

Acquired Haemophilia: A Case Report and Literature Review

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Case Report

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Abstract

Acquired haemophilia (AHA) is a rare coagulation disorder secondary to auto-antibodies against coagulation factor, most commonly factor VIII with potential for life threatening bleeding episodes. We report a case of an 88-year-old female presenting with frank haematuria three weeks after catheter insertion. Her background was of Alzheimer's Dementia, Asthma and Bullous Pemphigoid for which she was on low dose maintenance prednisolone (5mg). Laboratory tests showed haemoglobin 98g/dl and partial thromboplastin time (PTT) of 60s, with corrected prothrombin time 52s. Fibrinogen 5.39. As such coagulation factors were tested which revealed factor VIII of 0%. Her case was complicated by urinary tract sepsis, as such she was treated with oral prednisolone 60mg without immunosuppressive agent usage. A pan-CT scan revealed likely mesothelioma for which she declined further investigation. This case report will describe a rare presentation of AHA associated with bullous pemphigoid and mesothelioma, complicated by infection and frailty.

Background

AHA is a rare condition characterised by the generation of antibodies against a coagulation factor². This is contrasted to hereditary haemophilia in which patients are born without or with dysfunctional specific clotting factors and has a much higher incidence. The most common antibody acquired is against factor VIII, but it can be associated with factors V, IX, XI, XIII, II, VII, X, VWF¹

Breakdown of factor VIII results in failure to complete the intrinsic pathway of the clotting cascade, resulting in a high risk for catastrophic bleeding, without any other risk factors for bleeding. The severity of bleeding can vary significantly from patient to patient depending on antibody titre.

Aetiology^{1,3}

In approximately (46% - 51.9) of cases, acquired haemophilia presents idiosyncratically. There are, however, a wide variety of diseases that can lead to the disease, as follows;

Haematological/Oncological (6.4-18.4%)

As a paraneoplastic phenomenon, it is associated with leukaemia, multiple myeloma, lymphoproliferative disease, solid tumours, lymphomas and other oncological/haematological disorders

Obstetric

8.4% of cases can occur in the postpartum period up to 12 months of delivery

Autoimmune (9.4%-17.0%)

Rheumatoid arthritis, connective tissue disease, multiple sclerosis, giant cell arteritis, connective tissue disease, vasculitis, systemic lupus erythematosus, inflammatory bowel disease, Grave's disease, autoimmune hypothyroidism, myasthenia gravis

Dermatological

Pemphigus, pemphigoid, psoriasis

Medication

Antibiotics, interferon beta, amiodarone, rivastigmine, heparin, phenytoin, chloramphenicol, methyldopa, clopidogrel, Omalizumab, have been associated with cases

Other

Asthma, COPD, Hepatitis B, Hepatitis C, Diabetes, have also been associated with the disease

Epidemiology

The UK estimated incidence is 1.48 cases per million persons each year (for factor VIII).¹ The incidence is higher due to missed diagnoses and patients with only low levels of antibodies.

There is a biphasic age distribution for the condition. The majority occur in male 60-80 year olds, with the second smaller peak in women aged 20-30 years. Overall, there is equal prevalence in men and women, and race has not been identified as a risk factor. The increased incidence in young women is related to obstetric/post-natal acquired haemophilia.⁵

Pathophysiology

Factor VIII is converted to activated Factor VIII by thrombin as part of the intrinsic clotting cascade. This serves as a cofactor to factor IX, converting it to factor Xa. This leads to production of thrombin, activating further coagulation, releasing fibrin from fibrinogen and activating platelet surfaces. As such deficiency or absence of plasma FVIII prevents appropriate clot formation following vascular wall damage, with resulting bleeding.⁶

Autoantibodies produced belong to the IgG class. These bind to Factor VIII heavy and light chain domains, (commonly C2 and A2) preventing binding to factors IXa, X, phospholipids and von Willebrand factor.⁷

Unlike congenital haemophilia, there can be residual factor VIII activity. However, this lacks clinical benefit, as without sufficient levels of thrombin on activated platelet surfaces, significant bleeding cannot be coagulated.

Clinical Features

The majority of AHA cases present with sudden onset severe bleeding, with high mortality rate within weeks. Typical presentations are of subcutaneous bleeds (80% of patients)⁸ large haematomas, gastrointestinal or genitourinary bleeding, and bleeding following surgical/dental procedures. The bleeding following surgical procedures and wounds is often difficult to treat, therefore invasive procedures are best to be avoided. Large haematomas may result in irreversible nerve or blood vessel damage. Spontaneous intracranial bleeding and intra-articular bleeding are rare.

