SPLENIC INJURY IN HEMOPHILIC PATIENT WITHOUT ABDOMINAL TRAUMA

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BAKGROUND

Splenic rupture is well described as a complication of abdominal trauma and spleen is easily injured in children. Spontaneous splenic rupture is an unusual event, described in several conditions that cause splenomegaly. These conditions are generally grouped in the categories of infectious diseases like infectious mononucleosis, malaria and dengue, and hematological malignancies.

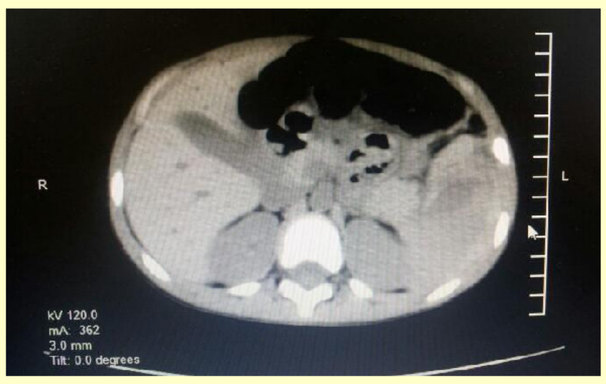
The consequences of spontaneous rupture of the spleen in this condition can be catastrophic.

Hemophilic children bleed frequently but splenic bleeding is not so common. Up to now, only few cases have been described.

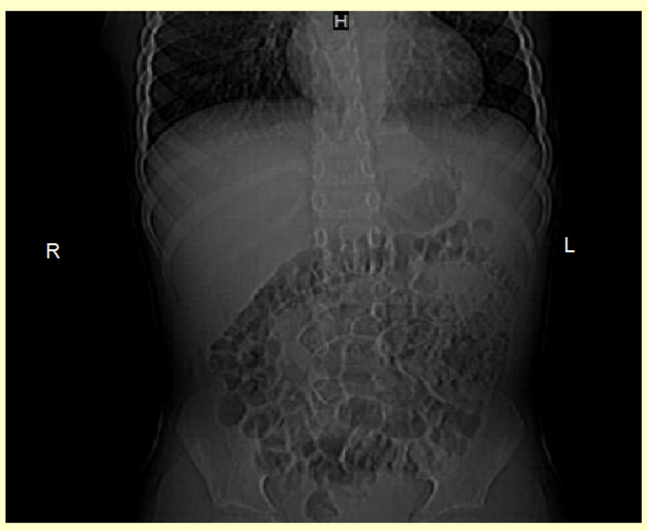
CASE DESCRIPTION

KGL is a four old year hemophilic patient, who has inhibitor since he was 18 months, and started immunotolerance treatment (ITI) six months before the bleeding episode that is being related. It was observed an Inhibitor ***title*** pick of 5,2 BU in March 2011, after ITI had started, it decreased to 2,6 BU.

In April 2013 after a cold and a severe coughing due to an asthmatic crisis, the patient presented an abdominal pain without any visible or described trauma. The parents brought him to the hemophilia emergency unit, after a CT scan that lead to a final diagnosis of peri splenic hematoma. The Hb level was 9,7 mg/dl fall to 7,4 mg/dl and maintained. Started receiving bypass agent therapy with NovoSeven in habitual doses, and pediatric surgeon surveillance, the patient remained at hospital under intensive pediatrics care. During this period, the hemoglobin level remained stable, and after four days a new CT scan indicated reduction in the hematoma area, as well as the absence of intra abdominal free liquid. At the next day, the patient had a compelling crying episode, followed by vomit and had a hemodynamic instability indicating that it should be necessary to undergo emergency splenectomy. The dose of NovoSeven was increased during the surgery and returned to pediatric doses every two hours at the first day and every three hours in the next day and was progressively reduced up to the eighth day after the surgery. The ITI process wasn´t canceled during the period of the surgery treatment. After that , the patient recovered his health, stayed at hospital for one more week and it was possible to continue the ITI at home. The inhibitor *title* had not increased anymore. Currently, the patient received prophylactic immunization and is continuing to receive oral penicillin. The inhibitor *title* was absent after six months and had total recovery of factor VIII after eight months and the ITI was achieved. Up to now prophylactic treatment at home is being performed.



CT April, 23



CT after 5 days

DISCUSSION

The mechanism of spontaneous splenic rupture is not clear. Three mechanisms have been proposed. The first one is that cellular hyperplasia and engorgement causes increased intrasplenic tension, and eventually rupture. The second one suggests that rupture is in fact not really spontaneous, but associated with episodes of increasing intra-abdominal pressure caused by coughing, vomiting, defecation, etc. Finally the third putative mechanism is that microvascular thrombosis and infarct weakens the capsule,resulting in rupture.

Only few cases of splenic rupture in patients with bleeding disorders have been described, the majority of them associated with a preceding traumatic insult. Spontaneous rupture of the spleen in hemophilia is very rare and too dangerous if not recognized in time to be treated aggressively. Diagnosis of spontaneous splenic rupture requires a high index of suspicion whenever a patient presents with any of the conditions known to be associated with splenic rupture. The presence of left upper quadrant pain, referred to the left shoulder (Kehr's sign), caused by blood irritating the diaphragm, might alert the examining doctor. Once suspected, further investigations can prove the diagnosis. The quickly ultrasound scan is increasingly playing a role in the emergency department, especially in the identification of potentially life-threatening conditions. Once a diagnosis has been confirmed and a decision has been made to manage the patient conservatively, ultrasound scan can be used to frequently re-examine the patient. As soon as the patient have condition, CT can be contemplated to specifically identify the pathology. Splenic rupture, unidentified, carries a significant morbidity and mortality, this can be significantly harmful to hemophilia.

CONCLUSIONS

The spontaneous splenic rupture is not a common hematoma in hemophilic patient, when it occurs, it is generally caused by abdominal trauma. Inhibitor is definitely a challenger to control bleeding episodes in hemophilia. The early treatment depends on high levels of clinical suspicion, early use of imaging, great involvement of hematology colleagues, expeditious surgery, and family cooperation are the most important points to hemophilics evolution to be succeeded.

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