

## Review

# Urological Manifestations of the Systemic Vasculitides—A Scoping Review

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**Abstract:** Background: Vasculitides are specific inflammations of the blood vessel wall that can take place in any organ system of the human body. They may occur as a primary process (primary systemic vasculitides, PSV) or may be secondary to another underlying disease. In general, in association with the specific type of vasculitis, affected vessels vary in size, type, and location. In the following scoping review, we present clinical characteristics and manifestations of PSV with reference to the genitourinary system. Materials and methods: A non-systematic search of the relevant literature was conducted using three electronic databases (PubMed, Embase, and Web of Science) up to 29 October 2021. Results: Urogenital manifestations of PSV are infrequent, with the most commonly reported findings as prostatic or testicular involvements. However, almost all other organs of the genitourinary system can be affected. Conclusions: Because of the clinical heterogeneity and non-specific symptoms, the proper diagnosis of PSV is often delayed and constricted. Fast identification of urological manifestations of vasculitides is essential in implementing appropriate therapy and avoiding unnecessary, harmful, and invasive surgery.

**Keywords:** vasculitis; urology; orchitis; epididymitis; prostatitis; ureteral stenosis



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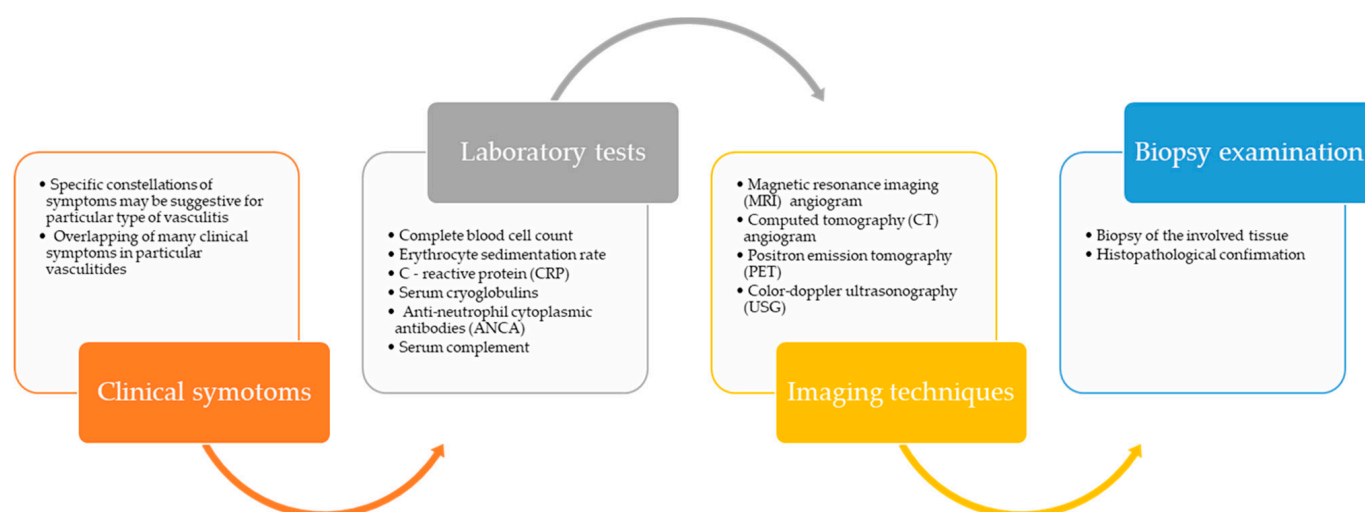
## 1. Introduction

Vasculitides are specific inflammations of the blood vessel wall that can take place in any organ system of the human body. They may occur as a primary process (primary systemic vasculitides, PSV) or may be secondary to another underlying disease. The pathogenesis of PSV is poorly understood. Three of the most common mechanisms of vascular damage are immune complexes, anti-neutrophil cytoplasmic antibodies (ANCA), and cell-mediated T-lymphocyte response [1]. In association with the specific type of PSV, affected vessels vary in size, type, and location. The most commonly used nomenclature system, including the names and definitions for most forms of vasculitides, was developed by the international Chapel Hill Consensus Conference (CHCC) in 1994 and was reviewed in 2012 [2,3].

