Case report

Vascular amyloidosis causing spontaneous mediastinal haemorrhage with haemothorax

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Abstract

Bleeding diathesis is a recognised complication of amyloid disease. Localised and generalised bleeding manifestations are usually associated with intravascular coagulopathy related to isolated or multiple coagulation factor deficiencies. Recently, there have been reports of haemorrhage due to amyloid deposition in blood vessel walls and in the perivascular region leading to increased fragility and poor haemostasis. We report a case of spontaneous mediastinal haemorrhage due to amyloid involvement of vascular tissue in the absence of coagulopathy.

Keywords: Amyloidosis; Haemorrhage; Bleeding diathesis

1. Introduction

Bleeding diathesis is a recognised complication of amyloid disease. Localised and generalised bleeding manifestations are usually associated with intravascular coagulopathy related to isolated or multiple coagulation factor deficiencies. Recently, there have been reports of haemorrhage due to amyloid deposition in blood vessel walls and in the perivascular region leading to increased fragility and poor haemostasis.

We report a case of mediastinal haemorrhage due to amyloid involvement of vascular tissue in the absence of coagulopathy.

2. Case report

A 50 year old housewife presented to the Accident and Emergency Department of a local hospital with a 4-day history of spontaneous sudden onset of sharp central chest pain and a 24-h history of neck swelling associated with progressive dysphagia. There were no precipitating events, predisposing factors or any history of trauma. The only past medical history was of a thyroidectomy for benign disease 13 years ago.

On examination she was haemodynamically stable; however, there was diffuse tender neck swelling with supra-ternal bruising. The ECG was normal and all blood indices were within normal limits except the haemoglobin of 10.3 g/dl; platelets were $318 \times 10^{12}$/l and the clotting screen was unremarkable (International Normalised Ratio normal and Activated Partial Thromboplastin Time normal). A chest radiograph revealed a widened mediastinum with bilateral basal effusions. Soft tissue cervical radiography revealed extensive retropharyngeal soft tissue swelling.

The patient was referred to the otorhinolaryngologists and a diagnosis of a large retropharyngeal abscess was made. She was taken to theatre, where a large haematoma was evacuated from the root of the neck, a tracheostomy performed and the retropharyngeal space drained. A more detailed haematological investigation failed to identify a haematological cause for bleeding. Angiography was carried out and revealed a normal aortogram with no obvious source of haemorrhage.

The patient gradually developed signs of early superior vena caval obstruction and worsening oxygenation with increasing bilateral pleural effusions. Computerised axial tomography revealed mediastinal and intrapleural blood (Fig. 1).

Referral was made to the thoracic surgeons and, after evaluation, an emergency endoscopy and right thoracotomy was undertaken. Bronchoscopy showed proximal submuco-
haemorrhage was due to amyloidosis. Despite further investigation, including serum electrophoresis to search for the presence of an underlying pathlogy such as multiple myeloma, no aetiological factor has been identified to date.

3. Discussion

Amyloidosis is a disorder of protein metabolism in which there is extracellular deposition of insoluble fibrillar protein either localised or widely distributed throughout the body. It may be sporadic in nature or reactive, and thus be secondary to a long-standing chronic infection, inflammation or malignancy.

The relation between amyloid disease and bleeding tendency has long been recognised. The underlying pathogenesis is varied, and may be related to malignancy-induced bone marrow depletion resulting in thrombocytopaenia, or isolated/multiple coagulation factor deficits. Recently there have been several reported cases of bleeding in amyloidosis secondary to infiltration of perivascular and vascular tissues in the absence of clotting defect [1].

Yood et al. [2] documented a series of 100 patients with amyloidosis. They reported that in 41% of cases the patients experienced one or more bleeding episodes, with fatal haemorrhage occurring in three cases. Forty-five percent of cases had abnormalities of clotting; however, there was no correlation between coagulation incompetence and incidence of haemorrhage. The high prevalence of observed bleeding tendency despite absence of coagulopathy was seen to suggest that the majority of bleeds were due to amyloid infiltration of vessel substance and not purely due to secondary clotting deficit. Such amyloid infiltration would presumably lead to increased fragility of blood vessels and a decreased ability for healing.

There have been many reports of unusual bleeding manifestations associated with amyloidosis including subconjunctival haemorrhage [3] and macroscopic haematuria [4]. A focal form of amyloidosis localised to the larynx has been reported to cause a fatal upper respiratory tract haemorrhage [5], gastrointestinal tract amyloidosis has been documented on several occasions to cause spontaneous upper and lower gastrointestinal haemorrhage [6–9], and Shaheen et al. [10] published a case of fatal bronchopulmonary haemorrhage due to unrecognised amyloidosis.

In the case we report that there were no obvious risk factors for coagulopathy and full clotting screen could identify no known pathology. The operative findings indicated an active bleeding process and the lack of focal pathology, combined with the histological findings, support the hypothesis of amyloidosis-induced spontaneous mediastinal haemorrhage due to perivascular and vascular wall involvement.

The wide variety of cases already reported suggests that such bleeding problems may well affect any organ of the body and thus, although rare, amyloid angiopathy must be
included in the differential diagnosis of spontaneous haemorrhage with no immediately identifiable cause.

References


