Bilateral spontaneous thrombosis of the pampiniform plexus mimicking incarcerated inguinal hernia: case report of a rare condition and literature review

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Abstract

Background: Pampiniform plexus thrombosis is a very rare disease (only less than 25 published cases are available till date), and it is a diagnostic dilemma. The present case is an unusual condition of an elderly gentleman who was finally diagnosed as a case of spontaneous thrombosis of bilateral pampiniform plexus and was managed conservatively. Literature was reviewed to explore potential etiologies and therapeutic strategies.

Case presentation: A 65-year-old afebrile gentleman, laborer (in brick industry), and non-smoker with no previous major health problems was admitted with swelling in the bilateral inguinal region. The swelling had started one and half months ago. He had developed severe pain over the swelling for last 1 day with tenderness and indurations. Neither he had history of previous surgeries, chronic cough, dysuria, prostatism, and trauma nor he presented any thrombogenic factors. There was no history of vomiting, abdominal pain, and obstipation. Physical examination revealed normotensive person with BMI of 22.5, was significant only for one tender, movable, and firm to hard 10 cm × 3 cm mass extending from the left deep inguinal ring up to the upper pole of the testis in the scrotum. Another 5 cm × 3 cm mass of similar characteristics was found extending from deep inguinal ring up to the root of the scrotum on right side. The testes and prostate were normal on palpation. On the contrary to preoperative USG, which clinched suspicion of incarcerated inguinal hernia, a thrombosed pampiniform plexus without any evidence of hernia sac was found on the left side during inguino-scrotal exploration. Wound was closed without doing any further procedure. Contralateral inguino-scrotal exploration was spared considering same nature of disease. Postoperative Doppler ultrasonography confirmed the diagnosis of bilateral thrombosed pampiniform plexus. MDCT of whole abdomen revealed no abnormality other than bilateral spermatic cord thrombosis. Blood thrombophilia screening came normal. The subject had an uneventful postoperative hospital course. With 2 years of follow-up, the gentleman is doing well, remaining asymptomatic and had returned to his usual life.

Conclusions: Due to extreme rarity, spontaneous thrombosis of the pampiniform plexus may be a diagnostic dilemma and requires a high index of suspicion. Doppler ultrasound is the initial investigation of choice. In the absence of other concomitant disease, beginning the treatment conservatively instead of excising the thrombosed segment is more suitable.

Keywords: Pampiniform plexus, Incarcerated inguinal hernia, Thrombosis

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Background
Spontaneous pampiniform plexus thrombosis is a diagnostic dilemma, and it is a very rare condition. Less than 25 cases of spontaneous thrombosis have been published in the literature till date [1]. Acute inguino-scrotal or testicular painful swelling is the usual clinical presentation [1], and commonly left spermatic cord gets involved [2]. Preoperatively, it may be misdiagnosed due to its non-specific presentation and as it is clinically indistinguishable from many other inguinal conditions. The present case is an unusual condition of an elderly gentleman, with idiopathic spontaneous thrombosis of bilateral pampiniform plexus. The present report is also the first ever reported case of bilateral pampiniform plexus thrombosis. Literature was also reviewed to explore potential etiologies and therapeutic strategies to manage this extremely rare condition.

Case presentation
A 65-year-old afebrile gentleman, laborer (in brick industry) and non-smoker with no previous major health problems, was admitted for painful swelling in the bilateral inguino-scrotal region. The swelling had started one and half months ago in the bilateral inguinal region, and later, it gradually involved the upper part of the scrotum bilaterally. The swelling was small initially, but gradually attained presenting size in the last 4–5 days. The swelling did not reduce on lying down, but it used to get prominent in standing position. Initially, there was mild dragging and aching pain over the swelling, but the pain was increased and became severe since 1 day with tenderness and indurations. There was no history of vomiting, abdominal pain, dysuria, and obstipation. Neither he had history of previous surgery, chronic cough, prostatism, and trauma nor he presented any thrombogenic factors.

