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Case Report Open Access

Management of Pregnancy in Woman with Heriditary Angioedema

Sreejith Kodakkattil*

Addenbrookes University Hospital, Rosie Maternity Unit, Cambridge, USA

Abstract

Hereditary angiooedema (HAE) is characterised by recurrence of cutaneous and mucous membrane swellings in any part of the body. Symptoms usually appear early in life and are normally accompanied by a family history because the disease is inherited in an autosomal dominant manner. The spontaneous mutation rate is about 25% and more than 1001 inhibitor gene mutations have been described′different C1 inhibitor gene mutations have been described.

Keywords: Hereditary angioedema; Pregnancy; Management

Introduction

Hereditary Angioedema (HAE) is characterised by recurrence of cutaneous and mucous membrane swellings in any part of the body. Symptoms usually appear early in life and are normally accompanied by a family history because the disease is inherited in an autosomal dominant manner. Abdominal attacks of HAE are mostly characterized by pain, vomiting, and diarrhoea. They are caused by transient edema of the bowel wall, leading to partial or complete intestinal obstruction, ascites, and hemoconcentration. Symptomatic involvement of the GI tract is an important clinical feature of HAE, since it is more distressing than the edema of the skin, and it occurs far more frequently than the life-threatening laryngeal edema [1,2].

Case Description

27 year old multiparous woman presented in her third pregnancy for booking in Addenbrookes University hospital. She has had two previous normal vaginal deliveries, one in 2004 and other 2008. She was diagnosed with HAE when she was sixteen years of age. She was on danozol 200 mg three times a day for prophylaxis of episodes of angioedema before her pregnancy. During careful history taking we understood that she may get episodes of abdominal pain every 3-4 months which may be mild or severe. If severe symptoms she normally attends emergency department and receive C1 esterase infusion with prompt relief of her symptoms. At booking there was past history of two episodes of laryngeal oedema which was successfully treated with intravenous C1 esterase infusion. There was history of allergy to fresh frozen plasma following treatment for an episode. There was no previous history of general anaesthetic. Patient had two successful epidurals for her previous vaginal births. The usual symptoms were episodes of abdominal pain. During this pregnancy, episodes occurred every 7-8 days during her first and second trimester. During third trimester these episodes occurred every 9-10 days. Overall patient required thirty administrations of C1 esterase till her delivery. These episodes were due to severe abdominal pain for which she received C1 esterase. This was performed in the emergency department and she didn't require any inpatient stay for these episodes.

During her antenatal period her care was shared by multidisciplinary team including obstetricians, anaesthetist, emergency medicine, allergy/vasculitis team, pharmacology and haematology team.

Anaesthetic plan was to give prophylactic C1 esterase before any regional or general anaesthetic if required in labour. The plan was to give an early regional and avoid a general anaesthetic if possible due to risk of laryngeal oedema during intubation. 1000 units (two vials of 500 units) of C1 esterase were stored in the theatres in delivery unit. A further supply was available from the central pharmacy if additional demands occurred. A regular automated measurement of blood pressure was discouraged as this may act as a trigger to complement activation. In the unlikely event of an episode of laryngeal oedema the plan was to administer 100 mg hydrocortisone (or 30 mg predinsolone), intravenous C1 esterase 500 units and adrenaline if necessary.

Our patient was induced at 40 weeks+10 days period of gestation due to failure of spontaneous labour. Our patient established spontaneous labour following rupture of membranes at 3 cm. A healthy boy baby weighing 4360 grams was delivered vaginally. Patient was comfortable with gas and air for pain relief. Patient was administered 500 units of C1 esterase intravenously immediately after artificial rupture of membranes due to abdominal pain and nonspecific symptoms. Post natal period during hospital stay way uneventful and patient was discharged on the 2nd postpartum day. Patient started danozol (200 mg) three times daily, following discharge. Our patient attended twice to emergency department with acute episodes requiring intravenous C1 esterase infusion during the first 6 weeks post partum.

Discussion

In the presence of C1 inhibitor deficiency the classical complement pathway can be inappropriately or prematurely activated. This leads to the activation of anaphylactoid-like substances and vasoactive peptides which leads to inflammatory and oedematous response [3]. The spontaneous mutation rate is about 25% and more than 100 different C1 inhibitor gene mutations have been described [1].

Airway obstruction is the most common cause of mortality. Airway trauma during intubation may worsen laryngeal edema and necessitate emergency tracheostomy [4]. Thus antenatal review and planning by anaesthetic team is an important part of multidisciplinary care during pregnancy. Less frequent or rare clinical symptoms include circulatory collapse and shock, non crampy permanent abdominal pain, dysuria, hemorrhagic diarrhea , tetany and intususception [2].

In a study of seven women with HAE conducted in Australia, Chinniah and Katelaris found reduced episodes of angioedema during the last two trimesters [5]. There was increased severity and frequency of episodes during the postnatal period [5]. Natani et al. reported a case where the antenatal episodes were minimal compared to our case [6]. Their case had only three self limiting episodes which didn't require

***Corresponding author:** Sreejith Kodakkattil, Addenbrookes University Hospital, Rosie Maternity Unit, Cambridge, Box 224, CB2 0QQ, USA, Tel: 44 1223 217617; E-mail: skodakkattil@gmail.com

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any C1 esterase during antenatal period. Our patient required thirty injections of C1 esterase during the antenatal period. During her first two pregnancies the episodes were frequent during first two trimesters, however there was significant reduction in episodes during her third trimester. In contrary, her current pregnancy (third) episodes were similar in all trimesters. Our patient had a relatively good delivery and immediate postpartum period even though the antenatal period was eventful with multiple episodes of symptoms. Gorman whilst managing a pregnant woman with angioedema, commenced her on 500 to 1000 units of C1 esterase every 2 weekly during her antenatal period [7]. Their patient didn't have any major episodes of angioedema during this regimen. Postnikiff and Prizker reported a maternal mortality in a woman with hereditary angioedema from irreversible shock, induced by perineal swelling after vaginal delivery [8].

Learning Points

1. It is important to manage woman with hereditary angioedema with a multidisciplinary approach.

2. It is important to have pre anaesthetic review of these patients because of the chances of acute laryngeal oedema.

3. It is important to ensure provision of C1 esterase by liaison with pharmacy department for emergency as well as for labour.

4. A/E department team should be informed about these patients due to increased frequency of episodes during pregnancy as prophylaxis with danazol is usually stopped. Majority of these patients presents to A/E for initial management.

5. It is important to recognise that even routinely performed automated blood pressure readings can act as a trigger to precipitate an episode.

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