CASE REPORT

Acquired haemophilia A presenting at a District General Hospital

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SUMMARY

We report a case of a 64-year-old man presented to A&E with a 2-week to 4-month history of right hip pain, right leg weakness and fasciculations, extensive bruising and a vesicular skin rash. He had a CT of the chest/abdomen/pelvis, which revealed multiple extensive haematomas including an iliopsoas haematoma causing a lumbar plexopathy and resulting in the right hip and leg symptoms. He had clotting studies showing a prolonged activated partial thromboplastin time. Haematology review together with mixing studies suggested a diagnosis of acquired haemophilia A. He was treated at the local tertiary centre with activated prothrombin complex concentrate and steroids and made a full recovery.

BACKGROUND

Acquired haemophilia has an incidence of 1–2 per million.[1] This presentation was classical of acquired haemophilia but the condition is not seen frequently at the presenting hospital. The diagnosis was made rapidly using general medical knowledge and appropriately utilising specialist resources.

Despite presenting at a District General Hospital, this patient received excellent care. By utilising basic knowledge of anatomy and physiology, the team and the available specialists managed to diagnose the condition within 24 h and transfer the patient to the tertiary centre within 36 h. This was also a fantastic example of good communication within and between healthcare teams.

CASE PRESENTATION

We report a case of a 64-year-old man with a history of hypertension, for which he takes ramipril, presented to A&E at Newham University Hospital with symptoms of leg weakness, rash and bruising. The rash and leg weakness began approximately 4 months prior to this presentation. He cannot remember any obvious trigger. The rash extended over his lower legs and was vesicular in nature. The natural history of the rash was formation of blood-filled vesicles, which would burst and heal leaving a temporary scar. The leg weakness began around the same time as the appearance of the rash and had been progressive in nature, affecting only his right leg and not the left. The weakness was associated with fasciculations of his right thigh. In addition to the 4-month history of weakness and fasciculations, he also reported a 2-week history of worsening right leg pain. He attended A&E for this pain 2 weeks previously and was discharged home with analgesia following negative investigations. On this occasion, he called an ambulance because the pain was so severe that he was unable to move. Furthermore, since the A&E attendance he developed extensive bruising over his whole body and bleeding from his gums. This bruising was not associated with any trauma that he could recall.

He does not drink alcohol and has never smoked. He lives alone and prior to this episode he was fully independent. Systems review revealed no further symptoms other than mild constipation related to tramadol use for his leg pain. He had had no recent foreign travel and no other change in his regular medications.

On examination, he appeared pale. He was tachycardic at 120 bpm. All other observations were within normal limits. Extensive ecchymoses were clearly visible over large areas of his face, trunk and limbs. Other than the bruising, abdominal and respiratory examinations were entirely normal. There were vesicles at varying stages of resolution visible over both legs. Hip flexion power was reduced to 3/5 on the right and 4/5 on the left, although he reported no subjective left leg weakness. There was also increased tone and hyper-reflexia on the right leg. Neurological examination was otherwise normal.

INVESTIGATIONS

Blood tests showed that since his A&E attendance 2 weeks previously, his haemoglobin had fallen from 120 to 60 g/L. His platelet count was normal and white cells were raised at 18.5 (the differential showed a neutrophilia of 15.7). Clotting screen revealed a normal international normalised ratio but a prolonged activated partial thromboplastin time (APTT) of 46. D-dimer was 8.8. C reactive protein was raised at 47.8 on admission, which then rose to 176 2 days later. Electrolytes and liver function tests were normal. A large number of other investigations were carried out, which were normal.

CT chest/abdomen/pelvis revealed iliopsoas haematoma as well as several other moderate-sized haematomas throughout the chest, abdomen and pelvis. Finally, mixing studies were requested including APTT 50:50 mix, which showed no correction of APTT on mixing with control plasma.

DIFFERENTIAL DIAGNOSIS

It was immediately evident that the patient had disordered coagulation. Following discussion with the haematology team it was felt that acquired haemophilia A was most likely due to the features of the presentation including the age of the patient. Hereditary haemophilias and other acquired

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Learning points

Effective communication between healthcare professionals is vital and in this case resulted in timely management of a potentially life-threatening condition.

- Referrals to the appropriate specialists were prompt and clear and the subsequent reviews and clinical decisions were communicated effectively between teams.
- Clear communication with the laboratory staff was required to ensure the correct specialist tests were performed.
- Liaison with pharmacy was necessary for early administration of specialist medication.
- On the day of transfer, swift referral to the tertiary centre occurred as a result of good communication between medical, nursing and managerial staff.

A good knowledge of basic sciences including anatomy and physiology are vital in a member of a general medical team.

- The diagnosis was made not based on extensive knowledge of rare haematological conditions but on basic knowledge of the anatomy of the lumbar plexus.

clotting disorders were also considered, including acquired factor VIII deficiency.

TREATMENT

The patient was given FEIBA, an activated prothrombin complex concentrate, and tranexamic acid and immediate transfer to the local tertiary centre was arranged. At the tertiary centre, he was started on a course of corticosteroids.

OUTCOME AND FOLLOW-UP

The patient was transferred and treated. He has since been seen in follow-up clinic having entered remission and made a full recovery.

DISCUSSION

Acquired haemophilia A is a rare condition that is not widely reported in the literature. Until last year there was limited guidance available on diagnosis and management.[2 3] The United Kingdom Haemophilia Centre Doctors' Organisation (UKHCDO) guidelines[4] have attempted to fill this gap, but due to the low incidence and poor reporting of cases, sample sizes still remain small and the conclusions can not necessarily be relied on. With rare conditions such as acquired haemophilia A are difficult to diagnose and treat, especially in smaller hospitals.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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