Physical examination revealed normotensive person with BMI of 22.5, was significant only for one left sided elongated, tender, movable, and firm to hard 10 cm (vertical) × 3 cm (horizontal) mass (above the crease of groin and medial to pubic tubercle). It was extending from the left deep inguinal ring up to the upper pole of the testis in the scrotum. Local temperature over the swelling was raised with mild erythema. There was no visible or palpable cough impulse. “Get above the swelling” was not possible. As the swelling was irreducible, the deep ring occlusion test could not be performed. Dull note was found on percussion with no audible gurgling sound. Another 5 cm (vertical) × 3 cm (horizontal) mass of similar character was found extending from the right deep inguinal ring up to the root of the scrotum. Both the testes were normally positioned in the scrotum, but the left one was mildly swollen. Prostate size and penile position were normal. Umbilicus was in normal position in scaphoid abdomen without any tenderness, visible peristalsis, or palpable mass.

Contrary to pre-operative grayscale USG finding (hernia containing tubular loops), which clinched the suspicion of incarcerated inguinal hernia, on exploration of left inguino-scrotal region under spinal anesthesia, left spermatic cord was found to be thick, multi-lobulated, blackish-red colored, tubular mass of firm to hard consistency (Fig. 1a, b). This was thrombosed pampiniform plexus without any evidence of hernia sac, and the testis was found to be mildly congested. Decision of no further intervention was taken. Wound was closed. Contralateral inguino-scrotal exploration was spared considering the same nature of disease (Fig. 2).

Post-operative period was uneventful. The patient was put on anti-inflammatory drugs. Oral feeding was started from the next day, and early ambulation was encouraged. The swelling and pain started to get reduced gradually. There was no episode of shortness of breath or
chest pain, tachycardia, and tachypnea in post-op period. BP was normal throughout the post-operative period. There was no development of calf tenderness. Post-operative ultrasonography with color Doppler study (Fig. 3a, b) confirmed the diagnosis of bilateral thrombosed pampiniform plexus, showing hyperechoic soft tissue mass lesions in bilateral spermatic cords. Very few vessels were seen within the mass with colored flow. Bilateral testis was normal. MDCT and MRI scan of the whole abdomen (Fig. 3c, d, e) revealed no abnormality other than bilateral spermatic cord thrombosis. Blood thrombophilia screening (factor V Leiden, prothrombin time, antithrombin assay, protein C and S, lupus anticoagulant, anticardiolipin antibody) came normal. ECG and urine analysis were normal. There was no surgical site infection. The patient was discharged in a stable condition after 7 days. The subject, with 2 years follow-up, is doing well, remaining asymptomatic and had returned to his usual life (Fig. 4a, b). He was advised regular check-up in surgical out-patients’ department.

Discussions

Spermatic vein thrombosis is an unexpected finding in the differential diagnosis of acutely painful inguinoscrotal region [3]. Most of these cases are initially tried surgically as if they had an incarcerated inguinal hernia [4]. Additionally, epididymitis, spermatic cord disease (such as torsion), or benign and malignant tumors of spermatic cord should be kept in mind in the differential diagnosis [5]. Hashimoto and Vibeto [2] noted that there was a preponderance of left-sided presentations with presumed, shared anatomical factors which can also predispose to varicocele formation. Right spermatic vein thrombosis is an important clinical sign to do detailed research at the renal hilus level or in the retroperitoneal region to rule out renal/retroperitoneal tumors with renal vein, vena cava thrombosis. The present report is also the first ever reported case of bilateral pampiniform plexus thrombosis. The author also studied the characteristics of all available cases [6] in chronological order (Table 1) which revealed (Table 2):

In the etiology of isolated spermatic vein thrombosis, there are many possible predisposing factors, such as trauma to the vascular endothelium, slow venous flow, and hypercoagulability [21]. Kayes et al. reported that spontaneous vein thrombosis could be related to prolonged vigorous activity (e.g., heavy weight lifting, sports, physical training), tumors of the genitourinary tract, infections, trauma, inguinal hernia surgery, long-hour flights, and the use of some drugs [22]. An increase in intra-abdominal pressure linked to these activities may decrease flow within the gonadal venous systems which may be compounded by specific anatomical factors. Most notably, in keeping with a left-sided predominance of this condition, one must consider meso-aortic compression of the left renal and spermatic vein(s), also known as “nutcracker syndrome” [23]. Examination with Doppler ultrasound should be the first-line investigation, while others outlined in previous case reports include a thrombophilia screen [24], MDCT of the abdomen to rule out causes of venous obstruction, incarcerated hernia, or malignancy [12]. As spermatic vein thrombosis is clinically indistinguishable from many other groin conditions, computed tomographic angiography may help to reveal whether the thrombus extends beyond the external inguinal ring. It also helps to find the etiology, such as nutcracker syndrome especially in young male.

