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Hypereosinophilia with an anterior mediastinal mass

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Hypereosinophilia with an anterior mediastinal mass (9153)

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Background

- HES characterized by blood eosinophilia of 1500 EOS/mL with evidence of end-organ damage attributable to eosinophilia and no other cause.
- Rare in adults but even more rare in children
- Causes are divided between primary or clonal versus secondary or non-clonal including allergic disease, infections, drug reactions

Case presentation

- A 3-year-old male with questionable history of asthma was admitted for newly found anterior mediastinal mass and left pleural effusion.
- Past surgical history: none.
- Family history: no family history of eosinophilic disorders.
- Medications: albuterol, Flovent
- CT chest showed large left pleural effusion, possible abscess, small pericardial effusion, and concern for an anterior mediastinal mass due to mediastinal shift.

Case presentation continued

- The patient's absolute eosinophil count suddenly increased from 600 to 25,048 and remained elevated above 20,000.
- He developed recurrent eosinophilic pericardial effusions requiring multiple pericardiocentesis.
- Despite high dose steroids and inpatient use of mepolizumab, his hypereosinophilia persisted.
- After two inconclusive fine-needle aspirations of his mediastinal mass, the patient's family agreed to a sternotomy with removal of the mediastinal mass.
- Pathology eventually revealed myeloid sarcoma, a variant of acute myeloid leukemia.

Figure 1: Coronal view of mass

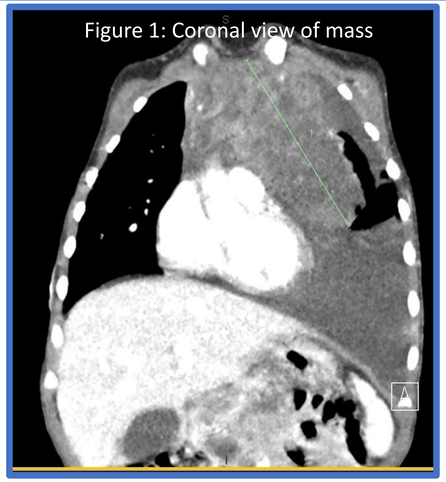
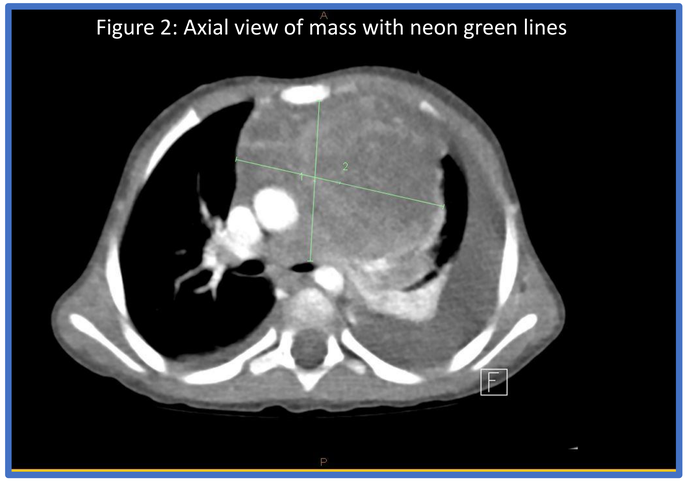


Figure 2: Axial view of mass with neon green lines



Management

- High-dose steroids in 1 mg/kg/day in children and adults
- Patients with MHES and LHES are least responsive to corticosteroids
- For those with FIP1L1-PDGFRA gene, imatinib is effective.
- Mepolizumab approved for 12 years old and greater with dose of 300 mg every 4 weeks particularly in patients who were FIP1L1-PDGFRA negative.

Conclusions

- Since HES is so rare, mainly case reports are available.
- Steroid resistant HES should raise concern for underlying malignancy especially when eosinophils are above 20,000.

References

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