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Case report

Bilateral spontaneous thrombosis of the pampiniform plexus; A rare etiology of acute scrotal pain: A case report and review of literature

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Abstract
Introduction: Acute testicular pain is frequent in urology. If torsion of the spermatic cord and orchiepididymites are usual, thrombosis of the pampiniform plexus is a very uncommon clinical entity. We present an unusual case and review the literature to explore potential etiologies and therapeutic strategies.
Observation: A rare case of bilateral thrombosis of the pampiniform plexus occurred in a 39 year-old male.The diagnosis was confirmed with doppler sonographie and Computer Tomography. Urethral infection and protein C deficiency were found as associated factors. The treatment was conservative with good result.
Conclusions: Anatomical factors are probably responsible for almost exclusive involvement of the left side. However, coagulation abnormalities and retroperitoneal tumors or absence of inferior vena cava must be sought especially in cases of right side or bilateral thrombosis of the pampiniform plexus. The management

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of pampiniform plexus thrombosis remains non surgical based on symptomatic treatment with good clinical and radiological evolution.

Introduction

Acute painful scrotum is the most common urologic emergency and may present a diagnostic challenge even the most experienced clinicians. It may be due to twisted spermatic cord, twisted testicular appendages or epididymitis. Most rarely it occurs as a result of a testicular trauma, orchitis, idiopathic scrotal edema, idiopathic infarction of testis and vaginalis tunica or testicular neoplasm. Rarely, acute painful scrotum may be associated with thrombosis of the internal spermatic vein or pampiniform plexus veins [1]. Varicocele thrombosis can affect children and adult. Mc Gavin [2] is credited with the first complete description in 1935. Only 20 cases of thrombosis of Varicocele were reported in the literature. We present a peculiar case of bilateral, spontaneous thrombosis of the pampiniform plexus and review the literature to explore potential etiologies and therapeutic strategies.

Case report

A 39 year-old male was evaluated in the emergency department 2 days after worsening, unprecipated scrotal pain and swelling. He had a past medical history of psoriasis and recurrent oral aphthosis. Examination showed large, firm, erythematous, and exquisitely tender scrotum, with the left hemiscrotum larger than the right, which remained unchanged when standing, supine, or with valsalva maneuver. The cord was swollen, tense and painful on palpation along the inguinal route. He had cutaneous lesions of psoriasis in the knee.

Scrotal ultrasound showed a left varicocele grade II partially thrombosed without focal testicular lesion (Fig. 1).

Figure 2 Computed tomography scan with reconstruction showing thrombosis of the right spermatic vein (arrow).

The patient underwent analgesic and preventive dose of enoxaparine (low-molecular – weight heparine) with clinical improvement. After six days of treatment he consulted again with the same symptoms (in the right scrotum) and purulent urethral discharge.

The Scrotal ultrasound showed an acute thrombus in the right gonadal vein. The CT scan (Fig. 2) showed thrombosis of the right spermatic vein into stoma of the vena cava with a heterogeneous aspect of the right gland seminal.

A treatment based on ofloxacin, analgesic and a curative dose of enoxaparine (LMWH) were initiated for 21 days with good clinical and radiological evolution.

Our patient underwent a coagulation check-up: protein S/C dosage, antithrombin III and resistance to activated protein c, homocysteine dosage, anti-cardiolipin b2 GPI.

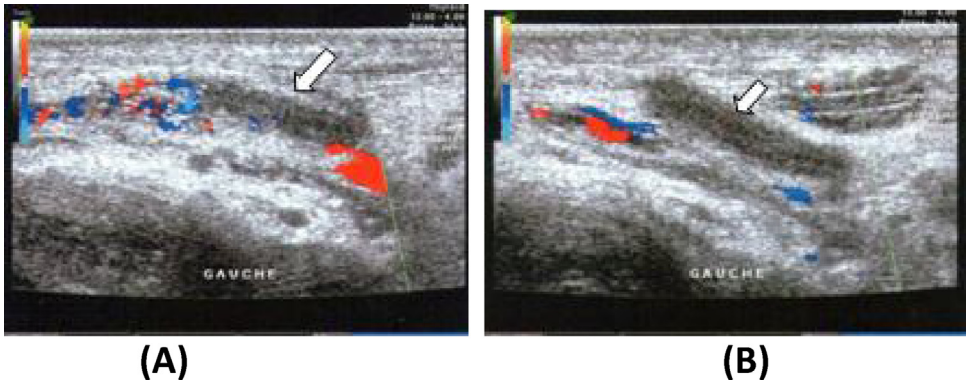

Figure 1 Sagittal color Doppler sonography of the left spermatic cord demonstrating echogenic intraluminal thrombus in a dilated vein pampiniform plexus (arrow) (A) and non depressible (arrow) (B).

Table 1 Reported cases of spontaneous thrombosis of the pampiniform plexus.

