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Case Report

Testicular infarction in a patient with sickle cell anemia: A case report

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ABSTRACT

Sickle cell anemia is one of the common inherited hemoglobinopathy disorders. It affects patient quality of life through different types of complications. Brain, lung, kidney, spleen, and bone marrow commonly affected; however, testicular infarction is rarely happened. This is a 29-year-old Saudi male who is known for sickle cell disease (SCD). He presented with left scrotal pain and swelling for 1 week which did not respond to antibiotic. Scrotal ultrasound showed swollen heterogeneous hypoechoic left testicle with no blood flow, suggesting testicular infarction. Despite medical treatment, the patient went for orchiectomy. Pathology revealed extensive testicular infarction. Up to our knowledge, this is the first case report of testicular infarction in SCD in Arab countries, and the sixth case reported worldwide. Early diagnosis and doing unnecessary surgical intervention still unavoidable as there is lack of information. We think writing the first case of testicular infarction in sickle cell patients with Arab haplotype is worthful and it will add to the literature.

Keywords: Testicular infarction, Sickle cell complication, Vaso-occlusive

Introduction

Sickle cell anemia is one of the common inherited hemoglobinopathy disorders in Saudi Arabia.[1] It carries major public health burden.[1] Vaso-occlusive complications have been reported in brain, lung, kidney, spleen, and bone marrow with rare involvement to the testes.[2] Sickling of erythrocytes secondary to polymerization of deoxygenated sickle hemoglobin results in loss of their pliability in microcirculation, leading to obstruction.[2] Although sickling process is central to the pathophysiology of vaso-occlusion, abnormalities in hemoglobin structure and function, red blood cell membrane integrity, erythrocyte density, endothelial activation, microvascular tone, inflammatory mediators, and coagulation factors have been established to be responsible too.[3] Suspicion of testicular infarction should be considered in sickle cell disease (SCD) patients who presented with acute painful scrotal swelling not responding to antibiotic.[4] Up to our knowledge, this is the first case report of testicular infarction in SCD in Arab countries, and the sixth case reported worldwide. Early diagnosis and doing unnecessary surgical intervention still unavoidable as there is lack of information. We think writing the first case of testicular infarction in sickle cell patients with Arab haplotype is worthful and it will add to the literature.

Case Report

A 29-year-old Caucasian man from East province of Saudi Arabia who's known for homozygous sickle cell anemia admitted to the hospital with acute left scrotal pain and swelling for 1 week that did not respond to antibiotic. The patient had been in his usual health until 1 week before this admission when pain in the left testis developed. The pain started suddenly, progressive and was associated with intermittent fever (38.2) and progressive enlargement of the left scrotum. There was no history of trauma or lower urinary tract infections symptoms or hematuria or urethral discharge. 1 week before this admission, he presented to another health center for evaluation where he was diagnosed with urinary tract infection and managed as outpatient with a prescription for 7-day course of amoxicillin/clavulanate potassium. The SCA course of this patient was remarkable for recurrent vaso-occlusive crisis and splenectomy at the age of 13 years, but there were no complications such as acute chest syndrome, cerebrovascular accidents, or genitourinary complications. He is not known to have any other medical problems, and he is not on any medications currently apart from intermittent use of analgesics. His vaccinations are up to date. There is additional history for discontinuation of hydroxyurea 4 months ago for improving fertility. He was married and worked as a pharmacist. He did not smoke, drink alcohol, use illicit drugs, or any kind of herbs. He had a family history of diabetes mellitus and hypertension. On examination, the temperature was 37.1°C and pulse 100 beats/min; the other vital signs were normal. The height of the patient was 163 cm and his weight was 55 kg. Swollen, red, and tender left scrotum was present. The examination was otherwise normal. Ultrasound scrotum showed heterogeneous hypoechoic left testicle with no blood flow [Figure 1].

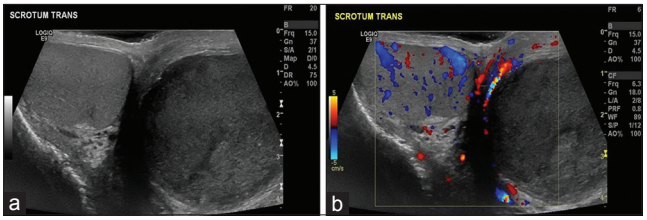

Figure 1: (a-b) Large left testicle with heterogeneous echogenicity and absent color flow, in compared to normal appearing right testicle