Because of the clinical heterogeneity and non-specific symptoms, the proper diagnosis of PSV is often delayed and constricted. The presence of the PSV may be mimicked by a vast number of other diseases, such as infections or neoplasms [4]. Hence, differential diagnosis requires careful assessment of all available clinical, laboratory, radiologic, and pathologic examinations (Figure 1) [5,6].

In the initial treatment of most types of PSV, glucocorticoids remain the basis, followed by the second line immunosuppressive drugs and biological agents [4]. Endovascular procedures, such as balloon angioplasty and stents or graft placement, constitute the main

forms of surgical treatment. The detailed review of all available forms of PSV treatment exceeds the scope of this review, and it was not performed.

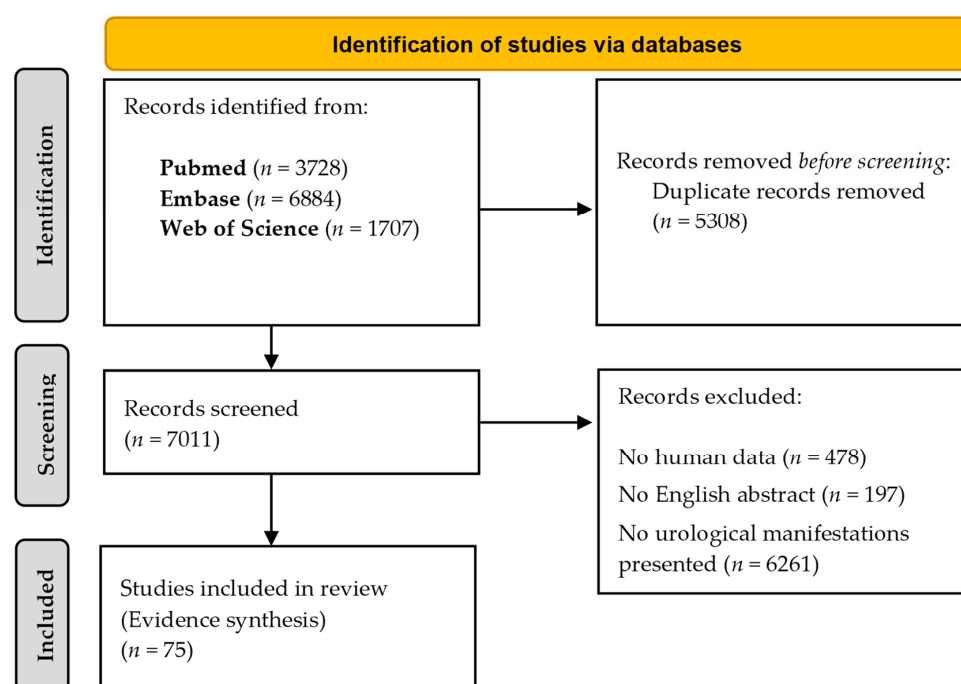


**Figure 1.** Basic methods used in diagnostic evaluation of primary systemic vasculitides.

To date, a single review article regarding urological manifestations of PSV was published in 2015 [7]. Therefore, we decided to perform an updated review including recently published data. We aimed to comprehensively present clinical characteristics and manifestations of PSV with reference to urinary system involvements.

## 2. Evidence Acquisition

A non-systematic search of the relevant literature was conducted using three electronic databases (PubMed, Embase, and Web of Science) up to 29 October 2021. The combination of key words used for search strategy included: ("Takayasu arteritis" OR "Giant cell arteritis" OR "Polyarteritis nodosa" OR "Kawasaki disease" OR "Granulomatosis with polyangiitis" OR "Microscopic polyangiitis" OR "Eosinophilic granulomatosis with polyangiitis" OR "Immunoglobulin A vasculitis" OR "Anti-GBM disease" OR "cryoglobulinaemic vasculitis" OR "Hypocomplementaemic urticarial Vasculitis" OR "Behçet's disease") AND ("urinary system" OR "genitourinary system" OR "urology" OR "kidney" OR "ureter" OR "bladder" OR "prostate" OR "urethra" OR "testicle" OR "penis"). Alerts in Medline were also run, as well as reference lists of original and review articles for further eligible data. Only publications with abstracts written in English were considered, and evidence was limited to human data. The vast majority of analyzed articles were single case presentations or case series studies. The flow diagram of the study selection process is presented in Figure 2.