Case N°	Reference	Age (years)	Involved side	Duration of symptoms	Associated factors	Treatment	Diagnostic tests	Preoperative diagnosis
1	13	NA	Left	NA	NA	NA	None	Orchitis
2	14	NA	NA	"Sudden"	None	Excision	None	Thrombosis
3	2	41	Left	5 weeks	None	Orchidectomy	None	Orchitis
4	2	57	Left	4 weeks	None	Orchidectomy	None	Orchitis
5	15	27	Left	16 h	None	vein biopsy	None	NA
6	16	7	Left	NA	None	Exploration	venography	NA
7	16	10	Left	NA	None	Anti-inflammtories	venography	Thrombosis
8	16	15	Left	11 days	walking	Excision	None	NA
9	3	44	Right	"Hours"	Playing sports	Excision	None	Inguinal mass
10	17	33	Left	10 days	None	Excision	IVP	Incarcerated hernia
11	12	33	Left	NA	Varicocele	Excision	None	NA
12	12	42	Cotralateral	1 week	None	Excision	IVP, Cavogram CT Scan	Incarcerated hernia
13	8	19	Left	"Hours"	Vigorous exercise	Excision	None	IH
14	1	23	Left	"Hours"	Heavy lifting	Excision	Doppler	Incarcerated hernia
15	9	27	Left	2–3 h	Heavy lifting	Exploration	None	Incarcerated hernia
16	18	33	Left	3 days	Cycling	Anti-inflammtries	Doppler	Thrombosis
17	19	NA	Cotralateral	NA	NA	NA	NA	NA
18	4	43	Right	2 days	Absence IVC+ Mutation F.V.Leiden	anticoagulation	Doppler + CT	NA
19	5	28	Left	14 days	Nutcracker synd	Excision	Doppler + CT	NA
20	Present Report	39	Cotralateral	3days	Infection Protein C deficiency	Atb + anticoagulant	Doppler CT Scan	Thrombosis

IVP: i.v pyelograpy, NA: information not available, CT: computed tomography, IH: incarcerated hernia, Atb: Antibiotherapy, IVC: inferior vena cava.

Bechet's disease was investigated because of the patient oral aphthosis. We performed:ophthalmic examination, pathergy test and HLA B51. These explorations were negative. We realised a complete blood count, and ferritin rate, to investigate a chronic intestinal inflammatory disease. Exploration has concluded a protein C deficiency and the patient remained therapeutic anticoagulation. The patient underwent treatment with Acenocoumarol.

The patient's symptoms resolved after 2 weeks. Ultrasonography at six months did not show thrombosis and varicocele. At 2-years follow-up, patient urinary tract symtoms and erectile function remain unchanged compared baseline.

Discussion

Spontaneous thrombosis of the pampiniform plexus is such an unusual condition that only 20 cases (including ours), have been reported in the literature (Table 1). The median age was 33 years ±13. Three broad categories of factors, known as Virchow's triad, contribute to thrombosis: blood stasis, coagulation factors and mural factors.

All reported cases, involved the left side, (as well as) except two (case N°9 [3] and N°18 [4] in Table 1). The etiology of **this condition remains unclear**. **In fact, there are well-known anatomical factor explaining the preponderance of varicocele in the left side**: compression of the left renal vein by superior mesenterique artery « nutcracker phenomenon » [17,5] and absent or incompetent valves in the left spermatic vein [15,6] that could also explain the same preponderance of the thrombosis.

Anatomical factors which are incriminated in the preponderance of varicocele on the left side can be responsible in the occurrence of the left pampiniform plexus thrombosis. These anatomical factors are: firstly the perpendicular disposition of the left spermatic vein when it joins the left renal one; contrary the right spermatic vein which enters the inferior vena cava obliquely. Secondly, the left renal vein enters the vena cava 8–10 cm above the right spermatic vein with greater column pressure in the left spermatic vein and as a consequence a reduction in blood flow [7,14].

Furthermore, in three reported cases (case N°13 [13–15,1,8,9]) an increased of intra-abdominal pressure due to heavy lifting or exercise are found, which can decrease the spermatic vein blood flow, creating stasis and thrombosis

The incidence of inferior vena cava anomalies reaches 5% in patients under the age of thirteen suffering from deep venous thrombosis (DVT). In some cases, concomitant clotting defects was reported, parting to indefinite anticoagulation therapy [4,16]. Among veina cava anomalies, the absence has a frequency of 0.2–1% [10,18]. This later associated to increased lower extremity venous pressure leads to DVT formation [11,19].

One other possible etiologic factor was ruled out in our patient. Retroperitoneal tumours can produce venous blood flow obstruction and thrombosis; however, the patient had a normal CT scan.

In our patient the presence of a hypercoagulable status, associated with infection (urethritis) would be risk factor for thrombosis. Our patient indeed is the first report of this kind of etiology.

