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A Case of Thymoma Presenting as Respiratory Failure

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Case Report

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Abstract: Thymomas are among the most common anterior mediastinal tumors that can uncommonly present with respiratory failure. This case – A 62 year old lady who presented with complaints of limb weakness and the development of type II respiratory failure, discusses in detail the problem posed and the workup leading to the diagnosis. Surgery was performed based on the radiological findings. The tumor was completely resected and the diagnosis given as Masoka classification-Type B2-predominantly cortical thymoma

Keywords: Thymomas, mediastinal tumors, respiratory failure.

INTRODUCTION

Thymomas are one of the most common anterior mediastinal masses with symptoms that indicate compression of the mediastinal structures. The characteristics of the tumor vary depending on the degree of neoplasia and can show metastasis to distant organs. Respiratory failure associated with thymoma is less common, which can be attributed to a number of causes. This case report sheds light on the course of respiratory failure in a patient with malignant thymoma without metastasis.

CASE REPORT

A 62 year old female presented to the ER with chief complaints of breathlessness and bilateral upper and lower limb numbness for a week, associated with altered sensorium past 2 days.

The patient was a hypertensive and hypothyroid on medications. She was married with two healthy children and was a homemaker. No history of tobacco, drug use or alcohol consumption were noted. Family history was unremarkable.

Upon examination the patient was drowsy, arousable and responding to oral commands. Vital signs in the emergency department revealed a pulse of 88 beats per minute, bp of 150/90mmhg, saturation of 99% with 6L O_2 via Hudson mask. Respiratory system examination revealed bilateral diminished basal air entry. The cardiac and abdominal examination was within normal limits. The neurologic exam was normal except for decreased muscle power bilaterally in both the upper and lower extremities (lower limb greater than upper limbs).

Initially laboratory values included an ABG taken in the ER which showed a pH of 7.27, pCO2 88, pO2 107, HCO3 of 39.4. Basic serum chemistries and complete cell count with differentials, Renal and liver function tests were found to be within normal limits. Chest radiograph demonstrated a suspicious mass in the suprahilar region. There was no evidence of pleural effusion, parenchymal lung disease, osseous deformities or cardiac decompensation.

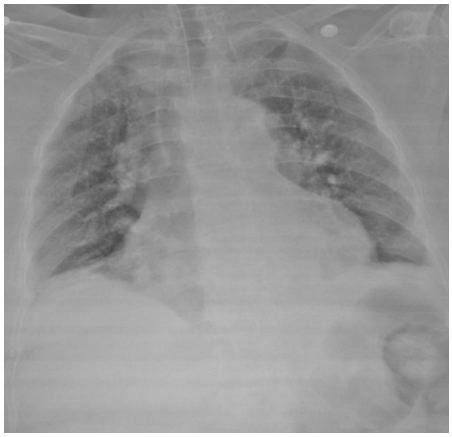

Fig-1: An anterio-posterior view of the chest demonstrating mediastinal widening

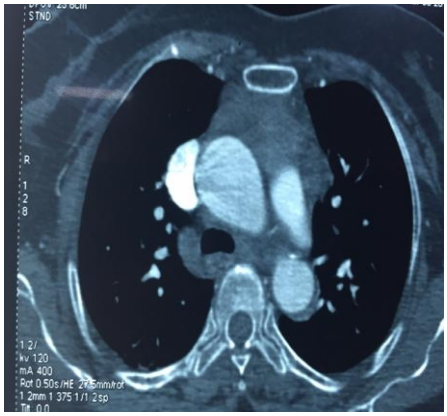

Fig-2: An axial CT image of the thorax obtained with IV contrast material demonstrating an anterior mediastinal mass

The patient was started on non-invasive positive pressure ventilation and transferred to the intensive care unit for further evaluation and treatment of type 2 respiratory failure of unknown etiology.

A CT of the chest with intravenous contrast material revealed an anterior mediastinal mass extending from above the aortic arch to the level of the left main pulmonary artery. The patient continued to be tachypnoiec and in view of impending respiratory arrest, the patient was intubated on the 3rd day post admission.