**Figure 2.** Flow diagram of study selection process.

### 3. Evidence Synthesis

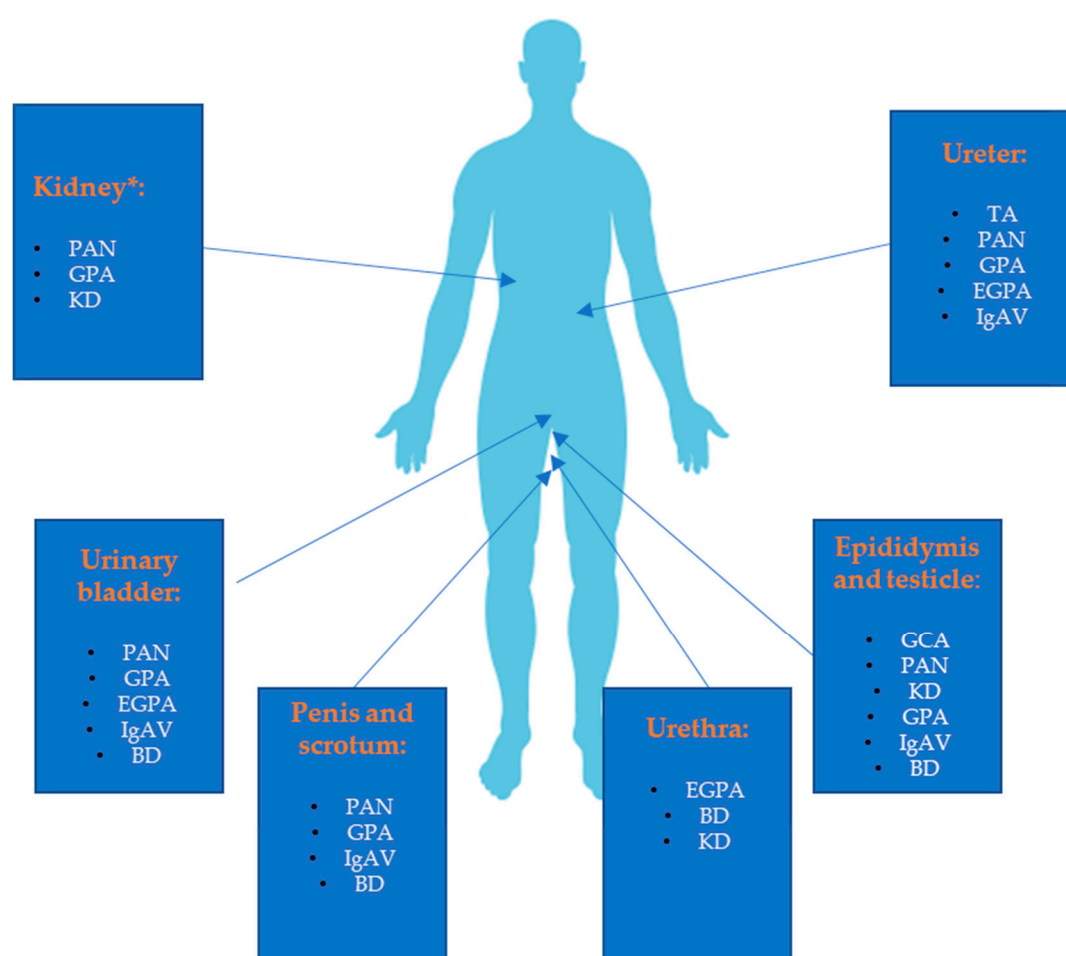
In Table 1, we summarize the main clinical symptoms of particular PSV and their possible urological manifestations. The types of PSV involving the specific parts of the urinary system are presented in Figure 3.

**Table 1.** Main clinical symptoms of particular primary systemic vasculitides and their possible urological manifestations.