Preoperative diagnosis of this condition is un-common. Among the 20 reported cases, only four were diagnosed preoperatively, both based on clinical examination alone. Complicated inguinal hernia was the most common. Despite ultrasound examination, a surgical exploration for strangulated hernia was indicated on the basis of the finding of mass without blood flow. In our opinion the treatment of pampiniform plexus thrombosis should be conservative. Infact, there is no need to excise the blood thrombosed plexus, as evidence of the good results in our case and at least three other cases. There has been only one case report of testicular infarction following pampiniform plexus thrombosis (Table 1, case N°12 [12,9]). This same patient had developed additionally thrombosis of the contralateral pampiniform plexus. Our patient is the third reported case of bilateral thrombosis of the pampiniform plexus.

Conclusions

Thrombosis of the pampiniform plexus is an uncommon clinical entity. Anatomical factors are probably responsible for almost exclusive involvement of the left side. However, coagulation abnormalities and retroperitoneal tumors or absence of inferior vena cava must be sought especially in cases of right side or bilateral thrombosis of the pampiniform plexus.

It should be suspected when we have swollen, tense and painful cord on palpation along the inguinal route. The clinician must roll out differential diagnosis such as a complicated inguinal hernia. The diagnosis would be confirmed by the Doppler sonography. It allows clinician to differentiate cases of warranting surgery from those who would benefit from conservative management alone in patients with acute scrotal pain

The management of pampiniform plexus thrombosis remains non surgical based on symptomatic treatment with good clinical and radiological evolution.

Conflict of interests

None.

Source of funding

None.

Consent from the patient

The consent of the patients was demand and approved.

Authors' contributions

Ktari Kamel, Sarhen Gassen, Mahjoub Mohamed, Ben kalifa Bader, Klaii Rim, Jelled Anis, Saidi Radhia, Saad Hamadi: all participated in collecting data and script writing. Bouslam Karim: major role in medical imaging analysis.

References

[1] Gleason TP, Balsara Z, Goff WB. Sonographic appearance of left spermatic vein thrombosis simulating incarcerated inguinal hernia. J Urol 1993;150:1513–4.

[2] Mc Gavin D. Thrombosis of the pampiniform plexus. Lancet 1935:368–9.
[3] Rothman D. Thrombosis of the pampiniform plexus. J Med Soc New Jersey 1981;78:681.
[4] Chi AC, Hairston JC. Acute right varicocele: a clue to congenital vascular anomaly. Urology 2015;85:e39–40.
[5] 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.
[6] Coolsaet BLRA. The varicocele syndrome: venography determining the optimal level for surgical management. J Urol 1980;124:833–9.
[7] Shneck FX, Bellinger MF. Abnormalities of the testes and scrotum and their surgical management. In: Walash PC, Retik AB, Vaughan ED, Wein AJ, editors. Campbell's urology. 8th ed. Philadelphia, PA: Saunders; 2002. p. 2353–94.
[8] Isenberg JS, Ozuner G, Worth MH, Ferzli G. Effort-induced spontaneous thrombosis of the left spermatic vein presenting clinically as a left inguinal hernia. J Urol 1990;144:138.
[9] Hashimoto L. Brett Vibeto spontaneous thrombosis of the pampiniform plexus. Scand J Urol Nephrol 2006;40:252–4.
[10] chee YL, Culligan DJ, Wtson HG. Inferieur vena cava malformation as a risk factor for deep venous thrombosis in the young. Br J Haematol 2001;114:878–80.
[11] Gil RJ, Perez AM, Arias JB, et al. Agenesis of the inferieur vena cava associated with lower extremites and pelvic venous thrombosis. J Vasc Surg 2006;44:1114–6.
[12] Roach R, Messing E, Starling J. Spontaneous thrombosis of left spermatic vein: report of 2 cases. J Urol 1985;134:369–70.
[13] Senn NA. Surgical clinic. Clin Rev 1903;4:241–5.
[14] Senn NA. Vesical calculus; thrombosis of the spermatic veins; cervical tuberculous lymphadenitis; sarcoma of the submaxillary gland; syndactylus; traction injury of the peroneal nerve; paralysis of the circumflex nerve; rachitis; acute osteomyelitis of the os calcis; adenomatous goiter; gaglion. Int Clin 1904;4:148–60.
[15] Anseline P. A case of spontaneous thrombosis of the testis. Aust N Z J Surg 1977;47:801–2.
[16] Coolsaet B, Weinberg R. Thrombosis of the spermatic vein in children. J Urol 1980;17:175–6.
[17] Vincet MP, Bokinsky G. Spontaneous thrombosis of pampiniform plexus. Urology 1981;17:175–6.
[18] Doerfler A, Ramadani D, Meuwly JY, et al. Varicocele thrombosis: a rare etiologie of testicular pain. Prog Urol 2009;19:351–2.
[19] Kayes O, Patrick N, Sengupta A. A pecular case of bilateral, spontaneous thrombosis of the panpiniform plex. Ann R Coll Surg Engl 2010;92(October (7)):W22–3.