Spontaneous haemothorax in Osler-Weber-Rendu disease

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Summary

A case of hereditary haemorrhagic telangiectasia (Osler-Weber-Rendu disease) is described who presented with severe, central chest pain mimicking acute myocardial infarction, a presentation which has not been described before. He was found to have developed spontaneous haemothorax which is a very rare complication of this disease.

KEY WORDS: Osler-Weber-Rendu disease, chest pain, spontaneous haemothorax.

Introduction

Hereditary haemorrhagic telangiectasia (Osler-Weber-Rendu disease) is an uncommon disease and, in over 98% of cases, the patients present with recurrent episodes of epistaxis (Harrison, 1964). It has been estimated that 15% of patients develop a pulmonary arterio-venous fistula during their lives (Hodgson *et al.*, 1959). The patient with a pulmonary arterio-venous fistula may suffer from various complications. According to Dalton *et al.* (1967), bleeding with haemoptysis occurs in approximately 25% of cases. Spontaneous haemothorax as a complication in Osler-Weber-Rendu disease is, however, a very rare event. We describe here a patient who presented with severe central chest pain due to spontaneous haemothorax.

Case report

A 37-year-old Kuwaiti male suddenly developed severe retrosternal pain at rest. The pain was stabbing in nature and radiated towards his left shoulder.

Examination revealed a well-built, normotensive man who was in severe pain, febrile and very restless with impaired resonance and decreased breath sounds on the left but no shift of trachea or apex beat was detected.

Telangiectases on his lips, tongue and palate were

noted. On further inquiry, he admitted to having had recurrent episodes of epistaxis in the past and bleeding from the tongue. It was also found that 2 of his daughters and one son also suffered from recurrent epistaxis. It became obvious that the patient had Osler-Weber-Rendu disease, but it was not clear how the sudden onset of chest pain, the chest signs, fever and leucocytosis (white cell count $20 \times 10^9/\text{litre}$, mainly polymorphs) were related to this diagnosis. Chest X-ray showed a diffuse opacity occupying whole of left hemithorax and 50 ml of blood was aspirated from the pleural space.

Next day, he was transferred to the thoracic surgery unit where 600 ml of blood was aspirated from the left pleural space. Antibiotics were continued and he was transfused with one pint of whole blood. On the 13th December, he again complained of chest pain and shortness of breath. Another 350 ml of blood was aspirated from the pleural space. Next day he again complained of severe retrosternal pain and became sweaty, breathless and restless. A number 28 Malecot catheter was inserted in the 7th left intercostal space and 1000 ml of blood was drained. He was given one more pint of blood. After this episode, however, he followed an uneventful course and improved gradually. Sixteen days after admission, he was afebrile and chest X-ray showed no evidence of haemothorax. He was discharged home on ethinyloestradiol, 0.05 mg, and methyltestosterone, 0.5 mg per day, along with iron and vitamin supplements.

Unfortunately, the patient refused to have pulmonary angiogram, but as the plain chest X-ray was normal, it may be presumed that he did not have a large arterio-venous malformation.

Conclusion

Hereditary haemorrhagic telangiectasia is an uncommon disease of unknown aetiology. The clinical

diagnosis is based on the triad of a history of recurrent bleeding, presence of multiple telangiectases, and a history of familial occurrence (Harrison, 1964).

Most commonly, these patients present with a history of recurrent bleeding. Epistaxis is especially common, but bleeding may originate from the telangiectases wherever they are: tongue, lips, respiratory, gastrointestinal or urinary tract. Haemorrhage in brain and retina has been attributed to the presence of telangiectases, but telangiectatic lesions have not been demonstrated in all instances (Wintrobe *et al.*, 1974). A large number of central nervous system symptoms such as headache, convulsions, loss of consciousness, mental confusion or transient sensory, motor, visual or speech disturbances have been attributed to telangiectases in the brain (Hodgson *et al.*, 1959). In an occasional patient, diffuse involvement of the hepatic and splenic vessels may produce hepatosplenomegaly (Bithell and Wintrobe, 1977).

Patients with hereditary haemorrhagic telangiectases may have associated vascular lesions in other viscera. Aneurysms of the splenic and hepatic arteries and arteriovenous fistulas of the retinal, cerebral and hepatic vessels have been described (Wintrobe *et al.*, 1974). A patient has been described who first presented with brain abscesses and was later found to have hereditary haemorrhagic telangiectases (Harkonen, 1981). Pulmonary arterio-venous fistula is not an uncommon association with this disease and may present with haemoptysis, recurrent pulmonary infections or significant cardiovascular features including dyspnoea, cyanosis, secondary polycythaemia and murmurs of various types (Stringer *et al.*, 1955).

Although a review of literature shows that these patients can present with almost any symptom, presentation with severe, central chest pain mimicking acute myocardial infarction has never been reported. The cause of chest pain in our patient was spontaneous haemothorax, which is a very rare complication of this disease. Dalton *et al.* (1967) found that in contrast to intrabronchial rupture which produces haemoptysis and occurred in approximately 25% of cases, intrapleural rupture of the aneurysm was a rare event and had been reported on only 8 previous occasions. Haemothorax may result from rupture of a tiny telangiectatic vessel on the pleural surface (Bowers, 1936; Livingston and Carr,

1956) but in some other reported cases it appears to have been due to rupture into the pleural space of a fairly large pulmonary arterio-venous fistula (Erf *et al.*, 1949; Armentrout and Underwood, 1950; Heyde, 1954; Moore, 1969).

Although epistaxis is the commonest presentation of the hereditary haemorrhagic telangiectasia, various unusual presentations and complications of this uncommon disease should be kept in mind.

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