

Abstract

Hereditary Haemorrhagic Telangiectasia (HHT) is an autosomal dominant disease with mucocutaneous telangiectasias and visceral arteriovenous malformations (AVM). The major visceral bleeding occurs due to AVMs in lungs, brain and liver. This patient had a life threatening bleed which needed intensive care.

Case presentation

A 41 year old patient with previous history of recurrent epistaxis presented with acute worsening of left sided chest pain and shortness of breath with left sided chest wall swelling for 3 days duration. On admission he had low oxygen saturation with poor breathing effort. He also noted to have multiple mucocutaneous telangiectasias involving bilateral hands, tongue, oral cavity and mucus membrane of the nasal cavity. Arterial blood gas showed type 2 respiratory failure. He was intubated and ventilated. CECT Chest, MRI Chest showed disseminated bleeding into left side of the chest wall, Pectoralis major and pectoralis minor muscles extending to insertion of muscles and superior mediastinal haematoma. Rigid nasal endoscopy revealed multiple sinonasal telangiectasias. Upper and lower gastrointestinal endoscopy was normal. His ICU stay was complicated with atrial fibrillation and left lower and lingular lobe pneumonia. He recovered with the supportive management and discharged on day 14 with further follow-up plan and family screening.

Conclusion

Hereditary Haemorrhagic Telangiectasia is a well known disease which complicates by visceral bleeding manifestation due to arteriovenous malformations. This case adds a different perspective to the disease, complicated with chest wall haematoma and mediastinal haematoma. Treatment is directed as palliation but effective preventive treatment modalities needs to be further evaluated.

Key words

Hereditary Haemorrhagic Telangiectasia, HHT, Arteriovenous malformations, mediastinal haematoma