Type	Main Symptoms	Urological Manifestations
<b>Large Vessel Vasculitis</b>		
Takayasu arteritis (TA)	Arthralgia, myalgia, carotidynia, absent or weak peripheral pulse, limb claudication, arterial bruit, discrepant blood pressure between arms, hypertension, angina, gastrointestinal symptoms (abdominal pain, diarrhea, gastrointestinal hemorrhage) skin lesions (erythema nodosum, pyoderma gangrenosum), respiratory symptoms (chest pain, dyspnea, hemoptysis, pulmonary hypertension), neurologic symptoms (light headedness, vertigo, syncope, orthostasis, headaches, convulsions, and strokes), visual impairment	Obstructive nephropathy
Giant cell arteritis (GCA)	Jaw claudication, ocular symptoms (transient visual loss, permanent vision loss, diplopia, Charles Bonnet syndrome), musculoskeletal symptoms (proximal polyarthralgias and myalgias, peripheral synovitis, distal extremity swelling with pitting edema), large vessel involvement (aneurysms and dissections of the aorta, stenosis, occlusion, and ectasia of large arteries), strokes	Epididymitis, orchitis, testicular masses
<b>Medium Vessel Vasculitis</b>		
Polyarteritis nodosa (PAN)	Skin lesions (tender erythematous nodules, purpura, livedo reticularis, ulcers, bullous or vesicular eruption), neurologic symptoms (mononeuropathy multiplex or asymmetric polyneuropathy affecting, e.g., radial, ulnar, peroneal nerves), gastrointestinal symptoms (abdominal pain, nausea, vomiting, melena, bloody or non-bloody diarrhea, life-threatening gastrointestinal bleeding), coronary artery disease (myocardial ischemia, heart failure, cardiomyopathy), myalgia and muscle weakness, ocular symptoms (ischemic retinopathy with hemorrhages, retinal detachment, ischemic optic neuropathy), glomerulonephritis	Orchitis, scrotal necrosis, bladder and testicular masses, ureteric stricture, spontaneous ureteric rupture, renal artery aneurysms, perirenal hematomas, spontaneous retroperitoneal hemorrhage

Table 1. Cont.

Type	Main Symptoms	Urological Manifestations
Kawasaki disease (KD)	Conjunctivitis, mucositis, skin lesions (rash, perineal erythema, redness or crust formation at the site of Bacille Calmette–Guérin inoculation), extremity changes (indurated edema of the dorsum of their hands and feet, diffuse erythema of their palms and soles), arthritis, lymphadenopathy (cervical lymphadenopathy), cardiovascular involvement (coronary arteries inflammation, formation of microaneurysms, myocarditis, pericarditis, valvulitis)	Sterile pyuria, hydrocele testis
<b>Small Vessel Vasculitis</b>		
Anti-neutrophil cytoplasmic antibody (ANCA) associated vasculitis		
Microscopic polyangiitis (MPA)	Elevated purpura, alveolar hemorrhage, glomerulonephritis	Prostatitis
Granulomatosis with polyangiitis (GPA)	Upper airways, eye and ear disease (conductive hearing loss, sensorineural hearing loss, strawberry gingivitis, underlying bone destruction with loosening of teeth, non-specific ulcerations throughout the lining of the mouth and nasal septum), renal involvement (rapidly progressive glomerulonephritis), subglottal stenosis, lower respiratory tract involvement (pulmonary nodules, cavitary lesions, bleeding in the lungs, bronchial stenosis), arthritis, skin lesions (subcutaneous nodule on the elbow, purpura), neurologic symptoms: (sensory neuropathy, mononeuritis multiplex)	Prostatitis, epididymitis, orchitis, ureteric stricture, penile ulceration, necrotizing urethritis, large renal and bladder granulomas, vesicovaginal fistula
Eosinophilic granulomatosis with polyangiitis (EGPA)	Respiratory symptoms (asthma, allergic rhinitis, nasal polyps, pulmonary bleeding), neurologic symptoms (mononeuropathy multiplex, symmetrical polyneuropathy), renal involvement (glomerulonephritis, hypertension), cardiovascular involvement (myocarditis, pericarditis), skin lesions (elevated purpura, subcutaneous nodules, livedo reticularis), gastrointestinal symptoms (eosinophilic gastroenteritis, diarrhea, gastrointestinal bleeding, colitis)	Prostatitis, cystitis, ureteric stricture, vesicovaginal fistula
Immune complex small vessel vasculitides		
IgA vasculitis (IgAV)	Skin lesions (symmetrical erythematous petechial or purpuric rash that almost exclusively starts on the lower limbs and buttocks), arthritis, gastrointestinal symptoms (nausea, vomiting, colicky abdominal pain, transient paralytic ileus, gastrointestinal hemorrhage, bowel ischemia and necrosis, intussusception, bowel perforation), glomerulonephritis	Epididymitis, orchitis, ureteric stricture, cystitis, priapism, spermatic vein thrombosis
<b>Variable Vessel Vasculitis</b>		
Behcet's disease (BD)	Oral ulcerations, skin lesions (acneiform lesions, papulo-vesiculo-pustular eruptions, pseudofolliculitis, nodules, erythema, palpable purpura), ocular disease (uveitis, secondary cataracta, glaucoma, macular edema), neurologic symptoms, arterial disease (hemorrhage, stenosis, aneurysm formation), venous thrombosis, cardiac disease, gastrointestinal symptoms (abdominal pain, diarrhea, gastrointestinal ulcerations, bleeding)	Genital ulceration, epididymitis, sterile urethritis, bladder wall ulceration, cystitis, vesicovaginal fistula, urethrovaginal fistula, priapism



