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Spontaneous Pelvic Hematoma- A Rare Manifestation of Sickle Cell Disease- A Case Report and Review of Literature

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Abstract
Hemorrhage in Sickle Cell Disease is not a common clinical entity and is most often limited to central nervous system bleeds, presenting with hemorrhagic stroke or extradural hematomas. We present an adolescent girl with Sickle Cell Disease who presented with acute abdominal pain and was diagnosed to have a spontaneous pelvic hematoma on non contrast CT. The girl had a normal coagulation profile with no other bleeding manifestations except for two episodes of epistaxis. She improved clinically after transfusion, and the pelvic hematoma resolved spontaneously with conservative management. To the best of our knowledge, this is the first reported manifestation of a spontaneous pelvic hematoma in a person with Sickle Cell Disease, presenting as acute abdomen, thus necessitating a high index of clinical suspicion of such an entity and appropriate management of the same.

Introduction

Sickle Cell Disease (SCD) is a form of haemolytic anemia resulting from autosomal recessive inheritance where glutamic acid is substituted by valine at position 6 of the beta-globin chain. This leads to the formation of sickle hemoglobin (HbS) which can occur either in the homozygous (HbSS) or heterozygous (HbAS) form[1,2]. Individuals with homozygous form of sickle cell haemoglobin manifest the disease, which is characterised by 'painful crisis'. This is caused due to vaso-occlusion by the abnormal sickle shaped red blood cells in the microvasculature, leading to tissue infarction[3].

Hemorrhagic manifestations are not very common in SCD and are mostly confined to the intracranial vessels resulting in cerebrovascular accidents or extradural hematomas. Although only 25 percent

of strokes in SCD are hemorrhagic, the mortality is as high as 25 to 50 percent within two weeks of the hemorrhagic event[3]. Gross hematuria is a well known entity occurring in persons with Sickle Cell trait, more often than Sickle Cell Disease[4]. The risk factors for hemorrhages in SCD are acute chest syndrome, acute hypertension, previous infarcts, low level steady state of haemoglobin concentration and high steady-state leucocyte count[5-9]. Haemorrhages occurring elsewhere in the body are seldom reported.

In the present case report, we describe a case of spontaneous pelvic hematoma in an adolescent with SCD. The clinical and radiological features of this condition along with the relevant review of literature are discussed. Assent from the patient and written informed consent from the parents were obtained for her findings to be reported.

Case Report

A 13 year old girl with Sickle Cell Disease presented with history of pain abdomen which was acute in onset for one day and gradually progressive. The pain was confined to the right lower quadrant and was dull aching in nature. It was associated with vomiting and loss of appetite. There were no fever, bowel or bladder disturbances, or gum bleeds. She had not attained menarche.

In the past, the girl had recurrent musculoskeletal aches and pains many of which required hospital admissions. She had not received any blood transfusions and had no history of tuberculosis, epilepsy, or surgeries.

She had been diagnosed with SCD through new born screening of hemoglobin and the diagnosis was confirmed by Hemoglobin electrophoresis. Both her parents were carriers of Sickle Cell trait and two of her siblings had Sickle Cell Disease.

On examination, the girl was afebrile, severely pale and icteric with typical hemolytic facies.

Her cardiovascular and respiratory systems were normal. The abdomen was soft with massive splenomegaly measuring 10 cm below the left subcostal margin. The right iliac fossa was tender on deep palpation. There were no musculoskeletal abnormalities and the central nervous system examination was normal.

Investigations

The blood counts revealed hemoglobin of 5.3 g/dl with a white blood cell count of 7900cells and platelet count of 2.15 lakhs. Her total bilirubin was 5.9mg/dl with an indirect bilirubin value of 5.5mg/dl. Her liver enzymes, renal function tests, lipid profile, bleeding time, clotting time and prothrombin time were within normal limits.

An ultrasound of the abdomen was performed and showed massive splenomegaly and a 8 X 7 cm mass with ill defined margins with mixed hyper and hypo echoic areas in the pelvis, that was separate from the uterus with minimal probe tenderness (fig1).

Non contrast CT of the pelvis showed a heterogeneous mass with high attenuation suggestive of acute pelvic bleed/ hematoma separate from the uterus with no intra-abdominal free fluid. (fig 2)

Course and treatment

Whole blood was transfused in view of the crisis and a significant drop in patient's haemoglobin concentration (5.3g/dl to 4g/dl). Abdominal pain relieved with opioid analgesics and she improved symptomatically over the next few days. The girl had two episodes of epistaxis on day 6 of admission which was controlled on local measures like nasal compression and head tilt.

The pelvic hematoma was serially monitored by ultra sonogram and showed resolution without surgical intervention. Follow up ultrasound after 2 weeks showed complete resolution and she was discharged on Hydroxyurea and vitamin supplements.

