A 43-year-old man, a known case of β-thalassaemia, intermediate type, from southern Iran, presented with a one-month long history of breathlessness and severe chest pain that was radiating to his back. He had undergone splenectomy 20 years back. On admission, his pulse rate was 88/min, respiratory rate 20/min, and blood pressure 155/80 mm Hg. The breath sounds were decreased and basal crackling rales were heard bilaterally. An apical 2/6 systolic murmur could be heard. His liver was palpable 2 cm below the right costal margin. The haemoglobin concentration was 10.5 g/dl (Hb A$_2$: 3.2%, and Hb F: 67%) with microcytosis, hypochromasia, anisocytosis, poikilocytosis, polychromatophilia and reticulocytosis evident on the peripheral smear. Chest roentgenogram showed multiple large intra-thoracic masses, the largest being located in the right lower lung field. There were also masses in left retro-cardiac space, in the superior mediastinum, para-vertebral region and infero-lateral aspect of the left lung. Ribs demonstrated widening, coarse trabecular pattern and osteopenia (Figure 1). Axial computed tomography (CT) scan of the lower chest showed bilateral para-vertebral and extra-pulmonary lobulated masses with soft-tissue density near the expanded ribs (Figure 2). Sonography-guided fine needle aspiration of the masses yielded highly cellular tissue consisting of cells resembling bone marrow cells that included myelocytes, lymphocytes, monocytes, segmented neutrophils, megakaryocytes, and normoblasts. The patient was diagnosed to have extra-medullary haematopoiesis (EMH) that was treated with low-dose radiation therapy. His symptoms of severe chest and back pain, limitation of movement and breathlessness subsided after receiving a cumulative radiation dose of 1500 cGy administered over 12 sittings. The chest radiograph done one month later demonstrated complete disappearance of the masses.

**Discussion**

The bone marrow and extra-medullary sites attempt to increase the production of erythrocytes to compensate for the chronic haemolytic anaemia present in individuals with thalassaemia. The EMH is commonly seen at sites such as the abdomen, chest, or epidural space.\(^1\) EMH accompanies a wide variety of diseases including thalassaemia, sickle cell anaemia, polycythemia rubra vera, chronic myelogenous leukemia, agnogenic myeloid metaplasia, and hereditary spherocytosis.\(^3\) There are two forms of EMH namely “para-osseous”— in which the normal medullary tissue of the bone marrow ruptures through the bone to present as a para-osseous mass, and “extra-osseous”— in which EMH occurs within soft tissue. Para-osseous EMH occurs more frequently in haemoglobinopathies whereas extra-osseous EMH accompanies predominantly with myelopro-
Patients with thalassaemia intermedia are not usually treated with regular transfusion therapy as the medullary and EMH are able to maintain haemoglobin concentration above a reasonable level, in contrast to individuals with thalassemia major whose life is dependent upon regular transfusion therapy. Consequently, subjects with thalassemia intermedia demonstrate severe grade of manifestations referable to medullary and extra-medullary haematopoiesis such as haemolytic facies and severe hepato-splenomegaly.

EMH can be prevented by the institution of regular transfusion therapy, which corrects anaemia and thereby, abolishes the stimulus for EMH. Surgical decompression has been the method of choice for the management of the disease because histological diagnosis can be established, and immediate decompression of the mass can be achieved. This is especially important in rapid decompression of spinal cord in patients with epidural mass caused by EMH. The disadvantages of surgical intervention include risk of excessive bleeding due to high vascularity of the mass and higher incidence of recurrence. Furthermore, total resection of the mass can lead to clinical decompensation and deterioration since these masses play a crucial role in maintaining an adequate haemoglobin level. To avoid this, incomplete resection of the mass can be attempted, followed by low-dose radiation therapy. Being very sensitive to radiation therapy, low doses of radiation (1000–3000 cGy), bring about a good clinical response. The major disadvantages of radiation therapy are the lack of histological diagnosis and the reduction in the bone marrow activity secondary to the procedure itself. Hydroxyurea, a myelosuppressive agent, has also been successfully employed in the management of EMH. The differential diagnosis of the posterior mediastinal locations of EMH includes neurogenic tumours, lymphoma, paravertebral abscess and metastatic carcinoma. In geographic areas where thalassaemia is prevalent, EMH should be considered in the differential diagnosis of patients who have chronic anaemia with an intrathoracic mediastinal mass.

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References