**Figure 3.** The types of primary systemic vasculitides involving the specific parts of the urinary system. TA—Takayasu arteritis; GCA—giant cell arteritis; PAN—polyarteritis nodosa; KD—Kawasaki disease; GPA—granulomatosis with polyangiitis; EGPA—eosinophilic granulomatosis with polyangiitis; IgAV—IgA vasculitis; BD—Behcet’s disease; \*—involvement other than glomerulonephritis.

### 3.1. Large Vessel Vasculitides

#### 3.1.1. Takayasu Arteritis (TA)

Jghaimi et al. presented a case report of a patient with suspected retroperitoneal fibrosis (RPF) and obstructive nephropathy requiring placement of double-J catheters. The diagnosis of TA was confirmed three years later when the patient presented with right upper extremity dysesthesia [8]. Aside from other cases of TA concomitant to RPF causing obstructive nephropathy, no more specific urological manifestations of TA have been reported in the literature [9].

#### 3.1.2. Giant Cell Arteritis (GCA)

Urological manifestations of GCA seem to be extremely rare. Epididymal involvement has been reported in a 66-year-old male with a 3-month history of generalized fatigue and right-sided epididymal tenderness. Epididymal biopsy demonstrated chronic inflammatory cell infiltration and the presence of “giant cells”. The patient subsequently underwent a temporal artery biopsy, which confirmed the diagnosis of GCA [10]. Testicular involvement has been reported in a 76-year-old male with weight loss, malaise, and right testicular swelling, suspected of malignant process on ultrasonography (USG). The patient underwent radical orchidectomy, and GCA was confirmed in histopathological assessment [11]. In the existing literature, the association between GCA as a paraneoplastic condition and the development of prostate cancer (PCa) was also hypothesized [12].

### 3.2. Medium Vessel Vasculitides

#### 3.2.1. Polyarteritis Nodosa (PAN)

Testicular involvement is the most characteristic urological manifestation of PAN. It is reported in 38–86% of patients based on autopsy studies, but only 2–18% of patients are symptomatic [13]. Clinically, the testicular involvement is usually characterized by pain, swelling, or erythema. Orchitis is predominantly unilateral and concomitant with systemic disease, whereas bilateral disease is extremely scarce [14]. Isolated testicular involvement, as the initial site of PAN, is rare, with only 39 cases presented in the literature up to date [15]. A heterogeneous clinical course of testicular involvement in PAN often leads to a misdiagnosis. Several authors have reported testicular masses suspicious for malignant tumors that have proven to be PAN [16,17]. In these cases, radical orchidectomies were performed, and subsequent pathological examinations revealed the presence of histological features consistent with a diagnosis of PAN. Clinical symptoms of testicular involvement in PAN can also mimic a testicular torsion [18]. In isolated testicular vasculitis in PAN, orchiectomy is considered the main therapeutic approach, with a low risk of developing systemic PAN requiring pharmacotherapy after surgery [17].

There is a single case report of a 54-year-old man with PAN affecting both the bladder and testis in the setting of cryptorchidism, without any signs of systemic disease. The patient presented a several-month history of urinary retention and intermittent gross hematuria. Cystoscopy revealed a superficial-appearing mass at the dome of his bladder. A radical right orchiectomy and transurethral resection of the bladder tumor (TURB) were performed. Histopathological examination revealed the presence of PAN vasculitis in both urinary bladder and undescended testis [15].

