Post partum flare in a patient with hereditary angioedema

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Abstract

Hereditary angioedema (HAE) is a rare autosomal dominant genetic disorder due to dysfunctional or deficiency of C1 esterase inhibitor (C1INH). This leads to episodic sub-cutaneous and submucosal oedema involving the upper respiratory and gastrointestinal tracts which can be life threatening. Stress and trauma are known to trigger angioedema attack[1]. As a result of hormonal changes in pregnancy some women may experience increase angioedema attacks. This is a case report of a 34 year old woman who developed life threatening post partum complications with a background history of HAE.

Key words: danazol, hereditary angioedema, C1 esterase inhibitor deficiency, pregnancy, C1INH concentrate.

Introduction

Hereditary angioedema (HAE) is a rare life-threatening disease that can occur in pregnancy. With respect to this disease and pregnancy, there is very few information available in the literature.

Case Report

Patient was a 34 year old gravida 2, Para 1 conceived by in vitro fertilization. She was diagnosed with HAE at the age of sixteen, with predominant gastrointestinal attacks. She has a strong maternal family history of HAE and was advised for Danazol prophylactic therapy but she was not compliant due to androgen related side effects[5]. Four years ago, she had an uncomplicated emergency cesarean section (CS) for failure to progress. In her current pregnancy, she had an uneventful antenatal period except occasional mild abdominal cramps and self limiting cutaneous angioedema. Due to this reason the C1INH concentrate was not administrated as prophylaxis. She delivered a female infant at term by an elective CS under spinal anaesthesia. She complained of abdominal pain on the first postoperative day, which rapidly developed into severe ileus with large and small bowel dilatation. (Fig. 1A) Day five post partum, she developed acute non oliguric renal failure (serum creatinine 311 μmol/L) and proteinuria (2.4 gm/24 hours) with difficulty in controlling blood pressure. A dose of C1INH concentrate was delivered in the intensive care unit with little improvement, following which she was transferred to a tertiary hospital for further management. A subsequent CT scan revealed small bowel wall oedema consistent with HAE (Fig. 1B). She improved with supportive therapy and was discharged 2 weeks later but represented to the emergency department with severe abdominal pain. A CT scan confirmed thrombosis of the left ovarian vein extending up to the inferior vena cava.

Patient is currently doing well. Her blood pressure and renal function has normalised two months post partum. She received three months of warfarin therapy and has recommenced on Danazol prophylaxis. She is closely followed up by an immunologist, has a Medic Alert bracelet, and was advised to present to the emergency department for C1INH concentrate infusion for future angioedema attack.

Discussion

Although the prevalence of HAE in the general population is 1:10,000 to 1:50,000, the condition can result in considerable morbidity and sometime even death[2,3]. HAE attack can occur anytime throughout the pregnancy but is more commonly documented during post partum period[1]. Therapy available during pregnancy is often limited[6]. Androgens prophylaxis is contraindicated and antifibrinolitic agents should be used with caution[3]. Even though C1INH concentrate is generally accepted as an effective therapeutic option, the best outcome often requires prophylactic administration[4]. Therefore pregnancy is often challenging and requires multidisciplinary approach during peripartum and postpartum period to ensure a good maternal and fetal outcome.

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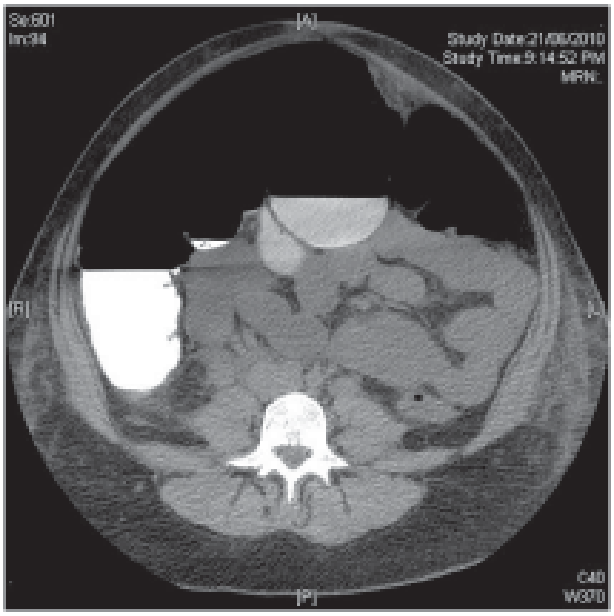

Figure 1A

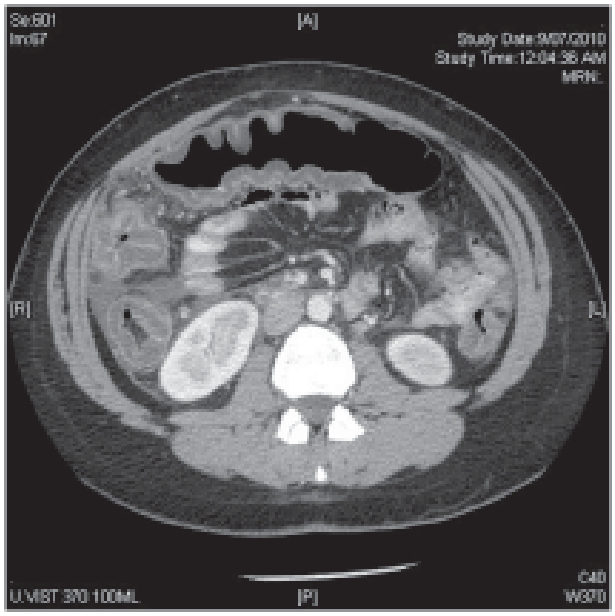

Figure 1B

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