Laboratory investigations showed white blood cells 40 (3.5–10.5 billion cells/L), hemoglobin 80 (135–170 g/L), mean corpuscular volume 80 (80–96 fl), platelets 1140 (150–450 billion/L), and reticulocytes 10.7%. Hemoglobin electrophoresis came out with hemoglobin S 79% and hemoglobin F 4.6%. Urine analysis was unremarkable. Conservative strategy has been followed with this patient. He received antibiotics (piperacillin/tazobactam 4.5 g IV q6 h), parenteral hydration, morphine, high-dose hydroxyurea (1000 mg oral daily), and aspirin (81 mg oral daily). There was no significant improvement of patient's symptoms for 7 days. Repeated ultrasound confirmed aforementioned findings. The urologist decided to proceed with the left orchiectomy. Pathology resulted in extensive infarcted testicular tissue with marked acute inflammatory changes and focal hemorrhage of the testis.

Discussion

On literature review of similar conditions, we noted only five cases reported internationally. This is a first case to be reported from Arab countries. The five cases were diagnosed at the age of 25, 37, 27, 57, and 20 years, respectively.[4-8] All of these cases of sickle cell disorders presented with acute testicular pain and swelling. However, silent testicular infarction could be more contributed to testicular dysfunction than symptomatic patients.[4] Consistent with other reports, our patient presented with acute left testicular pain and swelling that did not respond to antibiotic. Radiological features of four cases showed hypoechoic testicle with no blood flow[4-6,8] while one case showed hyperechoic intratesticular mass with anechoic rim and normal blood flow.[7] Our patient's scrotal ultrasound findings were similar to these cases' findings.[5-6,8] All of these cases treated with orchiectomy. Ultrasonographical features of heterogeneous hypoechoic mass in testicle with no blood flow in Doppler mode suggest the diagnosis but cannot differentiate it from tumor, thus orchiectomy mostly needed.[4] Testicular infarction in SCD is likely to be caused by cells sickling.[2] Other factors such as activation of coagulation and platelets and leukocytosis may result in conditions that facilitate the vaso-occlusion. This patient was having leukocytosis, thrombocythemia with additional risk of ceased hydroxyurea, all of these are likely reflecting his risk for getting acute vaso-occlusive complications.[3,9] The evidence of success of hydroxyurea in decreasing the frequency of acute complications in SCD is well documented in literature.[10] Therefore, high-dose hydroxyurea was given aiming to reduce the leukocytes. Reporting cases of similar clinical and radiological features will decrease the need for surgery and improve the diagnostic and preventive strategy.

Conclusion

Testicular infarction is a confirmed complication of SCD. Clinical and radiological features were consistent in all reported cases, although surgical intervention was the eventual approach. This case highlights more studies are needed to comprehend the knowledge of risk factors, prevention, and management.

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References

1. Memish ZA, Owaidah TM, Saeedi MY. Marked regional variations in the prevalence of sickle cell disease and β-thalassemia in Saudi Arabia: Findings from the premarital screening and genetic counseling program. J Epidemiol Glob Health 2011;1:61-8.
2. Claudino MA, Fertrin KY. Sickling cells, cyclic nucleotides, and protein kinases: The pathophysiology of urogenitaldisorders in sickle cell anemia. Anemia 2012;2012:723520.
3. Ilesanmi OO. Pathological basis of symptoms and crises in sickle cell disorder: Implications for counseling and psychotherapy. Hematol Rep 2010;2:e2.
4. Li M, Fogarty J, Whitney KD, Stone P. Repeated testicular infarction in a patient with sickle cell disease: A possible mechanism for testicular failure. Urology 2003;62:551.
5. Mueller FE. Segmental testicular infarction in sickle cell anemia. Urologe A 2014;53:725-7.
6. Gofrit ON, Rund D, Shapiro A, Pappo O, Landau EH, Pode D, *et al.* Segmental testicular infarction due to sickle cell disease. J Urol 1998;160:835-6.
7. Holmes NM, Kane CJ. Testicular infarction associated with sickle cell disease. Urologe 1998;160:130.
8. Urwin GH, Kehoe N, Dundas S, Fox M. Testicular infarction in a patient with sickle cell trait. Br J Urol 1986;58:340-1.
9. Turhan A, Weiss LA, Mohandas N, Coller BS, Frenette PS. Primary role for adherent leukocytes in sickle cell vascular occlusion: A new paradigm. Proc Natl Acad Sci U S A 2002;99:3047-51.
10. Nevitt SJ, Jones AP, Howard J. Hydroxyurea (hydroxycarbamide) for sickle cell disease. Cochrane Database Syst Rev 2017;4:CD002202.