Renal involvement in PAN comprises tissue infarction or hematoma, typically produced from the rupture of renal microaneurysms. Kidney infarcts might be clinically silent or manifest as hematuria or proteinuria [19]. Patients may rarely be complicated by aneurismal rupture leading to subcapsular and perirenal hematomas [20,21]. Spontaneous retroperitoneal hemorrhage (also known as Wunderlich syndrome, WS) is a potentially life-threatening entity, scarcely reported in the literature [22,23]. It is classically characterized by acute flank pain, abdominal tenderness, and signs of massive internal bleeding. Aneurysms can develop due to segmental necrosis, which correlates with the severity of the illness and may cause the development of thrombosis, ruptures, and massive hemorrhage [24]. In most cases, an urgent nephrectomy is a life-saving option, but in selected cases, WS can be treated conservatively [22]. Therapeutic arterial embolization could be an alternative to surgery in patients with hemorrhage from a ruptured aneurysm, especially those in critical condition [25].

Another single-reported, rare urological manifestations of PAN include ulcerative necrosis of the glans penis and scrotum, imprints on the ureteral wall, nodular appearance of the ureter, and spontaneous ureteral rupture (SUP) [26–28]. In the majority of these cases, resolution of the stricture could be achieved with systemic therapy for PAN, but surgical intervention such as resection of the stenosed ureteric segment or necrotic tissue might become necessary.

#### 3.2.2. Kawasaki Disease (KD)

There are very scarce reports of KD affecting the urinary system. Sterile pyuria is the most common abnormal finding, and some patients are misdiagnosed as having acute pyelonephritis. Sterile pyuria in KD is thought to be due to urethritis caused by a non-specific vasculitis of the urethra [29]. Kidney involvement also includes prerenal acute kidney injury (AKI), renal AKI caused by tubulointerstitial nephritis, hemolytic-uremic syndrome (HUS), immune-complex mediated nephropathy, or renal AKI associated with Kawasaki Disease Shock Syndrome (KDSS) [30]. The formation of renal artery pseudoaneurysms is an extremely rare complication of KD with an estimated 0.8% incidence rate [31].



There is a case report of a 13-year-old girl with a past medical history significant for KD and right ovarian torsion presented to the emergency department. In the physical examination, patient revealed a soft, non-tender abdomen and improvement of pain with right flank pressure. Urinalysis was positive for microhematuria, and subsequently performed computed tomography (CT) revealed right inferior pole intraparenchymal hemorrhagic mass, concerning for renal aneurysm. The patient underwent renal arteriography, which showed a 2 cm pseudoaneurysm. After confirmation of the diagnosis, embolization was performed with the good clinical outcome with preserved renal function [32]. Another urological manifestation of KD could be hydrocele testis, as described by Jibiki et al. [33].

### 3.3. Small Vessel Vasculitides

#### 3.3.1. ANCA-associated Vasculitides

##### Granulomatosis with Polyangiitis (GPA)

Urogenital involvement in GPA is observed in approximately 1–10% of patients [34]. Prostatitis is reported in about 12–37% of patients with urogenital involvement of GPA. GPA can involve the prostate gland causing acute obstruction with the potential to recur rapidly after surgical treatment [35]. Testicular vasculitis is observed in up to 36% of patients [34]. Scrotal hyperemia, necrotic ulcers, inflammatory masses, infarction, and necrosis have also been reported [36,37]. Testicular involvement in GPA is rather concomitant with a systemic disease, although limited disease has also been reported in the literature [38]. Epididymitis as the initial manifestation of GPA was described by several authors [36,39].

Bladder involvement is another possible urological manifestation of GPA. Patients typically present with urgency and dysuria, whereas urinary incontinence and obstructive signs leading to ureter dilatation and hydronephrosis are scarcely reported [40,41]. Cystoscopy usually reveals a diffusely thickened bladder with ulcerations and fibrosis, while CT images may show wall thickening or polyps. Some patients present micro- or macrohematuria. Vesicovaginal fistula is one of the rarest manifestations of GPA [42].

Ureteral stenosis due to retroperitoneal inflammation or segmental thickening of surrounding vessels can also be associated with GPA [43]. However, it is a rare manifestation as only 11 cases have been reported in the literature to date [44]. Anuria and acute renal failure may develop in patients with bilateral stenosis [45,46].

Ulcerations of the penis have been reported in a few cases. Ulcers are usually painless, recurrent, and sometimes accompanied by local edema and regional lymphadenopathy, simulating a neoplasm. There are also case reports describing aggressive necrotizing urethritis in both male and female elderly patients. This form of destructive urethritis may mimic invasive urethral carcinoma [47–49].

Renal granulomatous pseudotumors, which are fibro-inflammatory masses, could be present in GPA, mimicking kidney cancer. They are usually asymptomatic and discovered incidentally on imaging studies [50].

