

A Rare Presentation of Partial Segmental Thrombosis of the Corpus Cavernosum Secondary to Sickle Cell Anemia

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Abstract

Background: Partial segmental thrombosis of the corpus cavernosum, known as partial priapism, is an uncommon urological condition which predominantly affects young men in which the proximal part of one corpus cavernosum is thrombosed. Many risk factors have been described in the literature, however, the exact etiology of penile thrombosis and its pathogenesis remains unclear. Several treatment options are available ranging from conservative medical treatment, surgical intervention, or simple follow-up observation without treatment. **Aim:** In this study, we describe a patient with sickle cell anemia who presented with pain and a perineal swelling that was eventually diagnosed as partial priapism utilizing MRI scan and was treated conservatively with a successful outcome. We then performed a literature search of similar cases highlighting incidence, risk factors and management of this rare presentation. **Case Presentation:** A 23-year-old male who is known with sickle cell anemia presented to casualty with a 1-day history of perineal pain of a sudden onset associated with perineal swelling and vomiting. Genitourinary exam findings confirmed the absence of classic priapism. Careful examination of his perineal area revealed the presence of a fixed, hard, and tender mass at the proximal part of the penis. It was not attached to the overlying skin and no enlarged pelvic lymph nodes were felt. Once stabilized, MRI of the pelvis was performed showing right intra-tunicular corpus cavernosum features suggestive of hematoma in keeping with partial segmental thrombosis of the corpus cavernosum. Conservative treatment was initiated, and the patient was managed expectantly in which he improved gradually with eventual disappearance of the perineal mass. **Conclusion:** Partial segmental thrombosis of the corpus cavernosum is a rare urological condition. Pathogenesis and etiologies are poorly understood but risk factors have been advocated of which sickle cell anemia is one of them. MRI has a crucial role in the diagnosis under this condition. Conservative treatment appears to be a reliable initial therapeutic option.

Keywords

Partial Segmental Thrombosis of the Corpus Cavernosum, Risk Factors, Sickle Cell Anemia, Conservative Management

1. Introduction

Partial Segmental Thrombosis of the Corpus Cavernosum (PSTCC) is an uncommon urological condition which chiefly affects young men in which the proximal part of one corpus cavernosum is thrombosed [1]. Some risk factors have been described in the literature, however, the exact etiology and pathogenesis remain unclear. It is essential to clarify that PSTCC is different from classic priapism since it typically manifests without an erection [2]. Although partial priapism is the commonly used term to characterize this unusual clinical condition, PSTCC describes it in a better way [3]. Several treatment options are available ranging from conservative medical treatment, surgical, and simple follow-up observation without treatment [2]. Herewith we report a case of a Sickle Cell Anemia (SCA) patient presenting with perineal pain and swelling diagnosed as PSTCC and treated conservatively.

2. Case Report

A 23-year-old male known with SCA presented to casualty with a one-day history of sudden onset perineal pain, nausea, and vomiting without fever. The pain was associated with perineal swelling. There was no history of urethral discharge, sexual contact, trauma, substance abuse, or lower urinary tract symptoms. Past history was unremarkable except for a stable SCA and left pyeloplasty. Initial examination revealed stable vital signs and unremarkable abdominal examination. Genital examination was within normal limits with no signs of priapism. Nevertheless, careful examination of his perineal area revealed the presence of a fixed, hard and tender mass at the proximal part of the penis. It was not attached to the overlying skin and no enlarged pelvic lymph nodes were felt (**Figure 1**). Digital rectal examination was normal. Initial laboratory tests were normal except for mild elevation in the leukocytic count. Once stabilized, MRI of the pelvis was performed. It showed an intact Buck's fascia and tunica albuginea. The right intra-tunical corpus cavernosum showed altered signal intensity involving the root, proximal and mid third sparing the distal third with a maximum width of 26 mm (predominantly hyposignal intensity with few areas of hypersignal intensity) (**Figure 2**). Features were suggestive hematoma in keeping with PSTCC. Conservative treatment was initiated with bed rest, analgesia in the form of 6 hourly intravenous acetaminophen and regular bed-side examination. The response was observed over 48 hours in which there was gradual but steady decrease in the pain, swelling size and leukocytic count that normalized. Patient had an uneventful recovery and was discharged on the



Figure 1. Tender mass at the proximal part of the penis. The mass was not attached to the overlying skin.



Figure 2. T2 signal intensity drops suggestive of right corpus cavernosum hematoma (indicated by the red arrow).

third day. At follow up 2 and 4 weeks later, patient was pain free and the mass had disappeared completely. He also revealed that he had been having normal erection without any pain or prolonged erection. Patient was reassured and a hematology follow up was arranged.

