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RESEARCH ARTICLE

PRIAPISM AND CHRONIC MYELOGENOUS LEUKEMIA.

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Abstract

Chronic myeloid leukemia (CML) is a cancer of the white blood cells and the bone marrow. It is characterized by an increased and unregulated growth of myeloid cells in the bone marrow and accumulation of these cells in the blood. There are three stages of CML, and most patients are diagnosed incidentally with an elevated white blood cell count seen on the complete blood count (CBC). Occasionally, CML patients present with upper quadrant pain due to hepatosplenomegaly. An enlarged spleen may put pressure on the stomach, causing loss of appetite and weight loss. CML patients may also present with fever, night sweats, bleeding, or petechiae. An initial presentation of the CML patient with priapism is very unusual and rare. In this case report, we are documenting a case of a 22-year-old man who presented with a persistent painful erection of the penis for four days. After emergency treatment for the priapism, the patient underwent multiple investigations including blood morphology, bone marrow aspiration, and BCR-ABL molecular testing, which confirmed the diagnosis of CML. The purpose of this study is to highlight the rare presentation of CML and the importance of diagnosing the underlying cause responsible for the acute presentation.

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Introduction:-

Chronic myeloid leukemia (CML) is a cancer of the white blood cells and bone marrow. It is characterized by an increased and unregulated growth of the myeloid cells in the bone marrow and accumulation of these cells in the blood. Abnormal cells which are seen in CML are; myeloid, monocytic, erythroid, megakaryocytic, B-lymphoid, and occasionally T-lymphoid lineages [1].

The three stages of the CML are the blast phase, accelerated phase, and the chronic phase. Most of the patients are diagnosed incidentally with an elevated white blood cell count on the CBC during their chronic asymptomatic period. Common findings which are encountered in CML patients are fatigue, weight loss, abdominal fullness,

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bleeding, purpura, splenomegaly, leukocytosis, anemia, and thrombocytosis [1]. Less commonly, CML patients can present (mostly in chronic phase) with priapism and incidence of priapism in adult leukemic patients is about 1-5%. In the pediatric population, it is even rarer [2]. The following case illustrates priapism as an initial presenting symptom of an undiagnosed CML patient.

Case Presentation:-

An otherwise healthy 22-year-old man presented to the emergency department with persistent, and painful penile erection for four days accompanied by bleeding during micturition. There was no history of trauma, fever, night sweats, joint pain or rash. He did not have any antecedent infection. The patient's past medical, surgical and family history was unremarkable. He had no known allergic reaction to food or prescription medications. He did not use any illicit drugs.

His vital signs on examination were (1) Temperature: afebrile (2) Blood Pressure: 135/85 mmHg (3) Respiratory Rate: 21 breaths/min (4) Heart Rate: 108 beats/min. Pulse oximetry showed 98% oxygen saturation in room air. He was alert and oriented to time, place, and person. Physical examination of the patient revealed a palpable liver 5 cm below the right costal margin, and the spleen was palpable 6 cm below the left costal margin. The penis was erect, firm, and tender with superficial venous engorgement. The rest of the systemic examination was unremarkable.

Laboratory findings revealed the erythrocyte sedimentation rate (ESR) = 02 mm/hr (normal range: 0-22 mm/hr for men), hemoglobin = 8.2 mg/dl (normal range: 13.8-17.2 mg/dl in men), white blood cells = 218,600/cumm (normal range: 4000-11,000/cumm), platelets = 324,200/cumm (normal range: 150,000-450,000/cumm). His serum uric acid and creatinine levels were within the normal range.

The patient was scheduled for the aspiration of blood and irrigation of the corpora cavernosa at the emergency department for his painful, sustained erection. The patient felt relief after these procedures, and he was admitted to the Hematology department for further investigations of his hyperleukocytosis. The peripheral blood smear of the patient showed myeloid hyperplasia with neutrophilia, myelocytes, and metamyelocytes. The patient was scheduled for a bone marrow biopsy. The biopsy showed myeloid hyperplasia with the predominance of neutrophils and metamyelocytes. The patient's peripheral blood smear and bone marrow findings raised the suspicion of CML. Detection of BCR-ABL confirmed the diagnosis of Chronic phase CML, with priapism as the initial presentation. The patient was started on hydroxyurea 1.5 grams daily and one vial of interferon alfa-2a subcutaneously daily. Allopurinol 300 mg daily with adequate hydration was also started. Before discharge, his white blood cells dropped to 76000/cumm and hemoglobin raised to 10 mg/dl. In subsequent visits, recurrent priapism did not happen, and his leukemia was in remission.

Discussion:-

Priapism is defined as a persistent penile erection that continues at least four hours unrelated to sexual stimulation [3]. It is a urological emergency, which must be treated early to prevent erectile dysfunction. Priapism is a rare condition on its own with an incidence of 1.5:100,000 person-year [4]. In men, 20% of the cases are caused by the hematological conditions such as sickle cell anemia, chronic myelogenous leukemia, chronic lymphocytic leukemia, and acute lymphoblastic leukemia [2]. In adult leukemic patients, the incidence of priapism is 1-5%, and in the pediatric population, it is even rarer. However, as an initial presenting feature of CML, priapism is seen in 1-2% of cases only.

In patients with leukemia, priapism mainly occurs due to hyperleukocytosis which results in blockade of the veins [3]. Two mechanisms are proposed for the development of priapism in leukemic patients. In the first theory, sludging of leukemic cells in the corpora cavernosa and the dorsal penile vein. The second mechanism proposes that the sacral nerves and the central nervous system become infiltrated with leukemic cells. In this study, we report a case of a 22-year-old man who came to the emergency department with an initial presentation of priapism for CML. It is a rare clinical presenting feature and only seen in 1-2% of cases.

There are two types (high flow and low flow) of priapism, and the management varies with the type. Low flow (ischemic) priapism is mostly seen in CML due to occlusion of corpora cavernosa, and therefore it is considered of the urological emergency requiring urgent intervention. Because priapism in CML is a rare occurrence, there is no standard treatment according to the guidelines. However, the American Urological Association (AUA) has published some guidelines for the treatment of ischemic priapism. AUA recommends treating the ischemic priapism

and then doing investigations to find out the real cause of the priapism [5]. Once the cause is identified, then the next step is to treat the condition. According to the AUA guidelines, it is required to administer intra-cavernosal treatment concurrently.

In our case, the patient felt relief after the cavernosal aspiration and epinephrine irrigation. After that, the patient underwent multiple investigations and was diagnosed with chronic phase CML. The patient was started on medications for CML and also given allopurinol 300 milligrams and hydroxyurea 2 grams daily to ensure cytoreduction. The primary objective of this study is to highlight the importance of principal diagnosis and management of priapism, as there is a very high risk for impotence following this complication. Apart from the initial management of the priapism, the need for further investigations cannot be overemphasized as it can lead to the proper diagnosis of the underlying cause.

Conclusions:-

CML is a cancer of the white blood cells and bone marrow. It is usually diagnosed incidentally, in its chronic asymptomatic phase. The most common symptoms in its presentation are fatigue, weight loss, abdominal fullness, bleeding, purpura, splenomegaly, leukocytosis, anemia, and thrombocytosis. However, in rare cases (1-2% of adults) it can present with priapism as its initial presenting complaint. In this study, we would like to highlight the importance of principal diagnosis and management of priapism, as delayed treatment of priapism has a high risk of impotence. Also, we would like to emphasize the need to evaluate the patient further once priapism has been managed, to check for the underlying cause of the priapism and appropriately managing the condition to prevent relapses of the priapism.

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