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General Surgery

A RARE PRESENTATION OF ACUTE LYMPHOBLASTIC LEUKEMIA AS PRIAPISM IN PAEDIATRIC AGE GROUP

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ABSTRACT Priapism is a prolonged penile erection lasting for more than 4 hours, rarely seen in children. Our patient is a 12 year old previously healthy male patient who presented to emergency with complaints of painful Penile erection. On workup, hemogram coupled with bone marrow aspiration investigations revealed acute lymphoblastic leukemia L1 to be the root cause. Emergency decompression under penile block by aspiration from corpora cavernosa by a lateral approach (3 o clock and 9 o clock position) was done followed by injection of phenylephrine (sympathomimetic agent) in the cavernosa muscles to relieve the ischaemic priapism. The presence of dark, deoxygenated blood confirms ischaemic priapism. Priapism in paediatric age group is a very rare occurance. Ischaemic priapism is the most common type of priapism in children and is a surgical emergency.

KEYWORDS : Paediatric surgery, priapism, ischaemic, acute lymphoblastic leukemia, surgical emergency, corpora cavernosa.

INTRODUCTION:

Priapism is a prolonged penile erection lasting for more than 4 hours unrelated to sexual stimulus, rarely seen in children.***(1)*** There are 3 types of priapism: a) ischaemic priapism (Most common); b) stuttering priapism (recurrent, self-limiting prolonged erection); and c) non-ischaemic priapism (rarest, due to trauma). Ischaemic priapism is a surgical emergency causing fibrosis of the corpora cavernosa, subsequent erectile dysfunction and penile disfigurement. The commonest causes of priapism in children are sickle cell disease (65%), leukaemia (10%), trauma (10%), idiopathic (10%), and pharmacologically induced (5%).

Ischaemic priapism, commonest type seen in children, is typically painful. Sexual activity (including masturbation) and nocturnal erections are common precipitators. Typical features are marked rigidity of the corpora cavernosa with a flaccid glans and spongiosum. Elevated interstitial pressures ensue, causing microvascular compromise and ischaemia(a compartment syndrome within the tunica albuginea). ***(2)***

Stuttering priapism is recurrent "unwanted and painful erections" which are mostly self-limiting but may precede an "unrelenting" ischaemic priapism . Nocturnal erections are the most common trigger. It was first described in 1980 in Jamaican sickle cell disease (SCD) patients, in whom they often start in childhood.***(6)*** Stuttering priapism has a significant effect on quality of life: recurrent visits to healthcare providers (often at night), sleep deprivation, embarrassment, and sexual performance anxiety. ***(5)***

Non-ischaemic priapism is a partial erection due to unregulated cavernous arterial flow and is usually painless. Piesis sign (perineal compression resulting in penile detumescence which recurs after removal of perineal pressure) strongly suggests non-ischaemic priapism in children.***(7)***

The incidence of priapism in males of any age is estimated at 0.3–1.5 per 100,000 per annum, most frequently affecting men in their fifth decade. There is no data on the prevalence of priapism in all children, which is considered rare.***(3) (4)***

Case Report:

History:

A 12 year old, previously healthy male patient, presented with complaint of bilateral swelling in the neck which was painless for past 15 days, for which FNAC was done revealing reactive lymphoid hyperplasia. He also complained of painful penile erection/swelling for the past 1 day. He also had an associated history of fever for the past one day. He had associated history of acute retention of urine for which he took treatment at private hospital (given disodium hydrogen after which he could pass urine). He had no significant past medical/surgical history and no similar complaints in other family members. His birth history was not significant. His development was normal as per age and his immunisation was complete to his age.

Examination And Investigations:

The patient was vitally stable, with no focal neurologic deficits. Local examination revealed an erect and swollen penis which was tender. An inguinoscrotal ultrasound was done which showed corpora cavernosa appears bulky, heterogenous with multiple internal cystic spaces and cavernosal artery shows significant internal vascularity on colour Doppler. An urgent complete hemogram revealed total leukocyte count of 2.83 lakhs with 80% blasts undifferentiated with associated thormbocytopenia, suggestive of Acute Leukemia as the root cause of the priapism. A bone marrow aspiration study was done which revealed hypercellular marrow with increased lymphoblastic proliferation showing 80% lymphoblasts suggestive of Acute Lymphoblastic Leukemia (ALL-L1).