3. Discussion

This case highlights a rare yet interesting clinical entity. The literature has seen a rise in the reported cases due to the increase in the awareness of this clinical findings and the presence of advanced imaging modalities. PSTCC affects mainly young men and rarely affects other age groups [1]. Almost all cases described in literatures presented with unilateral painful perineal mass that is tender on palpation involving the proximal cavernosal region with or without partial priapism. The differential diagnoses include inflammatory processes and solid tumors [4]. Although several risk factors are described in the literature, the exact

etiology remains unclear in most cases. Risk factors for development of PSTCC include hematological abnormalities, blood transfusion, alcohol or recreational drugs abuse, and some medications [5] [6] [7]. Other possible causes include hypercoagulability associated with malignancy and long-distance flights and microtrauma after vigorous sexual intercourse or extensive bicycle riding [1] [2] [3] [8] [9] [10]. Ilicki *et al.* [1] postulated a two-hit model to explain the pathogenesis. Firstly, transverse membrane is required, dividing the corpus into a proximal and distal portion. In this line, Hillis and Weems described this as a transverse membrane separating the proximal thrombotic area from the distal flaccid corpora, the membrane is congenital or posttraumatic, unilateral or bilateral and could be identified on MRI study or during surgical exploration [11]. Secondly, obstructing the permeable membrane by trigger factors (possibly micro-trauma), that may lead to clotting of the blood that shut the proximal portion. Indeed, this model can justify several points e.g., its association with hematological disorders or micro-trauma, and how it may resolve spontaneously without treatment [1] [11]. However, it is a theoretical explanation since histological and pathological validation is lacking. In our case, the only risk factor that could have led to such a presentation was SCA.

The diagnosis is reached by detailed history, physical examination and advanced imaging studies, but the condition should be distinguished carefully from other penile diseases such as penile fracture, penile abscess, and penile cancer [12]. Clinical examination in the case we described was crucial to detect the perineal mass. In the past, a definitive diagnosis was possible during surgery. Currently, sonography, MRI and CT are standard imaging tools [13]. MRI is more definite and reliable than CT in diagnosing PSTCC as it displays the pathologic processes in soft tissue in greater visual details [1] [2] [3] [12] [14]. Utilizing MRI in the patient we encountered was of paramount importance in confirming the diagnosis. On MRI, the lesion is evident as a unilaterally distended segment of the corpus cavernosum, frequently compressing the contralateral corpus cavernosum due to mass effect of the thrombosed segment. Recently, contrast-enhanced ultrasound has been utilized, particularly in patients in which MRI is contraindicated [13]. It also helps to avoid invasive diagnostic tools, such as cavernosography, cavernous biopsy or surgical exploration [2]. PSTCC is not considered a urological crisis and several treatment options have been described. The first case of partial priapism was described by Hillis in 1976 [11]. At that time, surgical intervention was the norm including corporotomy, cavernosum-spongiosum shunt, and intracavernous injection [5] [15] [16].

Recent literature described some conservative medical approaches with fair success to preserve erectile function [11] [15] [16] [17]. Surgery is reserved for patients failing conservative measures [9]. Fortunately, PSTCC has an excellent prognosis without serious complications or shortcomings. Rare complications can occur such as failure to recover a complete erection (loss of rigidity) in one case, or difficulty with a full erection after surgical management in another case

[11] [12]. In the patient we reported, conservative measures were successful, recovery was excellent, and no complications were observed.

4. Conclusion

PSTCC is a rare urological condition which predominantly affects young men presenting with pain and perineal mass yet is not a urological emergency. Pathogenesis and etiologies are poorly understood but risk factors have been advocated of which SCA is one of them. MRI has a crucial role in the diagnosis under this condition. PSTCC has an excellent prognosis. Conservative treatment appears to be a reliable initial therapeutic option. Surgery is reserved for patients in whom conservative management fails.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