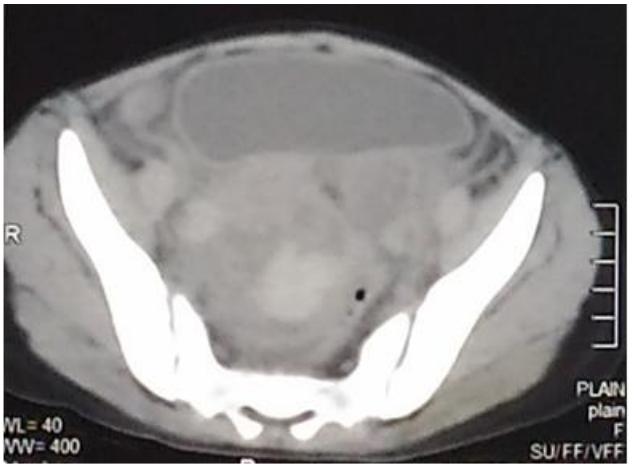

Fig 1. Ultrasound pelvis showing hyper echoic lesion with ill defined margins

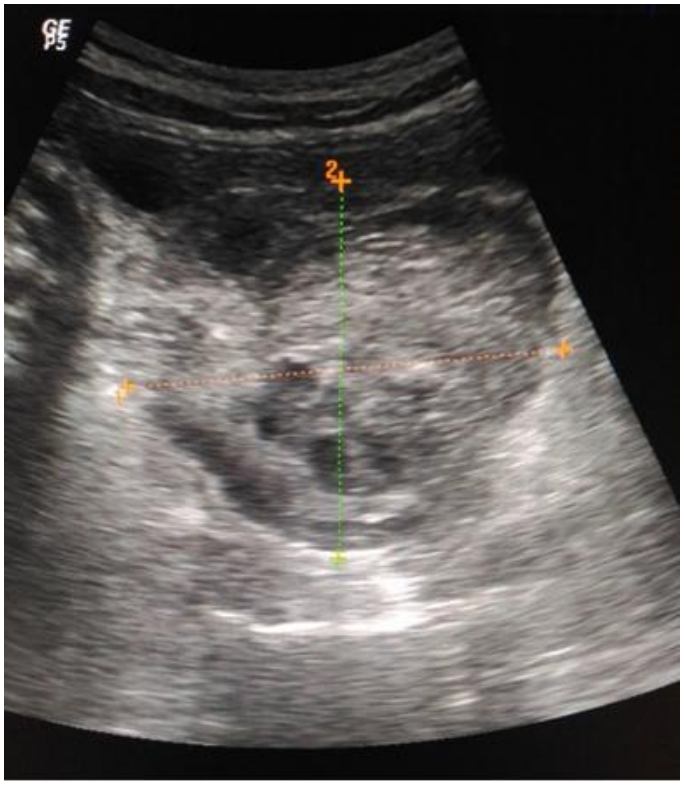

Fig 2 Non- contrast CT showing heterogenous mass with high attenuation

Discussion

Bleeding manifestations are not very common in SCD. There are case reports describing spontaneous extradural hematomas, the majority of which are attributed to skull bone infarction in the same areas as the extradural hematoma. Therefore most cases have suggested that the extradural hematomas are complications of periosteal elevation due to bone infarction with disruption of the cortical bone margin and bleeding into the extradural space[10]. Another mechanism described suggests that patients with SCD have abnormal skull anatomy due to chronic medullary hematopoesis[11] Renal structural and functional abnormalities are commonly associated with SCD and recurrent hematuria is often found in patients with Sickle Cell trait. Although the bleeding usually remits spontaneously, occasionally treatment with antifibrinolytics is necessary[12]. There has been one case report on spontaneous sub capsular splenic hepatoma in a 59 year old patient with Sickle Cell Trait while staying at high altitude and which improved on conservative management[13].

The patient in our report had no prior evidence of bleeding tendencies. The clinical presentation mimicked sickle cell crisis and mesenteric ischemia. Acute appendicitis and mesenteric lymphedenitis were also considered as the differential diagnosis until the CT scan suggested the pelvic hematoma as the definitive cause.

To the best of our knowledge, there have been no prior case reports on spontaneous pelvic hematoma in sickle cell patients. Although the patient improved well with conservative management, a surgical intervention could have be considered if the hematoma failed to resolve over the next few days or increased in size. The pathophysiology of spontaneous pelvic hematoma could be due to periosteal elevation secondary to bone infarction with disruption of the cortical bone margin and bleeding, as described for extradural hematoma by Catherine P et al[10], or from altered pelvic bone anatomy secondary to chronic medullary hematopoesis as described by Dahdeleh et al[11]. However, further research is required to confirm the same.

Conclusion

Spontaneous pelvic hematoma is an exceedingly rare manifestation of sickle cell disease and could present as acute lower abdominal pain in such patients. The possibility of this condition should be considered after ruling out the other common causes of abdominal pain. A non contrast CT of

abdomen and pelvis are necessary to establish the diagnosis and the management would vary based on the size and extent of the hematoma. Further research is required to study the pathophysiology of this condition and other associations.

Conflicts of interests

There are no conflicts of interests.

Supplementary material

1) Ultra sonogram of pelvis showing a hyper echoic lesion with ill defined margins.
2) CT cross sectional image of the pelvis showing heterogenous mass suggestive of hematoma in the pelvis. The hematoma is not in continuity with the uterus or the bony pelvic rim.

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