In the literature also, no report regarding recurrence was found after conservative management. There are no guidelines available for the management of this disease. Hashimoto and Vibeto reported that there is no need to excise the thrombosed plexus, as evidenced by the good results in their case [2]. Conservative management, including watchful observation and NSAID without anticoagulation, is acceptable for thrombosis out of external inguinal ring (pampiniform plexus). Yoko Kyono et al. proposed surgical excision, and anticoagulation may prevent pulmonary embolism in deep-seated spermatic vein thrombus inside the external inguinal ring and extending to the nearby renal vein [25]. Though the management remains unclear, proximal extension of the thrombosis is the most significant indication for further investigation.
Fig. 3  

a Grayscale ultrasonography demonstrate dilated, non-compressible, thrombosed tubular venous structure with increased wall thickness within the left spermatic cord. Within this tubular structure, focal echoes that belong to thrombus (green arrow) can be seen, but no vascular flow curve can be seen (red arrow).  
b On color Doppler ultrasound, no filling with the color was seen in the lumen of this vein within the left spermatic cord. On Doppler ultrasound, filling was seen within the neighboring arterial structure (green arrows), but not within the vein.  
c Computed tomography scan showing grossly distended and thrombosed spermatic veins (green arrows).  
d Sagittal section and e transverse section fat-compressed axial T1 magnetic resonance images demonstrated thrombosed tubular venous structure (green arrows) with increased wall thickness and focal diameter increase within the bilateral spermatic cord. Within this venous structure, intraluminal signal intensity was increased.

Fig. 4  

a Photograph of inguino-scrotal region on follow-up at 4 months.  
b USG of left spermatic cord shows no thrombus, and full compressibility was noted on 4 months follow-up.
| Serial no. | Age (years) | Location of lesion | Onset of pain | Predisposing factors | Diagnosis (provisional) | Investigations | Management | Publication year and author |
|-----------|-------------|-------------------|--------------|---------------------|------------------------|----------------|------------|-----------------------------|
| 1         | NA          | Left              | NA           | NA                  | Orchitis               | None          | NA         | 1903, Senn [7]               |
| 2         | NA          | NA                | "Sudden"     | None                | Thrombosis             | None          | Excision  | 1904, Senn [8]               |
| 3         | 41          | Left              | 5 weeks      | None                | Orchitis               | None          | Orchidectomy | 1935, Mc Gavin [9]          |
| 4         | 57          | Left              | 4 weeks      | None                | Orchitis               | None          | Orchidectomy | 1935, Mc Gavin [9]          |
| 5         | 27          | Left              | 16 h         | None                | NA                     | None          | Excision   | 1977, Anseline [10]          |
| 6         | 7           | Left              | NA           | None                | NA                     | Venography    | Exploration | 1980, Coolsaet and Weinberg [11] |
| 7         | 10          | Left              | NA           | None                | Thrombosis             | Venography    | NSAID      | 1980, Coolsaet and Weinberg [11] |
| 8         | 15          | Left              | 11 days      | Walking             | NA                     | None          | Excision   | 1980, Coolsaet and Weinberg [11] |
| 9         | 33          | Left              | 10 days      | None                | Incarcerated hernia    | VP            | Excision   | 1981, Vincent and Bokinsky [12] |
| 10        | 44          | Right             | "Hours"      | Playing sports      | Inguinal mass          | None          | Excision   | 1981, Rothman [13]           |
| 11        | 33          | Left              | NA           | Varicocele          | NA                     | None          | Excision   | 1985, Roach et al. [14]      |
| 12        | 42          | Contralateral     | 1 week       | None                | Incarcerated hernia    | VP, cavogram CT scan | Excision   | 1985, Roach et al. [14]      |
| 13        | 23          | Left              | "Hours"      | Heavy weight lifting | Incarcerated hernia    | Doppler USG   | Excision   | 1990, Isenberg et al. [15]   |
| 14        | 19          | Left              | "Hours"      | Vigorous exercise   | Incarcerated hernia    | None          | Excision   | 1993, Gleason et al. [16]    |
| 15        | 27          | Left              | 2–3 h        | Heavy weight lifting | Incarcerated hernia    | None          | Exploration | 2006, Hashimoto et al. [2]  |
| 16        | 33          | Left              | 3 days       | Cycling             | Thrombosis             | Doppler USG   | NSAID      | 2009, Doerfler et al. [17]  |
| 17        | NA          | Contralateral     | NA           | NA                  | Incarcerated hernia    | Doppler USG   | Anticoagulation | 2010, Kayes et al. [18]   |
| 18        | 28          | Left              | 14 days      | Nutcracker syndrome | NA                     | Doppler USG, CT scan | Excision   | 2014, Mallat et al. [19]    |
| 19        | 43          | Right             | 2 days       | Absence IVC, mutation factor V Leiden | NA | Doppler USG, CT scan | Antibiotics anticoagulant | 2015, Chi and Hairston [20] |
| 20        | 39          | Contralateral     | 3 days       | Infection protein C deficiency | Thrombosis | Doppler USG, CT Scan | 2018, Kamel et al. [6] |
| 21 (present case) | 65 | Bilateral           | 1 day        | Heavy weight lifting | Incarcerated hernia | Doppler USG, MRI, CT scan blood test | NSAID | 2020, Bakshi S |

