

CASE REPORT

A RARE CASE OF THYMOMA WITH PURE RED CELL APLASIA WITH INITIAL REMISSION OF ANAEMIA AFTER THYMECTOMY

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ABSTRACT: Although thymoma is a common neoplasm of mediastinum, its association with pure red cell aplasia is very rare. It is also rare to observe the initial remission of anaemia of pure red cell aplasia after thymectomy only.

KEY WORDS: Thymoma, Red Cell Aplasia

INTRODUCTION: Thymoma is a common neoplasm arising in the anterior mediastinum. Red-cell aplasia is a rare disorder that results in a severe normochromic normocytic anaemia and it occurs only in 5 percent of patients with thymomas as a paraneoplastic syndrome¹. We report a case of thymoma associated with pure red cell aplasia (PRCA) where initial remission of anaemia occur after thymectomy only.

CASE REPORT: A 37 year old male presented with gradually progressive breathlessness and nonproductive cough for 6 months, dull aching chest pain for 4 months with generalised weakness and fatigue for last 3 months without any fever, hemoptysis, wheezing or hoarseness of voice. Although he got six units of blood transfusion within past 3 months but there was no history of haematemesis, melaena or any other bleeding manifestations. He was a teacher by profession without addiction to smoking and alcoholism.

Except severe pallor and dependent oedema general examination was normal. On examination of Respiratory system there was diminished chest movement (Left>Right), dull mediastinal percussion note and decreased vesicular breath sound over the anterior chest wall (Left>Right). Patient had no peripheral lymphadenopathy or hepatosplenomegaly. Examination of other systems were normal.

A chest roentgenogram of the postero-anterior and lateral view (Figure 1A & 1B) suggested well circumscribed homogenous round opacity occupying lower anterior mediastinum of left side. Computed tomography (CT) scan of the thorax (Figure 2A & 2B) showed the presence of an anterior mediastinum soft tissue mass which was suspected to be a thymoma.

His hematocrit was 13.7% with a hemoglobin concentration of 4.7 g/dl. His reticulocyte counts were persistently abnormal at 0.1%, although the white blood cell and platelet levels of her peripheral blood were normal. His serum iron level was 160 µg/dl (normal range, 50-150 µg/dl), total serum iron binding capacity was 267 µg/dl (normal range, 300-360 µg/dl), serum vitamin B₁₂ and folate concentrations were normal and erythropoietin level was 860 U/L (normal range, 10-25U/L). Although both direct and indirect Coombs' tests were positive, there was no evidence of hemolysis since the serum bilirubin level, urine urobilinogen concentration and red cell fragility were all normal. Bone marrow aspirate (Figure 3) showed severe erythroid hypoplasia associated with normal myeloid and megakaryocytic cell lines. The diagnosis of PRCA was made. CT guided

CASE REPORT

FNAC of the mediastinum lesion suggested benign spindle cell neoplasm. Bronchoscopy didn't reveal any intrabronchial pathology. Electromyography & Motor conduction velocity of all four limbs were within normal limit. After several units of blood transfusion left exploratory thoracotomy was done and a huge variegated well encapsulated extraparenchymal mass detected with pedicle superomedially towards mediastinum. Pedicle ligated and a well circumscribed lobulated, encapsulated tumour measuring 14.5 cm × 9.5 cm × 6 cm was dissected out. Histological examination (Figure 4) revealed a mixture of neoplastic epithelioid cells and non neoplastic lymphocytes. Diagnosis of thymoma was confirmed.

Although the patient received no further treatment for PRCA, his hematological test results rapidly improved. He was discharged from the hospital on the 22nd postoperative day with normal chest X-ray (Figure 5) and a hematocrit of 33.2%, a hemoglobin concentration of 11 g/dl and a peripheral reticulocyte level of 1.9%. The patient required no further transfusions. He is doing well, six months after surgery, without any recurrence of PRCA.

DISCUSSION: The incidence of thymoma is 19% among all mediastinal tumours². It occurs in between the ages of 40 and 60 years, with equal gender predilection. Two thirds of patients are asymptomatic at the time of diagnosis. The rest of the patients typically have nonspecific chest pain, cough, or dyspnea³. However, from 40% to 70% have at least laboratory evidence of one or more of the two dozen systemic "parathymic" syndromes that have been recognized². Myasthenia gravis is the most frequent among these but red-cell aplasia occurs only in 5 percent of patients with thymomas^{1,2}. Presenting clinical and radiological feature along with cytological report guided us regarding possibility of thymoma which later on confirmed by post operative histopathology report. The presented case had mixed lymphocytic and spindle cell variety of thymoma. Pure red cell aplasia was suspected in this case because of the presence of marked anaemia in a background of several units of blood transfusion along with presence of thymoma. It was confirmed after bone marrow aspiration cytology. The pathophysiology of PRCA is not completely known but may be related to abnormal T cell function and the presence of IgG antibodies which target erythroblasts and erythropoietin⁴. Usually 30 to 50 percent of patients with red-cell aplasia have thymomas¹.