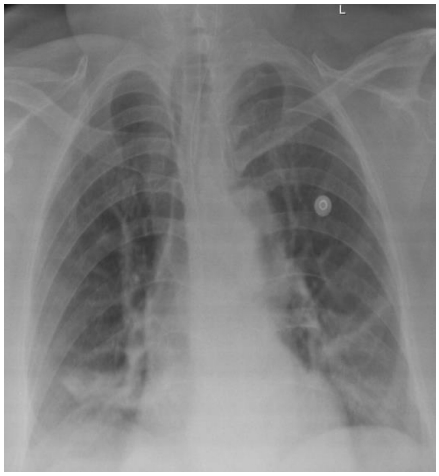

Fig-3: An XRAY taken post intubation

The next step in evaluation involved a CT – guided biopsy of the mass leision

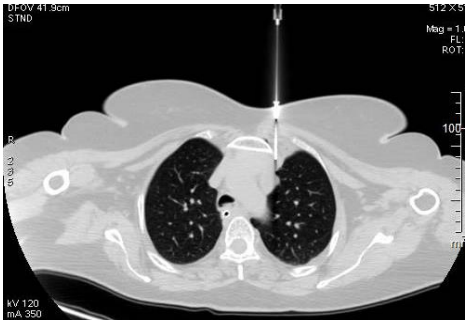

Fig-4: CT Guided Biopsy

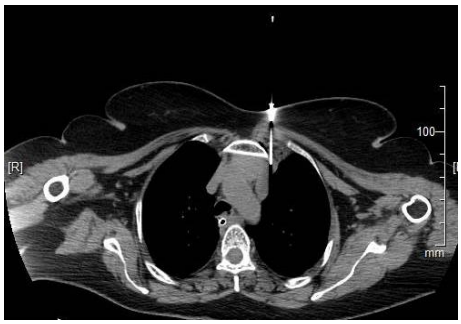

Fig-5: mediastinal window

The Biopsy was reported as Thymoma – B2 type

Other disease specific investigations included Anti Acetylcholine receptor antibodies which were found to be markedly elevated - 29.23 nmol/L.

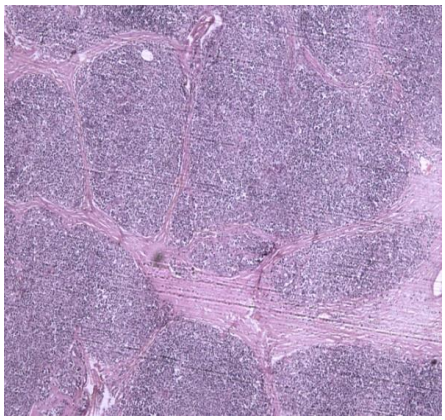

Fig-6: Tumour cells arranged in lobules separated by fibrous septa

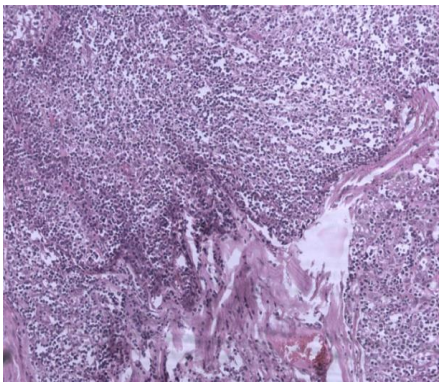

Fig-7: Mixed population of lymphocytes and plump cells

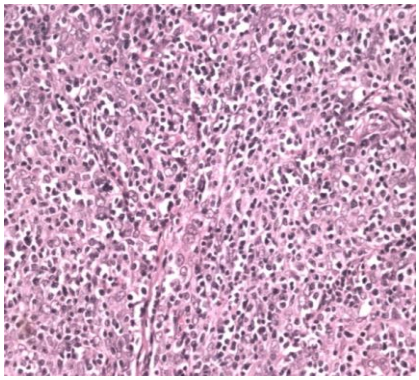

Fig-8: Lymphocytes with scant cytoplasm and hyperchromatic nuclei

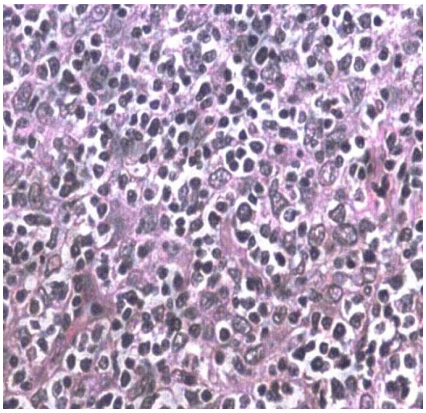

Fig-9: Plump cells have moderate cytoplasm, round to oval, irregular nucleus and clumped chromatin

Management

The cardiothoracic surgical team opinion was obtained and excision of the medistinal mass was done.

