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Research Article

HEREDITARY ANGIOEDEMA AND PREGNANCY: REPORT CASE

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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 edema 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 32 year old woman who developed several crisis during pregnancy with a background history of HAE.

Keywords: Hereditary angioedema, C1 esterase deficit, Pregnancy.

INTRODUCTION

Hereditary angioedema (HAE) is a rare disease (1/10,000–1/50,000) [2] in which episodes of edema of the skin and mucous membranes develop. Episodes may last 2 to 5 days and resolve spontaneously, but if they involve the larynx or oral cavity, they may lead to airway obstruction and death. However, the most common site of involvement is the abdo-men [3]. Close follow-up during pregnancy, labor, delivery, and breast-feeding is recommended since these events can affect the course of the disease in different ways.

CASE REPORT

A.N 32 year old woman, first gestation, followed for hereditary angioedema since pre-pubertal period following the onset of segmentary edemas self-limited. Recurrent characteristics of edemas and family context (father and sister having the same illness) encouraged us to conduct a study of complement system. C1 INH dosage was very low and it confirmed the diagnosis of hereditary angioedema-1 due to quantitative C1 INH deficit. The patient received corticoids during crisis with good progress. She had been followed up by a multidisciplinary team (obstetricians, anesthesiologists) since the beginning of her pregnancy. In the beginnig of pregnancy, the patient had one episod of threat early abortion treated by progestins at minimum dose per os. The first edema crisis followed this episod (Figure 1) and was resolved by oral corticoids (methylprednisolone 60mg/day per os during 3 days following the crisis). During the follow up, prenatal check-up was without abnormalities and the patient had mild crisis every month on the extremities. At 28 GW the patient was admited for threat of premature birth associated to moderate abdominal crisis confirmed on abdominal US by low abundance ascites. Obstetrical ultra-sound was normal. The patient received IV corticoides with good progress (Methyprednisolone 80 mg IV during 3 days). The threat of premature birth was treated with calcium channel blockers (IV Nicardipine followed by oral tabs). At 32 GW, an obstetrical ultra-sound was done showing fetal biometry between 10th and 25th percentiles without Doppler abnormalities. At 34 GW, control ultrasound showed intrauterine growth retardation with biometry under 3rd percentile with estimated weight at 1400g, without oligoamniosis, zero umbilical diastole, decrease in resistance index at cerebral Doppler with negative wave « a » in ductus venosus (Figure 2), which led us do emergency C-S. A healthy boy baby weighting 1400 grams was delivered and was transferred to neonatal intensive care unit. No particular event during perinatal period was noted and the patient went back home after 5 days and the new-born left neonatal ICU after 3 weeks. 3 months after delivery the mother and her new-born were well.

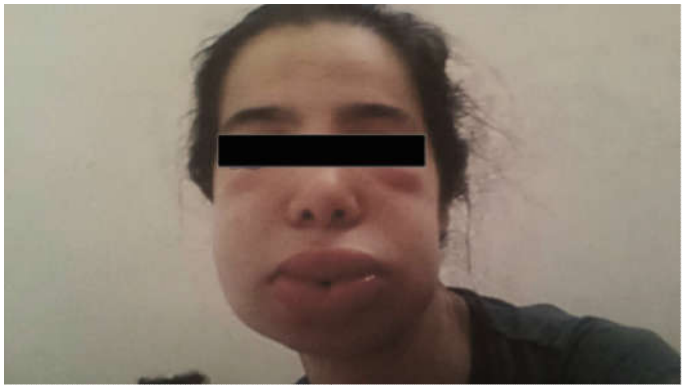

Figure 1. Hereditary angioedema crisis showing signs of facial edema

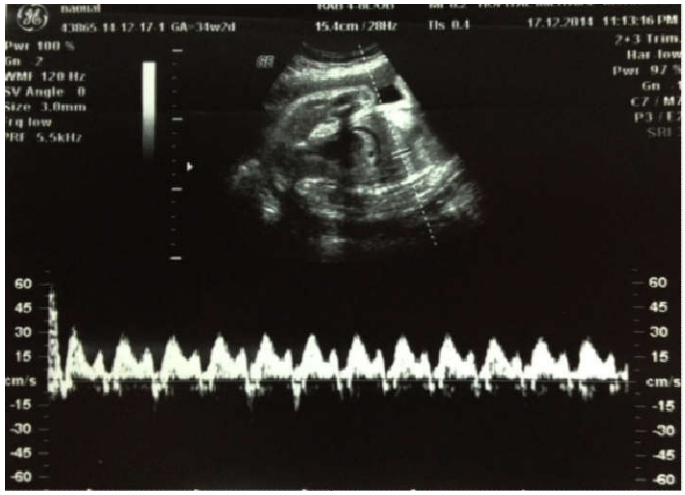

Figure 2. negative wave « a » on Ductus Venosus

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DISCUSSION

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 gastrointestinal 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 [4,5]. 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 [6,7]. 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 [8]. Androgens prophylaxis is contraindicated and antifibrinolitic agents should be used with caution[7]. Even though C1INH concentrate is generally accepted as an effective therapeutic option, the best outcome often requires prophylactic administration [9]. The anatomical and physiological primarily hormonal changes of pregnancy may influence the manifestations of HAE and interfere with the diagnosis and the treatment of HAE. Pregnancy can either mitigate or aggravate HAE; alternatively, it may have no impact whatsoever. Infrequently, the manifestations of HAE first occur during pregnancy. Attack frequency observed during previous pregnancies is not predictive of HAE events during any subsequent pregnancy. Pregnant HAE-1/2 patients require vigilant care and meticulous monitoring by an HAE expert. Patients should be managed in close cooperation by professionals from relevant medical specialties. Labor and delivery themselves only rarely induce an attack, which may occur either during labor or within 48 hours of delivery.

Close follow-up is recommended for at least 72 hours postpartum after uncomplicated vaginal delivery. Breastfeeding may be associated with an increased number of maternal edematous attacks, with abdominal symptoms and facial edema, but is still recommended [9].

REFERENCES

1. Chinniah N, Katelaris CH. Hereditary angioedema and pregnancy. Aust N Z J Obstet Gynaecol 2009; 49: 2-5.
2. Cicardi M, Agostoni A. Hereditary angioedema. N Engl J Med. 1996;334:1666.
3. Agostoni A, Cicardi M. Hereditary and acquired C1-inhibitor defi- ciency: biological and clinical characteristics in 235 patients. Medi- cine (Baltimore). 1992;71:206–15.
4. Tosi M (1998) Molecular genetics of C1 inhibitor. Immunobiology 199: 358-365.
5. Bork K, Staubach P, Eckardt AJ, Hardt J (2006) Symptoms, course, and complications of abdominal attacks in hereditary angioedema due to C1 inhibitor deficiency. Am J Gastroenterol 101: 619-627.
6. Hermans and Cedric. Successful management with C1- inhibitor concentrate of hereditary angioedema attacks during two successive pregnancies. Archives of Gynaecology and Obstetrics 2007; 276: 271-6.
7. Davis AE. New treatments addressing the pathophysiology of hereditary angioedema. Clinical and Molecular Allergy 2008; 6: 2.
8. Bingham CO. UpToDate [database on the Internet] Waltham, MA: UpToDate; [Accessed 2006 January 13]. Overviewofangioedema.
9. Nathani F, Sullivan H, Churchill D. Pregnancy and C1 esterase inhibitor deficiency: a successful outcome. Arch Gynecol Obstet 2006; 274(6): 381-4.

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