Around 30% of cases initial remission of PRCA is found after thymectomy⁵. It works by the excision of autoreactive follicles. Complete surgical resection of the entire thymus with en bloc resection of all involved adjacent structures remains the mainstay of treatment of patients of thymoma and can be achieved in up to 88% of cases^{6,7}. However, in contrast to myasthenia gravis which is known to respond to thymectomy even in the absence of a thymoma, in PRCA there is currently no role of thymectomy in the absence of identifiable thymic mass⁸. Remission may require some additional therapy. Corticosteroids are often considered the first line of pharmacotherapy⁵. Other treatment regimens include azathioprine, erythropoietin, methotrexate, rituximab & antithymic globulin^{5,9}. Plasmapheresis or lymphocytapheresis and autologous and nonmyeloablative allogeneic peripheral stem cell transplantation have also been used¹⁰. Median survival of PRCA patients is approximately 12 years, although the prognosis and response to treatment are significantly related to associated disorders⁴. PRCA associated thymoma has a worse prognosis stage for stage than non PRCA thymoma¹¹.

CASE REPORT

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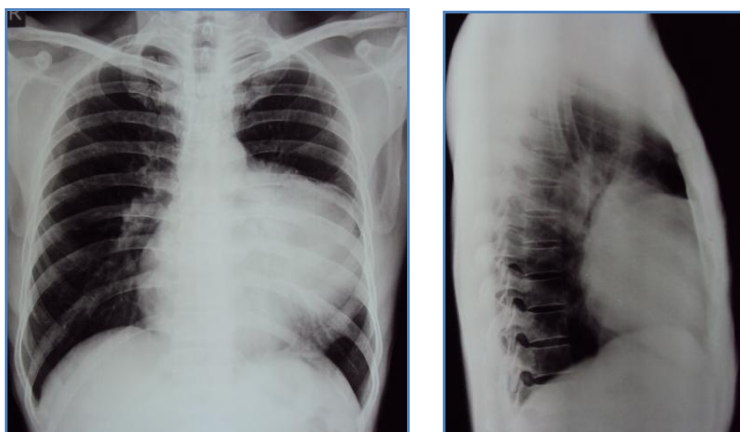


Fig. 1A & 1B: Well circumscribed homogenous round opacity occupying lower anterior mediastinum of left side.

CASE REPORT

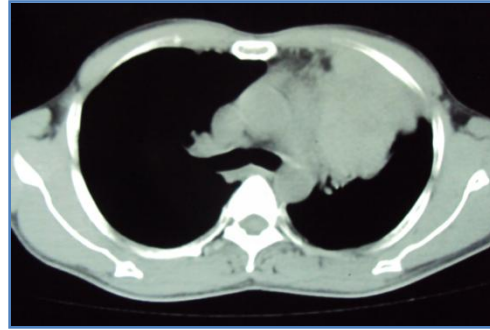
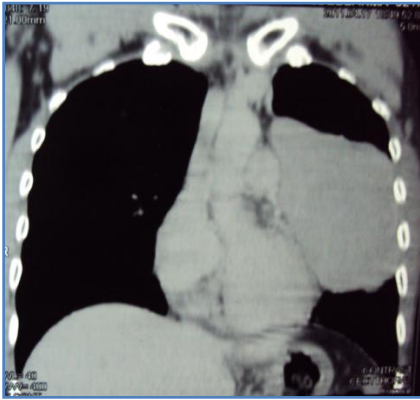


Fig. 2A & 2B: CT scan of the thorax showed the presence of an anterior mediastinum soft tissue mass.

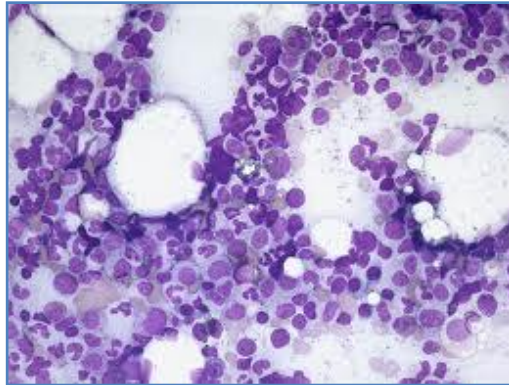


Fig. 3: Bone marrow aspiration cytology showed severe erythroid hypoplasia associated with normal myeloid and megakaryocytic cell lines.

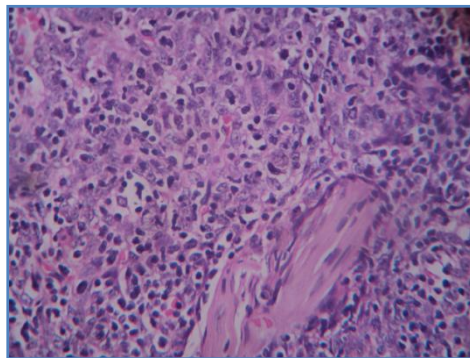


Fig 4: Histological examination of the mass revealed a mixture of neoplastic epithelioid cells and non neoplastic lymphocytes.

CASE REPORT

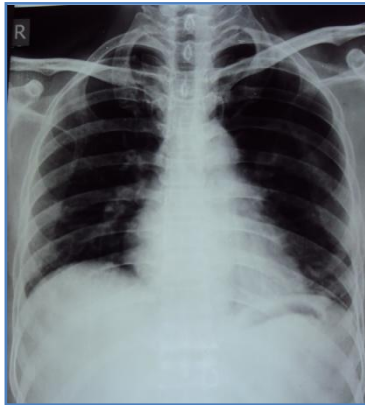


Fig. 5: Post operative chest X-

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