The paraneoplastic phenomenon of myasthenia gravis in association with a malignant thymoma was then postulated as the likely etiology of the patient's neuromuscular weakness and episodes of respiratory failure secondary to phrenic nerve associated diaphragmatic weakness. The excision biopsy revealed 39gm mass of thymoma of B2 cortical type and staged as type pT2 as the tumour invaded perivascular connective tissue and clinically no metastasis was observed.

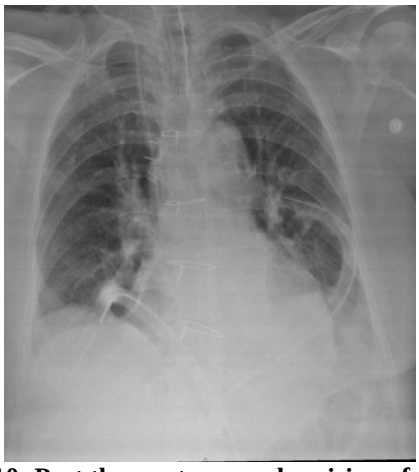

Fig-10: Post thoracotomy and excision of mass

On day13 patient extubated and contininued on NIV, Day 17 patient underwent tracheostomy in view of repeated weaning failure. Patient was started on T. Pyridostigmine 60mg TDS, with steroids in high doses and intermittent pressure support and spontaneous mode of ventilation, Failure to wean off ventilator support prompted plasmapheresis of 5 cycles. Also immunosuppressive therapy with T. Azathioprine 50mg OD was added. Corticosteroids gradually tapered off. On day 35 repeat anti AchR antibody assay was done

which revealed levels of 11.78 nmol/litre significant reductions compared to the values on admission. Serial levels of anti AchR antody levels showed a downward trend that correlated with improvement of her symptoms. Day 53 tracheostomy was decanulated and patient was discharged home on T. Azoran 50mg BD, T. Pyridostigmine 120mg TDS and tapering doses of steroids.

DISCUSSION

Thymoma, the most common of anterior mediastinal tumours, originates within the epithelial cells of the thymus. It accounts for 20–25% of all mediastinal tumors and 50% of anterior mediastinal masses. They usually present during the fourth or fifth decades of life and are mostly observed equally in men and women with a slight male preponderance. Majority of patients experience symptoms including cough, chest pain or dyspnoea and patients with more advanced disease may experience weight loss, fever night sweats or symptoms related to superior vena cava syndrome. Posteroanterior (PA) and lateral chest radiographs can detect most thymomas. On the PA view, the lesion typically appears as a smooth mass in the upper half of the chest. The mass usually projects predominantly into one of the hemi thoraces. On CT scan, thymoma usually appears as a well-defined round or oval mass located anterior to the great vessels and heart, below the left innominate vein. The staging of the disease is based on these radiographic studies as well as findings during surgical exploration. The most widely used staging system was developed by Masoka and colleagues and is based on the extent of tumour invasion into medistinal structures. The most common paraneoplastic syndrome associated with thymoma is myasthenia gravis which occurs approximately in 30 % of these patients. The disease is caused by autoantibodies to the nicotinic acetylcholine receptors at the neuromuscular junction. Maggi and associates [11] demonstrated a better prognosis for patients with MG, linked to an earlier thymoma diagnosis and less invasive thymoma in this group. Monden and colleagues [6] found a lower recurrence rate among patients with MG, even in cases with similar clinical stages. The diagnosis of MG can be made by detecting abnormally high levels of serum antibodies to the Ach receptors or by perfoming the tensilon test. Dushay *et al*. **[**12] has reported the first case of myasthenia gravis that presented with respiratory failure due to selecvtive involvement of respiratory muscles. In contrast to our patient, their patient showed lack of response to pyridostigmine and corticosteroid therapy. Surgical removal of all neoplastic thymic tissue is the mainstay of treatment. In patients with noninvasive well capsulated (stage 1 disease), the 5 year survival approaches 90%. The treatment of Thymoma MG consists of pyridostigmine titrated to an effective dose with addition of corticosteroids in poorly controlled patients. IV immunoglubin or plasma exchange can also be used. The beneficial effects of plasmapheresis are seen within days of initiating treatment. The second choice is immunosuppressive drugs whenever additional pharmacological treatment is needed before or after thymectomy. Immunosuppressive drugs, such as corticosteroids, azathioprine, cyclophosphamide, cyclosporine, methotrexate, mycophenolate mofetil, rituximab, and tacrolimus are generally used.

CONCLUSION

This is a case of Respiratory failure which is an uncommon presentation of thymoma, was managed on mechanical ventilation along with a surgical approach for resecting the tumour combined with plasmapheresis and immunosuppressive therapy leading to remission of symptoms.

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