rare anterior mediastinal tumor. Locally advanced thymoma with superior vena cava obstruction (SVCO) is an uncommon presentation. We here present a case of locally advanced case of thymoma with SVCO, which was managed with radiotherapy (RT) and chemotherapy (CCT). The patient had radiological complete response and is disease free for eight years.

Key words: Chemotherapy, mediastinal tumor, radiotherapy, superior vena cava syndrome, thymoma

INTRODUCTION

Thymoma is a rare anterior mediastinal tumor.[1] It rarely presents as a locally advanced tumor with superior vena cava obstruction (SVCO).[1] Though surgery is the primary treatment in localized disease, locally advanced thymoma with SVCO possesses a therapeutic dilemma with regards to management approach. Initial therapy is directed towards relief of symptoms of SVCO with localized mediastinal radiotherapy (RT) followed by systemic chemotherapy (CCT) and assessment for resection of residual tumor.

CASE REPORT

A 37-year-old male patient was presented in August 2004 with complains of difficulty in breathing for two months. He also had feeling of fullness in chest and had engorged neck vessels with swelling of face and neck for one month. There was no history of trauma, hemoptysis or melena. Past and family history was not significant. He was non-smoker and non-alcoholic. On general physical examination, he had bilateral dilated engorged neck veins and swelling of face and neck. No clinical neck nodes were present. On respiratory system examination, a stony dull note was felt in right infrascapular region on percussion with diminished breath sounds in right infrascapular region on auscultation. Cardiovascular and abdominal examinations were normal. Chest X-ray showed mediastinal widening. CECT – Chest revealed a large soft tissue mass measuring $4.6 \times 8.2 \times 9.0$ cm in superior part of anterior mediastinum with loss of fat planes with adjacent great vessels and bronchi [Figure 1]. CECT – neck did not showed any mass or lymphadenopathy. Ultrasound of abdomen and pelvis were essentially normal. Following this an ultrasound guided FNAC was done from the mediastinal mass, and it showed cluster of malignant epithelial cells with presence of few lymphocytes in background (Haematoxylene and Eosin stain) and was reported as Thymoma [Figure 2]. So, the final diagnosis of thymoma (Masoaka Stage III) with SVCO was made. He was immediately started on medical decompressive measures and mediastinal RT was delivered in the doses of 30 Gy in 10 fractions in 2 weeks. He had significant improvement in symptoms and at the completion of RT had approximately 50% symptomatic relief. He underwent repeat CECT – Chest for response evaluation, and it showed a 60% decrease in size of the mediatinal

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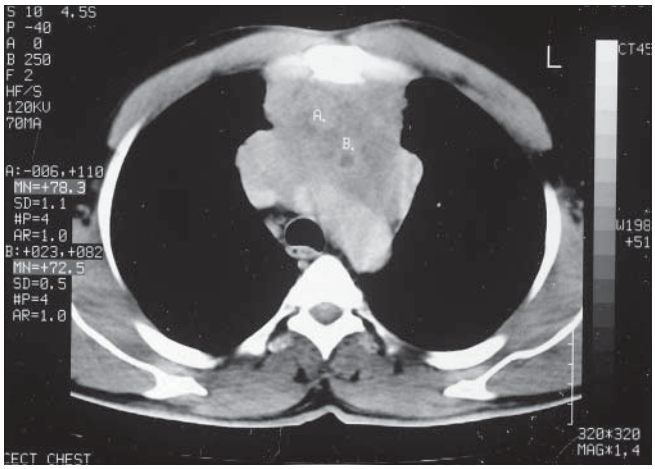

Figure 1: CECT – Chest showing a large soft tissue mass superior part of anterior mediastinum with loss of fat planes with adjacent great vessels and bronchi

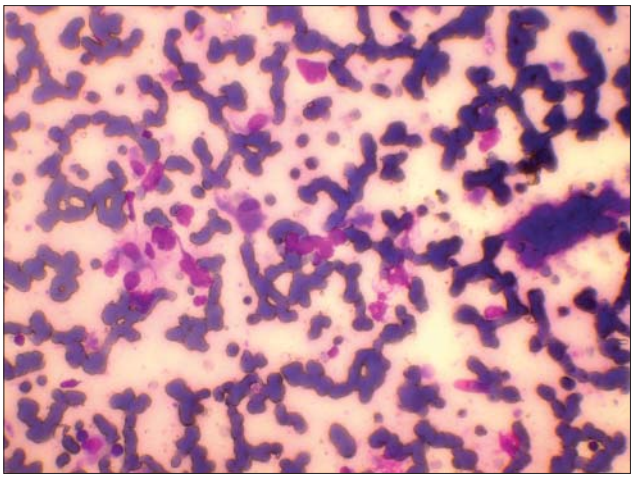

Figure 2: Microphotograph showing cluster of malignant epithelial cells with presence of few lymphocytes in background (Haematoxylene and Eosin stain)

mass, but it was still adherent to major vessels. Surgical opinion was sought regarding the operability of the residual mass and it was regarded as inoperable. In view of residual mass and surgical inoperability, he was further given systemic chemotherapy with cyclophosphamide, adriamycin, and carboplatin every three weeks for four cycles. Post chemotherapy CECT – Chest was essentially normal without any significant residual mediatinal mass. He was further kept on regular follow-up with periodic clinical and radiological evaluation. His last visit in the outpatient department was in June 2012 and was clinically and radiologically free of disease. This is a rare case of invasive thymoma with SVCO being treated with combined modality treatment (sequential RT followed by chemotherapy) and having complete cure persisting for eight years.

DISCUSSION

Thymic malignancies are relatively uncommon, with an incidence of approximately 2.5 to 3.2 per 10^6 people.[1] Thymoma arise from epithelial cells of thymus and comprises 50% of anterior mediastinal tumors.[1] It is associated with myasthenia gravis (MG) in 45% of cases.[2] Peak age of incidence is 7th decade but seen early in patients with MG (4th decade). Forty percent of the patients are asymptomatic, whereas 40% presents with local compressive symptoms and the rest 30% have systemic signs.[1] The index case presented in the 4th decade of life with local compressive symptoms, but did not had any parathymic syndromes (MG).

The spread of tumor is mainly regional, with rare lymphatic and hematogeneous metastasis. Different staging systems have been used, but the Masaoka-Koga system is most widely used.[3,4] The index case was Masaoka Stage III, in view of invasion in the neighboring structures macroscopically.

Forty percent of the patients presents in locally advanced or in-operable stages with a significant proportion with SVCO as in the index case.[5] The ideal approach is combined modality treatment using all of the three modalities (RT, CCT, and Surgery). CCT and RT in neoadjuvant approach leads to significant tumor regression and finally have complete resections in approximately 60-75% of cases.[6-8]

Thymomas are the 3rd most common cause of SVCO.[5] Early local RT is indicated in patients with SVCO and high doses per fraction is given for early palliation of symptoms (30 Gy in 10 fractions or 20 Gy in 5 fractions).[5] At presentation, the index case had SVCO with distressing respiratory symptoms, and so, was given early mediastinal RT (30 Gy in 10 fractions).

With combined modality approach, the pathological complete response rates ranges from 6-31% in different series.[6] The index case had radiological complete response to the combined modality treatment and is radiologically and clinically disease free for eight years post treatment completion.

For follow-up, yearly computed tomography (CT) scans of the thorax is recommended for five years and then alternating annually with a chest radiograph until year 11, followed by annual chest radiographs alone.

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