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One Case of Hereditary Angioedema Characterized by Pleural Effusion

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Abstract

Hereditary angioedema is a rare autosomal dominant disorder with prevalence estimated at 1/50,000, C1-INH levels may be decreased or normal, with an accompanied decrease in functionality, characterized by non-itchy and non-pitting swelling of subcutaneous and sub mucosal tissues of the face, extremities, genitalia, gastrointestinal tract, and upper airways, which can be life-threatening when located to the airways. It has been reported hereditary angioedema with ascites, but characterized with pleural effusion is rare. We describe a case of 30-year-old woman with pleural effusion for attracting attention of medical staff.

Keywords: Hereditary angioedema • Pleural effusion • Allergy

Introduction

Hereditary angioedema is a rare autosomal dominant disorder with prevalence estimated at 1/50,000 [1], C1-INH levels may be decreased or normal, with an accompanied decrease in functionality, characterized by non-itchy and non-pitting swelling of subcutaneous and sub mucosal tissues of the face, extremities, genitalia, gastrointestinal tract, and upper airways, which can be life-threatening when located to the airways. It has been reported hereditary angioedema with ascites [2,3], but characterized with pleural effusion is rare. We describe a case of 30-year-old woman with pleural effusion for attracting attention of medical staff.

Case Report

A 30-year-old female was admitted to the hospital because of abdominal distending pain, nausea and vomiting for 28 hours, without diarrhoea, swelling, rash or dyspnea. The patient complaint of abdominal pain on the left upper quadrant abdomen in the beginning, 4 hours later spread to the whole abdomen, the companied symptoms were abdominal distention, vomiting a lot of black digestive food and low fever, the temperature was 37.5°C.

Family study confirmed her father, grandma, two uncles, aunt, cousin and her son have herediaty angioedema (Figure 1).

The physical examination revealed stable vital signs, soft abdomen, tenderness on the left upper quadrant abdomen and shifting dullness positive. Laboratory investigations included leukocyte, hemoglobin, platelet counts of 4.79 × 10^9/L, 146 g/L, 93 × 10^9/L, respectively. Her C4 (0.07 g/L, normal: 0.1-0.4) and C1-INH (0.05 g/L, normal: 0.21-0.39) levels were low, C3 (1.05 g/L, normal: 0.9-1.8) and CH50 (25.0 U/mL, normal: 23-46) were within normal ranges. Meanwhile, hepatonephric function and coagulation function were detected. ALT (123 U/L, normal: 7-40), AST (99 U/L, normal: 13-35), ALP (227 U/L, normal: 35-100), GGT (121 U/L, normal: 7-45), LDH (387 U/L, normal: 0-250), HBDH (288 U/L, normal: 80-220), CRP(29.48 mg/L, normal: 0-5), PT (13.8 s, normal: 8.8-11.8), INR(1.25, normal: 0.86-1.14), D-Dimer (1713 ng/mL, normal: 0-550) levels were high, while PT% (63%, normal: 80-130) was low. The hepatonephric function and coagulation function returned to normal after the symptoms relieved.

Laboratory findings also included as follows. Abdominal enhancement CT scan revealed the second and third group of small intestinal edema, possibility of the second group of small intestinal volvulus, ascites, peritonitis, cystic low density focus on the fundus uteri, ovarian cyst, and accessory spleen, no pleural effusion initially (Figure 2). Ultrasound revealed ascites 3.5 cm in pelvic floor, 1.5 cm in spleen fossa, 1.0 cm in Morrison's crypt, 1.2 cm in the left lower quadrant abdomen, 0.8 cm around the left lobe of liver. Standing abdominal plain film showed incomplete intestinal obstruction. The color Doppler ultrasound revealed no abnormal sign of superior and inferior mesenteric artery. Light yellow liquid was drained by abdominal paracentesis. Chest X-Ray showed cardiac enlargement after 2 days.

The patient complaint of relief of abdominal pain and vomiting, but chest tightness, short of breath and dyspnea appeared 1 week later. Subsequently, abdominal CT scan was rechecked after 8 days' acute attack, which revealed mild swelling of the second and third group of intestine on the left upper

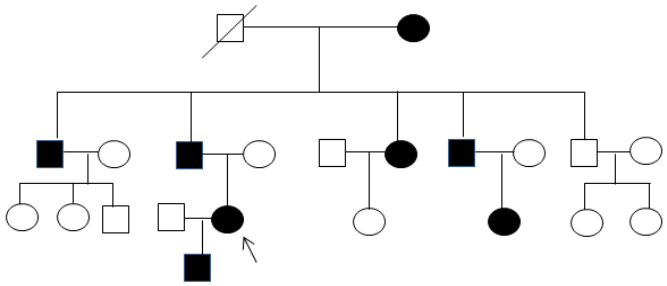

Figure 1. Family tree revealed her father, grandma, two uncles, aunt, cousin and her son were the patients with hereditary angioedema.

