ANTERIOR MEDIASTINAL MASS AND PURE RED CELL APLASIA - A RARE CLINICAL ENTITY

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ABSTRACT

BACKGROUND
Thymomas are rare tumours with a worldwide incidence of 0.15 per 100,000 person-years.1 They usually occur after 40 years of age and have no particular sex predilection. They are associated with a variety of paraneoplastic syndromes like Myasthenia gravis, hypogammaglobulinaemia, Graves’ disease and pernicious anaemia. Pure red cell aplasia is rarely reported paraneoplastic syndrome associated with Thymoma. Its incidence is 10%.2

KEYWORDS
Pure Red Cell Aplasia, Thymoma, Anterior Mediastinal Man.


BACKGROUND
Case Presentation
A 34-year-old non-smoker male presented to Pulmonary Medicine Department, GMCH, Chandigarh with complaints of fever and shortness of breath for 2 - 3 months. On respiratory system examination there were reduced breath sounds in left infrascapular, infra-axillary and infra-mammary region. Routine blood investigations were within normal limits. Sputum staining was negative for acid fast bacteria. His chest radiograph depicted a homogeneous opacity in the left lower zone with positive Silhouette sign and obliteration of left costophrenic angle. Contrast Enhanced Computed Tomography of the thorax (CECT) scan revealed a well-defined lesion measuring 14.2 x 11 cms occupying the left lingular and the left lower lobe. His ultrasound abdomen revealed changes of congestive hepatomegaly. A trucut biopsy of the lesion was performed, which was inconclusive. Later, patient underwent thoracotomy and excision of the mass. A well-encapsulated mass weighing 2.15 kg was excised. Histopathological examination diagnosed it as encapsulated thymoma type AB. The thymoma was Masoka stage I, so no adjuvant therapy was administered to the patient. Post-operative period was unremarkable. After 5 months, patient complained of pallor and generalised weakness. On examination, there was no splenomegaly. Haemogram revealed haemoglobin of 4.6 with a reticulocyte count of 0.2%, total leucocyte count was 7600 and platelets were 2.76 lakhs. A bone marrow biopsy was performed, which revealed pure red cell aplasia.

Figure 1. Chest X-Ray Pre-Op

Figure 2. Chest X-Ray Post-Op

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DISCUSSION

Thymoma are tumours of thymic epithelial cells. They are usually indolent, but poses malignant potential for pleural dissemination, local invasion and rarely systemic metastasis. Thymomas are usually diagnosed by a biopsy or on histopathology. Cytokeratin is a useful marker for epithelial cells. Histologically thymomas are classified as per the WHO classification into type A, B, AB and C. It is staged as per the Masoka staging into four stages. Surgery is the cornerstone in the management of thymomas. Adjuvant therapies consisting of chemotherapy and radiotherapy are employed for invasive tumours. A 98% 5-year survival rate are reported after surgical resection of stage I and II thymomas. In one of the study out of 166 subjects of thymoma which were studied, only 6 subjects developed PRCA; 3 subjects before the resection of the tumour and 3 subjects developed it post-operatively. Due to its rarity, there are no definitive studies or protocols for its management. Specific therapy is indicated with systemic steroids or cyclosporins and other agents. The efficacy of these treatments is not known. The presentation of thymoma with PRCA has been associated with a bad prognosis. The rare incidence, pure red cell aplasia occurring after the resection of thymoma, pure red cell aplasia being associated with thymoma type AB are some of the fields that needs to be addressed. However, more studies are required to postulate a definitive management strategy for this rare clinical entity.

CONCLUSION

Pure red cell aplasia occurring after the resection of thymoma. The present case reinforces the need for clinicians to be vigilant with thymoma patients, even following thymectomy. However, more studies are required to postulate a definitive management strategy for this rare clinical entity.

REFERENCES