Clinical Picture During Presentation Showing Painful, Erect Penis.

Surgical Intervention:

An urgent decompression of the penis was done wherein a wide bore needle was inserted into the corpus cavernosa muscles and the blood

was aspirated out, after which the penis became flaccid and tenderness was relieved. This was followed by injection of phenylephrine in the cavernosa muscles. The child withstood the procedure well and was free of pain after the procedure. No adverse reactions to the phenylephrine injection was noted.

Surgical Aspiration Of Corpora Cavernosa

DISCUSSION:

Ischaemic priapism is the most common type of priapism seen in children and is typically painful. Most common precipitators are sexual activity(including masturbation) and nocturnal erections. There is marked rigidity of the corpora cavernosa with a flaccid glans and spongiosum. Sickle cell disease is the most common cause of priapism in children followed equally by Leukemia, trauma and idiopathic causes.

Hinman's classic theory that "congestion and slowing of the blood stream" increases blood viscosity and subsequent ischaemia cause ischaemic priapism is supported by the presence of dark, deoxygenated blood when aspirating the corpora (as observed in our case).***(1)*** Childhood leukaemias may cause priapism: hyperleucocytosis and thrombocytosis are commonly seen. In hyperleucocytosis , direct interaction between leukaemic blasts and endothelial cells causes a loss of vascular integrity, activating prothrombotic mechanisms, hence an increased risk of pulmonary or cerebral leucostasis. Cavernosal leucostasis may similarly lead to thrombus formation and venous outflow obstruction, activating the common pathway and hence ischaemic priapism.***(8)***

Management:

Ischaemic Priapism require urgent surgical management as it may cause future ED, anxiety, attenuated sexual aversion behaviour, and intimacy avoidance due to a fear of recurrent priapic pain.

Initial management aims to assess the type of priapism and achieve detumescence . Opiate analgesia is usually required in ischaemic priapism, which may inhibit tumescence.***(9)*** This is followed by urgent surgical decompression by Corporal aspiration and lavage followed by injection of phenylephrine or adrenaline and observation for hypertensive episodes.

If repeated sympathomimetic ICI is unsuccessful, then a surgical fistula ("shunt") should be formed, bypassing the pathological veno-occlusion and allowing blood drainage. Shunts can be distal (cavernoglanular: percutaneous – Winter/T-shunt, or open – Al-Ghorab), proximal (cavernospongiosal – Quackels) or cavernovenous (saphenous – Greyhack). ***(1) (12)***

Corporal aspiration and lavage: Lateral (3/9 o'clock) mid-shaft corporal needle access is obtained, avoiding the urethra or dorsal neurovascular bundle. A 23–21 G (blue/green) butterfly needle should be used in pre-pubescent boys and 19 G (white) needle in adolescents. Minimizing needle passages using a three-way tap and applying pressure for 5 min after removal reduces haematoma rates. Rare complications include infection, urethral lesions and non-ischaemic priapism.

The presence of dark, deoxygenated blood ($pO_2 < 40$ mmHg) confirms ischaemic priapism. In ischaemic priapism the corpora should be immediately decompressed: 3–5-mL aliquots should be aspirated until bright red (oxygenated) blood is seen (not exceeding 10% of the circulating blood volume; 7.5 mL/kg in children aged ≥1 year) . The corpora should then be flushed with warmed 0.9% saline. ***(10) (11)***

CONCLUSION:

Ischaemic priapism in paediatric age group is a rare finding and a surgical urological emergency which requires urgent decompression by a paediatric surgeon followed by close monitoring and further workup of the child to find the root cause of priapism. If left untreated, it leads to necrosis, fibrosis and invariably future ED. This patient had a timely intervention which relieved the ischaemic priapism and follow-up bone marrow aspiration study revealed Acute Lymphoblastic Leukemia as the root cause of priapism.

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