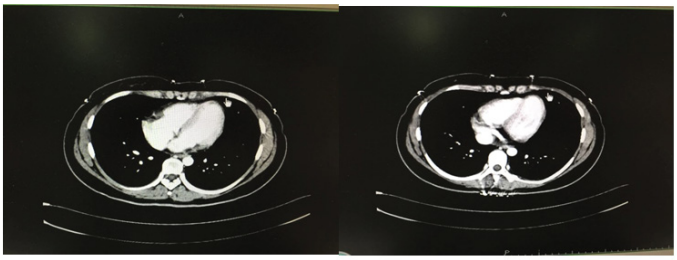

Figure 2. No pleural effusion in the abdominal enhancement CT scan on the first day acute attack.

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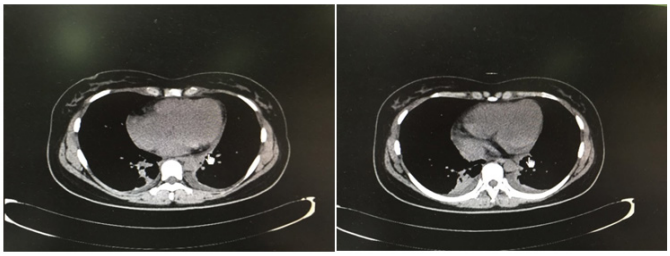

Figure 3. Abdominal CT scan revealed pleural effusion and atelectasis 8 days later (Abdominal pain lightened, but chest tightness, short of breath and dyspnoea appeared).

quadrant abdomen, and the second group of small intestinal tortuosity, Swelling and tortuosity improved obviously. Abdominal and pelvic effusion decreased remarkably. Retroperitoneal enlarged lymph nodes, accessory spleen and splenomegaly were same as before. However pleural effusion and atelectasis were progressive (Figure 3).

The symptoms were failure to respond to antibiotics and glucocorticoids, then relieved gradually after taking danazol.

Discussion

Hereditary angioedema is a rare congenital disease caused by SERPING1 gene mutation, which results in deficient C1 inhibitor (type 1-HAE) and dysfunctional C1 inhibitor (type 2-HAE). Other different forms of HAE are genetically unidentifiable: HAE with mutation in the F12 gene (HAE-FXII), angiopoietin-1 gene (HAE-ANGPTI), plasminogen (HAE-PLG) and unknown mutation (HAE-UNK) [4]. The patient presents a history of painful abdominal symptoms, a positive family history, failure to respond to antihistamines and glucocorticoids, and the laboratory investigation revealed C1-INH function, C1-INH protein and C4 levels are abnormally low, so type 1-HAE is definitely diagnosed.

Conclusion

The clinical manifestation of HAE is recurrent angioedema attacks, painful abdominal symptoms, probably emergency surgery for misdiagnosed by acute abdomen or life-threatening for laryngeal edema. Intestinal obstruction and ascites maybe appeared when acute attack, but pleural effusion is rare. The patient complaint of chest tightness, short of breath, and dyspnea, CT scan revealed pleural effusion, atelectasis, ascites and swollen intestinal mucosa. The common cause for pleural effusion is infection, tumor, autoimmune disease and so on. But for the HAE patient characteristic by pleural effusion, constant vigilance is essential in order to decrease the mortality, morbidity and increase the quality of life.

References

1. Zanichelli, Andrea, Francesco Arcoleo, Maria Pina Barca and Paolo Borrelli, et al. A nationwide survey of hereditary angioedema due to C1 inhibitor deficiency in Italy. *Orphanet J Rare Dis* 10(2015).
2. Huali, Ren and Zhang Hongyu. One case of hereditary angioedema with abdominal pain and temporary ascites. *Chinese Med J*. 85(2005): 272.
3. Xuemin, Yan and Zhang Hongyu. One case of intermittent vomiting and ascites- C1 inhibitor deficiency. *Chinese J Clin Immunol and Allergy*. 3(2009): 63-65.
4. Maurer, Marcus, Markus Magerl, Ignacio J Ansotegui and Emel Aygören-Pürsün, et al. The international WAO/EAACI guideline for the management of hereditary angioedema-The 2017 revision and update. *Allergy*. 73(2018): 1575-1596.

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