

Acquired haemophilia presenting with tongue swelling and dysphagia

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DESCRIPTION

A 78-year-old woman presented to the emergency department with rapid swelling and bruising of the tongue and anterior neck over a 12 hours period. Other symptoms included dysphagia, dysphonia and tightness in her neck. There was no stridor or clinical evidence of airway compromise. She did not report any trauma or other injuries that could account for the swelling. She had recently suffered an upper respiratory tract infection. Her medical history included osteoarthritis, hypertension and gastro-oesophageal reflux disease. Her medication history consisted of candesartan, simvastatin, omeprazole and simple analgesia as required.

Examination of the patient revealed an obviously discoloured swollen tongue (figure 1), with oedema to the floor of the mouth displacing the tongue superiorly. There was also diffuse ecchymosis of the anterior neck. There were no signs of airway compromise and the patient had full pain-free range of neck movement.

Flexible nasendoscopy was performed to assess airway patency. This showed a patent airway with symmetrical cord movement, and there was some postcricoid oedema suggestive of venous congestion (figure 2) and no evidence of deep neck space collection or haematoma.

On initial laboratory investigations, haemoglobin and platelet counts were normal. Her initial activated partial thromboplastin time (APTT) was elevated at 64.6 s (reference range 25–35 s), as was her APTT ratio at 2.2. Her factor VIII (FVIII) assay was very low at 4.2 iu/dL (reference 50–150 iu/dL).

The low FVIII levels and raised APTT suggested a diagnosis of acquired haemophilia A.

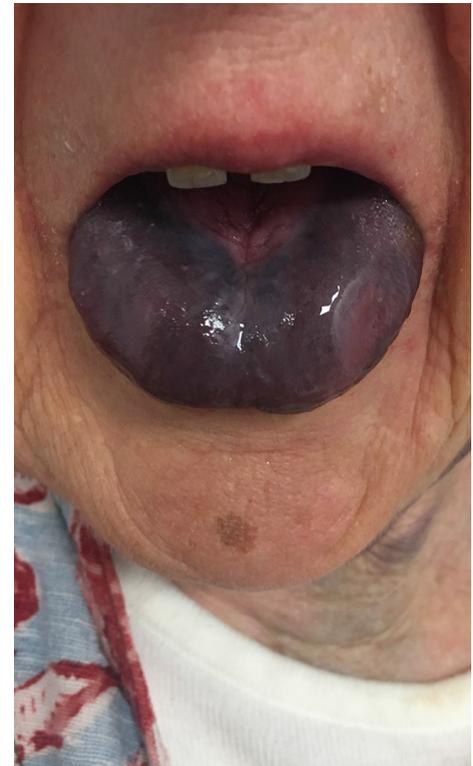


Figure 2 Appearance of larynx with postcricoid oedema.

Acquired haemophilia A is a medical emergency causing potentially life-threatening bleeding due to an autoimmune process.¹ The body produces auto-antibodies, known as FVIII inhibitor, which inhibit the procoagulant activity of FVIII. In approximately 50% of acquired haemophilia A cases, there is no cause identified, and the remaining 50% is made up of autoimmune conditions, malignancy, pregnancy and the postpartum period, dermatological conditions and rarely drug reactions.²

This patient was treated with factor eight inhibitor bypass activity (FEIBA) intravenously followed by prednisolone and cyclophosphamide. In 30% of cases, prednisolone alone can induce elimination of FVIII inhibitor and the addition of cyclophosphamide raises this up to 70%. Following administration of FEIBA, her FVIII inhibitor levels fell from 32.6 Bethesda units (BU) to 4 BU. After several days of prednisolone and cyclophosphamide, her FVIII inhibitor test was negative, her APTT returned to within normal limits and her APTT ratio returned to 0.9. Her FVIII levels rose from 4.2 iu/dL pretreatment to 162 iu/dL.



Figure 1 Tongue swelling.



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Learning points

- ▶ Acquired haemophilia A is a medical emergency with potentially fatal consequences secondary to uncontrollable bleeding. Rapid treatment of acquired haemophilia with bypassing agents prevents further deterioration with.
- ▶ Early immunosuppressive treatment with corticosteroids required to eradicate existing autoantibodies and return coagulation status to normal.
- ▶ Surgical specialties covering the affected areas should review patients to assess for local complications associated with bleeding.

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