In pregnancy and the post-partum period, symptoms can occur immediately after delivery, presenting

with uterine or vaginal haemorrhages, or can present up to 12 months following delivery with haematomas, gastrointestinal or genitourinary bleeding. In 70% of cases, spontaneous remission is expected, with 30% requiring immunosuppression and haemovascular support. Complete remission approaches 100% at 30 months.^{8,9}

Diagnosis

A coagulation panel shows prolongation of activate prothrombin time (aPTT) and normal prothrombin time (PT), thrombin time (TT), closure time (CT), platelet count, plasma fibrinogen level. APTT mixing tests can confirm the factor inhibitor, and an assay can then be undertaken.^{1,2}

Investigations should be undertaken to look for the underlying cause including CT image, peripheral blood film, LDH, viral serology, TFTs, ANA, Anti-DNA, RF, pregnancy test.^{2,3}

Management

There are two arms of management of AHA, that of acute haemovascular stasis and that of inducing remission of autoantibodies.¹

To begin with haemostatic agents; the most effective management of severe bleeding is by anti-inhibitor coagulant complexes, commonly referred to as Bypassing Agents (BPA). These include recombinant activated factor VII or activated prothrombin complex concentrate, with aims to bypass the FVIII dependant stage of clotting.^{3,9} The downside of these drugs is a considerable increase in thromboembolic events which must be considered, and as such the drug should be discontinued promptly following remission of AHA. BPA can effectively stop bleeding in around 86% of cases.⁹

A new medication has been developed – recombinant porcine factor VIII, with a goal to mimic effects of factor VIII without being inhibited by autoantibodies.

Tranexamic acid can be used for mucosal bleeding, however, should be avoided in haematuria due to risk of clot retention. It is considered safe to use in conjunction with bypass agents.

Moving onto immunosuppression to cause complete inhibitor factor eradication. This should be initiated as soon as the diagnosis is confirmed, however consideration needs to be undertaken due to the significant risk of infection. This is especially of note in the elderly population, whose outcome will be significantly worse when concomitant with sepsis.

The usual first line treatment is corticosteroid monotherapy or combined with cyclophosphamide. Prednisolone dosing at 1mg/kg/day for 4-6 weeks, cyclophosphamide dosing at 1-2mg/kg/day for up to 6 weeks.⁹ One study suggests defining remission as increase in FVIII above 50iU/Dl and lowering the inhibitor titre to below threshold. This regimen is effective in 68% of patients.^{9,1}

If ongoing at 4-6 weeks, then second line immunosuppression should be instigated. Currently Rituximab is recommended at dose 375mg/m²/week for 4 weeks. Alternatively, ciclosporin/tacrolimus/azathioprine/vincristine/mycophenate mofetil have been trialled for use

It is suggested to follow up these patients for 2 years after remission with monthly FVIII levels for first 6 months, then 6 monthly afterwards. Relapse occurs in 20% of patients.¹¹

It should be of note that despite treatment, mortality for AHA ranges from 6.7% to 38%.¹⁰

The Case

An 88-year-old female presented to the Emergency Department with frank haematuria and dysuria, three weeks after catheter placement. She had a background of advanced Alzheimer's dementia, asthma and bullous pemphigoid for which she was on long term prednisolone 5mg daily. Importantly she

was not anticoagulated

Laboratory tests showed haemoglobin 98g/dl, partial thromboplastin time (PTT) of 60s, with corrected prothrombin time 52. Fibrinogen 5.39.

Clotting factors subsequently showed:

Factor VIII: 0%, Factor IX 83%, Factor XI 66%, Factor XII 61%. Inhibitor screen positive with time dependant inhibitor present. Consistent with acquired haemophilia A.

This is consistent with a diagnosis of acquired haemophilia. Given her significant frailty status and possible infection she was deemed not suitable for aggressive immunosuppression (FEIBA or immunosuppression), instead started initially on 30mg prednisolone. The lose dose was given initially to reduce delirium side effects.

Mid-stream urine subsequently grew *E. coli* and she was started on nitrofurantoin 100mg BD and a one-off dose of gentamicin.

USS renal tract showed global cortical thinning and no evidence of hydronephrosis.

Bloods two days later: Hb 92g/dl, PTT 55.6 – Increased steroids to 60mg as tolerating well. Significant clinical improvement with reduction in haematuria, stable blood sugars and no delirium.

Bloods on Day 9: PTT 51, Hb 87

A Pan-CT revealed Pleural plaques and suspicious left sided pleural thickening, with possibility of mesothelioma. Given her functional status and co-morbidities she was deemed for best supportive care, without further investigations. For similar reasons she did not undergo cystoscopy to investigate the haematuria.

Treatment duration was for prednisolone 60mg 14 days total, then to reduce by 5mg every two weeks until at 10mg OD.

She remained clinically well and was discharged home. A decision was made that if she were to relapse her prednisolone could be increased back to 60mg, but palliative care should be considered.

As such this case demonstrates a patient with severe frailty and multiple possible triggers for her AHA but showed a good response to steroid monotherapy.

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