NA no available information, IVP intra-venous pyelogram, CT scan computed tomography scan, IVC inferior vena cava
Conclusions

Isolated spermatic vein thrombosis is a rare event and requires a high index of suspicion. Although present case is bilateral, spermatic vein thrombosis is almost always found at the left side. Doppler ultrasonographic examination is the procedure of choice in the diagnosis of the varicocele thrombosis with higher sensitivity and specificity. Exploratory surgical approach may be needed initially in the absence of Duplex study, to exclude an acutely infarcted testis or incarcerated hernia. But in the confirmed absence of other concomitant disease that necessitates urgent surgical intervention, beginning the treatment conservatively instead of excising the thrombosed segment is more suitable. Although conservative management, including watchful observation and NSAID without anti-coagulation, is acceptable for thrombosis out of external inguinal ring (pampiniform plexus), surgical excision and anticoagulation may prevent pulmonary embolism in deep-seated spermatic vein thrombosis out of external inguinal ring (pampiniform plexus), surgical excision and anticoagulation may prevent pulmonary embolism in deep-seated spermatic vein thrombus (proximal to the external inguinal ring) and extending to the nearby renal vein. Surgeons should be aware of this rare clinical entity for prompt management of potential morbidity.

Abbreviations

BMI: Body mass index; BP: Blood pressure; ECG: Electrocardiography; MDCT: Multi-detector computed tomography; MRI: Magnetic resonance image; NSAID: Non-steroidal anti-inflammatory drugs; USG: Ultrasonography

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Author’s contributions

All works regarding this case report was solely done by Dr. Sabyasachi Bakshi, who is also the corresponding author. The author read and approved the final manuscript.

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Availability of data and materials

Presented within the manuscript. Please contact author for additional data requests.

Table 2 Comparative characteristics of present study

| Parameters                        | Findings after literature review                                                                 | Findings of the present case                                                                 |
|-----------------------------------|---------------------------------------------------------------------------------------------------|---------------------------------------------------------------------------------------------|
| Age at presentation               | Mean age was found 32.27 years (range 7–65 years)                                                | Present case is the eldest of all reported subjects till date                               |
| Location (side)                   | Left sided in 70% cases, 25% in right side                                                       | Present case is the only reported case of bilateral thrombosis                              |
| Duration of pain                  | Varied duration. Ranges from hours to 5 weeks                                                     | In the present case, mild dragging pain started 6 weeks ago                                 |
| Predisposing factors              | Majority reported heavy physical works                                                            | Subject in the present case was also an active physical labor                                |
| Initial diagnosis                 | Majority was diagnosed preoperatively as incarcerated inguinal hernia                            | Present case was also diagnosed as incarcerated inguinal hernia in the emergency department |
| Primary investigation and management| USG Doppler flow study confirmed majority of the cases, and majority were managed by surgical excision | USG Doppler confirmed diagnosis. But the case was managed conservatively                     |

Consent for publication

Written consent to publish was obtained for the publication of all clinical details and images, and the consent form is available for review by the editor of the journal.

Competing interests

The author declares that he has no competing interests.

