X-Ray Quiz

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This is a 2-year-old boy suffering from poor weight gain and unexplained iron deficiency anaemia. He had no recent infections and is afebrile. Physical examination showed a small sized child with no dysmorphic facial features. He was stable in room air with no respiratory distress. He was tachycardiac with regular heart rate of 120 beats per minute. There was no jaundice, no clubbing, no rash, but marked pallor, eczema and multiple shotty cervical lymph nodes. Chest, cardiovascular and abdominal examinations were unremarkable with no organomegaly.

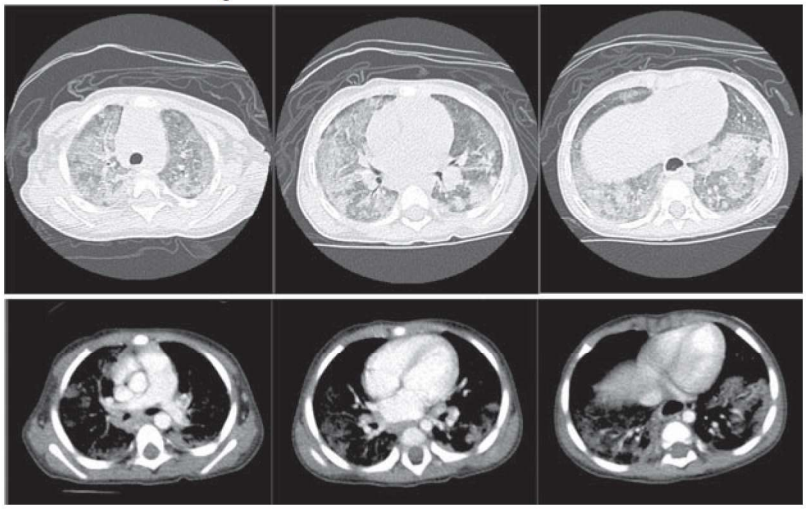
Significant initial investigation results were as follows:
- Haemoglobin level: 4.6 g/dL; Hb pattern: no haemoglobinopathy
- LDH 412 IU/L (normal); Haptoglobin 0.51 g/L (normal)
- Iron: 2.8 umol/L (low); Total iron binding capacity: 84.7 umol/L (high); Saturation 3%; Ferritin 53 pmol/L (normal)
- Faecal occult blood: positive for one sample

Extensive workup covered sources of gastrointestinal bleeding, including Meckel's scan, oesophageogastroduodenoscopy, ultrasound abdomen and red cell scan were unremarkable. Three more faecal occult blood samples came back to be negative.

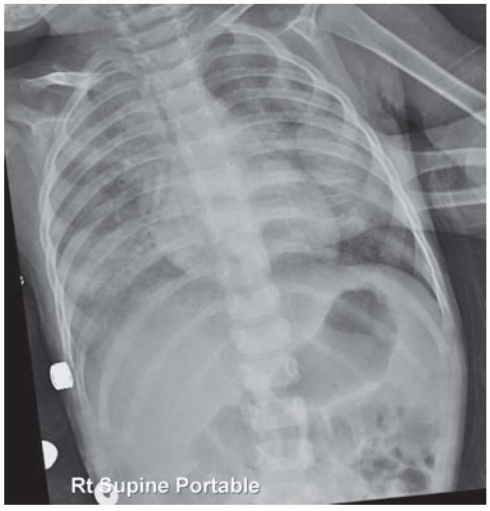
Haemolysis workup was performed twice over 3 months were unremarkable. CRP was raised at 10.6 mg/L. There was marked reticulocytosis and eosinophilia. Bone marrow aspiration showed active bone marrow with no infiltrative changes but absent iron stores.

Infection screening for Parvovirus DNA PCR, Tuberculosis, HIV, TORCH, Histoplasmosis and Yersinia were all negative. Despite iron supplements and five blood transfusions given over 9 months, the boy still showed recurrent anemia with lowest Hb 4.5 g/dL. CT abdomen showed non-specific terminal ileum short segment mural thickening, likely of inflammatory or infective changes, as well as incidental finding of consolidative lung changes.

CT thorax with contrast was performed in view of the incidental CT abdomen findings and is shown below:



Retrospective review of chest X-ray on admission was as follows:



Questions

1. What are the Contrast CT Thorax and Chest X-ray abnormalities?
2. What is the radiological diagnosis?
3. Base on the clinical history and radiological findings, what is the patient's diagnosis?

(Answer on page 17)

Answers to X-ray Quiz on page 15

1. CT anomalies:
   a) Bilateral diffuse consolidative changes and ground glass opacities with prominent lymph nodes are noted at right paratracheal, bilateral hilar and subcarinal regions
2. Bilateral diffuse pulmonary infiltrate which can be haemorrhage or infective changes
3. Pulmonary Haemosiderosis

The patient was diagnosed to have pulmonary haemosiderosis. Pulmonary haemosiderosis is a rare disease entity caused by recurrent diffuse alveolar haemorrhage, and is predominantly found in the paediatric population.[1] Haemoptysis, iron-deficiency anaemia and pulmonary infiltrates with haemosiderin-laden macrophages are the most salient features of the disease. Chest X-ray (not mentioned above) typically show a "butterfly" or "batwing" pattern.

Bronchoscopy and lung biopsy is considered the gold standard for diagnosis. Transbronchial and transthoracic lung biopsy was sent for histopathology to rule out vasculitic pulmonary disease. Bronchoalveolar lavage (BAL), gastric lavage (GL) was also sent for haemosiderin laden macrophage. Both BAL and GL came back to be positive for the pathognomonic haemosiderin laden macrophages. There was no evidence of occult infections like Pneumocystic carini or tuberculosis from BAL specimens.

Known trigger factors for pulmonary haemosiderosis include gluten proteins, moulds and cow's milk proteins.[2] Associated disease entities include collagen vascular diseases, Wegener's granulomatosis, Henoch-Schonlein purpura, Goodpasture syndrome and coeliac disease.

Prognosis is highly variable but has been vastly improved with the advent of immunosuppressants and corticosteroids, though comorbidities including rheumatoid, coeliac disease and other respiratory problems are common in survivors.[3-5] Blood transfusion and intravenous methylprednisolone is the mainstay of treatment for acute symptomatic haemorrhage.[1] Oral use of regular corticosteroids and/or immunosuppressants[5] is followed when the acute haemorrhage is controlled.

References

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