References

- [1] Ilicki, J., Krauss, W. and Andersson, S.O. (2012) Partial Segmental Thrombosis of the Corpus Cavernosum: A Case Report and a Review of the Literature. *Urology*, **79**, 708-712. <https://doi.org/10.1016/j.urology.2011.11.032>
- [2] Goeman, L., Joniau, S., Oyen, R., Claes, H. and Van Poppel, H. (2003) Idiopathic Partial Thrombosis of the Corpus Cavernosum: Conservative Management Is Effective and Possible. *European Urology*, **44**, 119-123. [https://doi.org/10.1016/S0302-2838\(03\)00217-3](https://doi.org/10.1016/S0302-2838(03)00217-3)
- [3] Horger, D.C., Wingo, M.S. and Keane, T.E. (2005) Partial Segmental Thrombosis of Corpus Cavernosum: Case Report and Review of World Literature. *Urology*, **66**, 194. <https://doi.org/10.1016/j.urology.2005.01.011>
- [4] Gentile, G., Broccoli, A., Brunocilla, E., Schiavina, R., Borghesi, M., Romagnoli, D., *et al.* (2013) An Isolated Penile Mass in a Young Adult Turned Out to Be a Primary Marginal Zone Lymphoma of the Penis. A Case Report and a Review of Literature. *Anticancer Research*, **33**, 2639-2642.
- [5] Dubois, F., Lesur, G., Azzouzi, A.R., Beurrier, P. and Chautard, D. (2007) Partial Thrombosis of the Corpus Cavernosum. Must a Clotting Disorder Be Systematically Investigated? *Progrès en Urologie*, **17**, 866-868. [https://doi.org/10.1016/S1166-7087\(07\)92310-6](https://doi.org/10.1016/S1166-7087(07)92310-6)
- [6] Hoyerup, P. and Azawi, N.H. (2013) Partial Priapism. *BMJ Case Reports*, **2013**, bcr2013200031. <https://doi.org/10.1136/bcr-2013-200031>
- [7] Kilinc, M., Piskin, M., Guven, S., Gurbuz, R., Odev, K. and Kaynar, M. (2009) Partial Priapism Secondary to Tamsulosin: A Case Report and Review of the Literature. *Andrologia*, **41**, 199-201. <https://doi.org/10.1111/j.1439-0272.2008.00908.x>
- [8] Blaut, S., Schneider, M., Zschuppe, E., Günl, U. and Steinbach, F. (2008) Partial Unilateral Penile Thrombosis of Corpus Cavernosum Due to Hyperhomocysteinemia. Case Report and References. *Der Urologe*, **47**, 748-752. <https://doi.org/10.1007/s00120-008-1650-4>
- [9] Lewis, J.H., Javidan, J., Keoleian, C.M. and Shetty, S.D. (2001) Management of Partial Segmental Priapism. *Urology*, **57**, 169. [https://doi.org/10.1016/S0090-4295\(00\)00851-7](https://doi.org/10.1016/S0090-4295(00)00851-7)

- [10] Schneede, P., Schmeller, N., Müller-Lisse, U.G., Reiser, M.F. and Hofstetter, A.G. (1999) Partial Priapism. Case Report and Review of the Literature of Diagnostic and Therapeutic Measures. *Der Urologe*, **38**, 179-183. <https://doi.org/10.1007/s001200050263>
- [11] Hillis, R.S. and Weems, W.L. (1976) Priapism: An Unusual Presentation. *The Journal of Urology*, **116**, 124-125. [https://doi.org/10.1016/S0022-5347\(17\)58710-8](https://doi.org/10.1016/S0022-5347(17)58710-8)
- [12] Pepe, P., Panella, P., Candiano, G., Garufi, A., Priolo, G. and Aragona, F. (2012) Partial Priapism Secondary to Idiopathic Segmental Thrombosis of Corpora Cavernosa. *Archivio Italiano di Urologia, Andrologia: Organo Ufficiale [di] Societa Italiana di Ecografia Urologica e Nefrologica*, **84**, 101-103.
- [13] Sauer, S., Goltz, J.P., Gassenmaier, T., Kunz, A.S., Bley, T.A., Klein, D., *et al.* (2014) Partial Segmental Thrombosis of the Corpus Cavernosum (PSTCC) Diagnosed by Contrast-Enhanced Ultrasound: A Case Report. *BMC Urology*, **14**, Article No. 100. <https://doi.org/10.1186/1471-2490-14-100>
- [14] Patel, R.P., Mucksavage, P., Ramchandani, P., Hanno, P.M. and Malkowicz, S.B. (2010) Idiopathic Partial Thrombosis of the Corpus Cavernosum. *Urology*, **76**, 1373-1374. <https://doi.org/10.1016/j.urology.2009.12.058>
- [15] Albrecht, W. and Stackl, W. (1997) Treatment of Partial Priapism with an Intracavernous Injection of Etilefrine. *The Journal of the American Medical Association*, **277**, 378. <https://doi.org/10.1001/jama.1997.03540290030027>
- [16] Chary, K.S., Rao, M.S., Kumar, S., Palaniswamy, R., Chandrasekar, D., Vaidyanathan, S., *et al.* (1981) Creation of Caverno-Glandular Shunt for Treatment of Priapism. *European Urology*, **7**, 343-345. <https://doi.org/10.1159/000473261>
- [17] Burkhalter, J.L. and Morano, J.U. (1985) Partial Priapism: The Role of CT in Its Diagnosis. *Radiology*, **156**, 159. <https://doi.org/10.1148/radiology.156.1.4001404>