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References

1. Castilho OA, Díaz M, Vitagliano GJ, et al. Pulmonary thromboembolism secondary to left spermatic vein thrombosis: a case report. Urol Int. 2008;80:17–8.
2. L. Hashimoto Brett Vibeto. Spontaneous thrombosis of the pampiniform plexus. Scand J Urol Nephrol, 40 (2006), pp. 252–254.
3. Kleinlauss F, Della Negra E, Martin M, et al. Spontaneous thrombosis of left varicocele. Prog Urol. 2001;11:95–6.
4. Campagnola S, Fessati P, Fasoli L, et al. A rare case of acute scrotum. Thrombophlebitis from ectasia of the left pampiniforme plexus. Minerva Urol Nephrol. 1999;51:163–5.
5. Gleeson MJ, McDermott M, McDonald G, McDermott TE. Spontaneous thrombosis of the left spermatic vein. Br J Urol. 1992;70(5):567.
6. Kernel K, et al. Bilateral spontaneous thrombosis of the pampiniform plexus; A rare etiology of acute scrotal pain: A case report and review of literature. Afr J Urol. 2018;24:14–8. https://doi.org/10.1016/j.afrju.2017.09.006.
7. Senn NA. Surgical clinic. Clin Rev. 1934:241–5.
8. Senn NA. Vesical calculus; thrombosis of the spermatic veins; cervical tuberculous lymphadenitis; sarcoma of the submaxillary gland; sandtectly; traction injury of the peroneal nerve; paralysis of the circumflex nerve; raschis; acute osteomyelitis of the os calcis; adenomatous goiter; ganglion. Int Clin. 1904:4:148–60.
9. Mc Gavin D. Thrombosis of the pampiniform plexus. Lancet. 1935;226(5842):368–9. https://doi.org/10.1016/S0140-6736(00)77242-4.
10. Anseline P. A case of spontaneous thrombosis of the testis. Aust N Z J Surg. 1977;47:801–2.
11. Coolsaet B, Weinberg R. Thrombosis of the spermatic vein in children. J Urol. 1980;175:4–5.
12. Vincent MP, Bokinsky G. Spontaneous thrombosis of pampiniform plexus. Urology. 1981;7:175–6.
13. Rothman D. Thrombosis of the pampiniform plexus. J Med Soc New Jersey. 1981;78:681.
14. Roach R, Messing E, Starling J. Spontaneous thrombosis of left spermatic vein: report of 2 cases. J Urol. 1985;134:369–70.
15. Eisenberg J, Oster G, Worth WH, Feizl G. Effect-induced spontaneous thrombosis of the left spermatic vein presenting clinically as a left inguinal hernia. J Urol. 1990;146:138.
16. Gleason TP, Balsara Z, Goff WB. Sonographic appearance of left spermatic vein thrombosis simulating incarcerated inguinal hernia. J Urol. 1993;150:1513–4.
17. Doerfler A, Ramadani D, Meuwly JY, et al. Varicocele thrombosis: a rare etiology of testicular pain. Prog Urol. 2009;19:351–2.
18. O. Kayes, N. Patrick, A. Sengupta. A peculiar case of bilateral, spontaneous thrombosis of the pampiniform plex. Ann R Coll Surg Engl, 92 (2010), pp. W22-W23.

19. Mallat F, Hmida W, Ahmed KB, et al. Spontaneous spermatic vein thrombosis as a circumstance of discovery of nutcracker syndrome: an exceptional entity. Int J Case Rep Images. 2014;5:519–23.

20. Chi AC, Hainston JC. Acute right varicocele: a clue to congenital vascular anomaly. Urology. 2015;85:e39–40.

21. Virchow RLK. Gesammelte Abhandlungen zur Wissenschaftlichen Medicin. Frankfurt: Meidinger Sohn & Co; 1856.

22. Kayes O, Patrick N, Sengupta A. A peculiar case of bilateral, spontaneous thromboses of the pampiniform plexi. Ann R Coll Surg Engl. 2010;92:W22–3.

23. Rudloff U, Holmes RJ, Faust GR, Moldwin R, Siegel D. Mesoaortic compression of the left renal vein (nutcracker syndrome): case reports and review of the literature. Ann Vasc Surg. 2006;20(1):120–9. https://doi.org/10.1016/j.avsg.2005.07.011.

24. Kayes O, Patrick N, Sengupta A. A peculiar case of bilateral, spontaneous thrombosis of the pampiniform plexi. Ann R Coll Surg Engl. 2010;92:22–3.

25. Mallat F, Hmida W, Ahmed KB, Mestiri S, Mosbah F. Spontaneous spermatic vein thrombosis as a circumstance of discovery of the nutcracker syndrome: an exceptional entity. Int J Case Rep Images. 2014;5(7):519–23.

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