Life threatening haemoptysis in cystic fibrosis: an alternative therapeutic approach

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Abstract

Desmopressin and vasopressin were used to control massive haemoptysis in a patient with cystic fibrosis. After bolus doses a continuous infusion of vasopressin was maintained for 36 hours and haemoptysis stopped.

Small recurrent haemoptyses are common in patients with cystic fibrosis.[1] During seven years in a clinic of 150 patients only three presented with severe bleeding requiring aggressive intervention. Conservative management consists of blood transfusion, correction of impaired clotting, provision of oxygen and positioning the patient head down and lying on the side of the bleeding lung. Intravenous antibiotics should also be started. More invasive management consists of fibreoptic bronchoscopy to locate the site of bleeding followed by embolisation of the appropriate bronchial artery,[2] although recurrent bleeding may follow embolisation.[3] Furthermore, bronchoscopy may be difficult if the patient is very breathless and it often fails to locate the site of bleeding.[4] Facilities for rigid bronchoscopy to maintain airway patency and prevent asphyxiation may not be immediately available, as required by such a life threatening condition, and bronchial artery embolisation is not available in many hospitals. If bleeding can be controlled temporarily the patient should be transferred to a thoracic centre for consideration of bronchial artery ligation or lobectomy, though most adult patients have poor respiratory function and would be unfit for surgery.

We report a further conservative measure to control profuse haemoptysis in a critically ill patient with poor lung function.

Case report

A 23 year old man with cystic fibrosis presented with massive recurrent haemoptysis. He had attended Monsall Hospital for seven years and had had repeated admissions for infective exacerbations requiring intravenous antibiotics. His compliance with physiotherapy and outpatient attendance had always been poor. Despite the severity of his lung disease (FEV_1 1·4 l, vital capacity 2·2 l) he had managed to work full time in a sedentary job. When he was first referred (1982) he had hepatosplenomegaly and in 1984 was found to be anaemic. An abdominal ultrasound examination and barium swallow and meal confirmed portal hypertension and oesophageal varices. He had a blood transfusion but refused treatment of his varices. In 1987 hypogonadotrophic hypogonadism was diagnosed. In 1988 he presented with collapse after a large haematemesis and melaena. His haemoglobin was 5·4 g/dl. He received six units of blood and at gastroscopy grade 4 varices and a small duodenal ulcer were found. The varices were sclerosed on four occasions. In December 1989 he presented with further haematemesis requiring sclerosis of his varices. He refused intravenous antibiotics for a concurrent chest infection.

Two weeks after discharge he reported coughing up large amounts of bright red blood. On admission he was hypotensive and breathless. His haemoglobin concentration was 11·2 g/dl, platelet count 8·2 × 10^9/l, and prothrombin time 1·4 seconds. The bleeding settled; he had a transfusion of one unit of blood and was given intravenous vitamin K and oxygen, but 12 hours later he had a fresh haemoptysis of 650 ml. The bleeding continued; he became cyanosed, breathless, hypotensive, and preterminally ill. He was given 4 μg of intravenous desmopressin with immediate cessation of the bleeding. Eight hours later he had further haemoptysis of 500 ml and was given an intravenous infusion of vasopressin 20 units over 15 minutes. The acute bleeding ceased; in view of this success an infusion of vasopressin 0·2 units/min was started and continued for 36 hours. No further bleeding occurred but he continued to produce small amounts of darker blood mixed with sputum. Intermittent abdominal cramp, pallor, and chest tightness, relieved by nebulised bronchodilators, was noted. Diuretics were given to prevent fluid overload. After 14 days' intravenous antibiotic treatment he was discharged with no further haemoptysis.

Discussion

The initial management of massive haemoptysis may present a considerable clinical challenge in a district hospital. A recent editorial suggested guidelines but concluded, "In the management of massive haemoptysis no hard and fast rules have been proved to apply."[5] Subsequent correspondence and a review of management approaches confirmed the diversity of opinion.[6 7]

Owing to the lack of local facilities for performing embolisation we decided not to locate the site of bleeding by fibreoptic bronchoscopy. We fortuitously used two vasopressor agents to control the severe haemoptysis. The fact that vasopressin was not at first available led us to use desmopressin. Although its effect on smooth muscle is less than that of vasopressin it shortens bleeding time and decreases blood loss in patients undergoing cardiac surgery.[8] It probably acts by increasing the concentration

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Accepted 2 October 1990

of factor VIII and it also releases plasminogen activator from endothelial cells.[9]

Vasopressin has been used to control bleeding from oesophageal varices. Its plasma half life is about 24 minutes and it is most effective when given by infusion. The site of action is probably arteriolar smooth muscle, through an increase in the intracellular concentration of inositol phosphates, which mobilise intracellular calcium, causing contraction. The bronchial and mesenteric arteries both arise directly from the aorta. We hoped to reproduce the effect of pressor agents on the mesenteric vasculature in the bronchial circulation. The effect of the pressor agents in stopping pulmonary bleeding may have been fortuitous; but the immediate termination of profuse bleeding on separate occasions, initially with desmopressin and subsequently with a bolus and infusion of vasopressin, was impressive. We are not aware of any publications describing the action of pressor agents on the bronchial circulation in either man or animals (personal communication from Parke Davis).

There was no difficulty in distinguishing between a large haemoptysis and a haematemesis in this patient. He was observed to cough up a large amount of blood and was grossly breathless with a much smaller reduction in haemaglobin than when he had previously been admitted for haematemesis.

Severe haemoptysis in chronic lung disease is uncommon and pressor agents should not be used routinely owing to the side effects of water retention and bronchoconstriction. They may, however, have a useful conservative role in the management of patients with cystic fibrosis who have severe lung and liver disease.

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Thorax 1990;**45**:976–978

Intercostal arteriovenous fistula due to pleural biopsy

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Accepted 11 June 1990

Abstract

A 32 year old woman had a pleural biopsy for a left pleural effusion, which showed caseating granuloma typical of tuberculosis. When the fourth biopsy specimen was removed considerable bleeding occurred from the puncture site. Four days later a bruit was audible over the punctured area, radiating to the back. Eight days after the procedure the patient had a massive bleed into the left pleural space. Selective aortic angiography showed an arteriovenous fistula between the 9th intercostal artery and vein and a pseudoaneurysm in the intercostal punctured area. Thoracotomy showed bleeding from the site of the pleural biopsy. The intercostal vessels were ligated and pleural decortication was performed, and the patient recovered uneventfully.

Recently we encountered a previously unreported complication of closed pleural biopsy—namely, the occurrence of a traumatic arteriovenous fistula of the intercostal artery and vein.

Case report

A 32 year old woman was admitted with fever (38·3°C) and a productive cough. There was no history of previous lung disease, chest trauma, or excessive bleeding. She was very slim (36 kg). Physical examination disclosed nothing abnormal apart from dullness to percussion and decreased breath sounds over the left lower lung field. Diagnostic thoracocentesis was performed and 20 ml yellowish serous fluid removed.

The next day a closed pleural biopsy was performed through the 9th intercostal space at the posterior axillary line with a Cope needle. Before biopsy the patient appeared tense and fearful. After injection of a local anaesthetic three pleural specimens were obtained without incident; but when a fourth specimen was