Sometimes surgical intervention may become necessary in patients with GPA. Placing ureteral catheters as a temporary measure relieves obstruction symptoms in ureteral stenosis. In the case of urethral stricture, dilation may be required. Some patients undergo surgical procedures (orchiectomy, prostatectomy, and nephrectomy) as part of the diagnostic protocol for suspected malignant neoplasms [36].

##### Microscopic Polyangiitis (MPA)

Renal involvement, characterized by rapidly progressive glomerulonephritis (RPGN), is the major clinical feature of MPA with the occurrence of 80–100%. Renal manifestations can range from asymptomatic urinary sediment to end-stage renal disease requiring dialysis. Consistent with glomerulonephritis, the most common clinical symptoms of renal involvement in MPA are proteinuria, microscopic hematuria, and urinary granular or red blood cell casts [51]. Lower urogenital tract involvement in MPA is virtually not reported in the literature, except for two isolated case reports of prostatitis [52,53].

### Eosinophilic Granulomatosis with Polyangiitis (EGPA)

There is a single case report describing urinary bladder involvement in association with EGPA. A 76-year-old male with a medical history of seasonal asthma, symptoms of sensory polyneuropathy in both feet and calves, cutaneous lesions on his arms and legs consistent with leukocytoclastic vasculitis, and chronic diarrhea due to ileocecal resection underwent radical cystectomy in view of high-grade cancer and severe bladder symptoms. Histopathological examination after cystectomy revealed an urothelial carcinoma, as well as intense tissue eosinophilia besides the tumor. A review of the patient's complete medical history and the available pathology samples led to the diagnosis of EGPA [54].

Famokunwa et al. presented a case 55-year-old woman with a vesicovaginal fistula as a manifestation of EGPA. An attempt at surgical closure of the fistula failed, and a flare of the EGPA subsequent to surgical intervention resulted in rapid deterioration of her condition with acute renal failure and sepsis. Administration of methylprednisolone resulted in improvement in her pain and normalization of CRP, allowing sufficient conditions in which an ileal conduit diversion was possible [55].

Prostatic involvement is also reported in EGPA. In one case, a 74-year-old male, who had previously been treated for asthma, underwent transurethral resection of the prostate. Post-operatively he developed pyrexia and eosinophilia. The biopsy specimens showed eosinophilic prostatitis in keeping with EGPA. The patient's symptoms responded to oral prednisolone [56]. In the second case, a 60-year-old man presented with poor urinary flow, hesitancy, and nocturia underwent TURP. Histology showed fibromuscular hyperplasia and florid eosinophilic prostatitis. Two weeks after discharge, he was admitted as an emergency with a rash affecting both ankles and legs. The clinical and laboratory findings suggested EGPA with cutaneous, prostatic, and possibly renal involvement [57].

Urological manifestation of EGPA may also concern ureteric obstruction secondary to calcification and stenosis [58,59], bilateral ureteral stenosis with anuria [60], ulceration of the mid-penile urethra [61], and funiculitis [62], but only a few case reports are available in the literature.

### 3.3.2. Small Vessels Vasculitides Associated with Immune Complex Deposition Immunoglobulin A Vasculitis (IgAV)

Involvement of urinary tract in IgAV was reported in several cases, including scrotum, testicle, ureter, bladder, prostate, and penis. Common testicular manifestations of IgAV include acute scrotum, epididymitis, orchitis, and spermatic cord complications [63]. It may require assessment by an experienced pediatric surgeon to exclude a testicular torsion, which requires surgical emergency instead of conservative management. Orchitis is usually gradual in nature with associated nausea and vomiting, whereas scrotal involvement due to IgAV presents with instantaneous pain, typically without nausea or vomiting [64]. Hematomas and edema of the spermatic cord are rare urological complications of IgAV and they could be also confused with testicular torsion and acute scrotum. Some patients with spermatic cord involvement suffer from thrombosis of the spermatic veins; however, it is not typically seen in children [65]. Penile manifestations of IgAV include thrombosis, priapism, and purpuric lesions on the penis. Such lesions may appear before or after the onset of IgAV [66]. Ureteral involvement often results in ureter obstruction. Obstruction is either unilateral or bilateral and may be partial or complete depending on the severity of the case [67]. Residual stenotic lesions and subsequent urinary tract obstruction frequently require surgical intervention [68]. Bilateral and unilateral ureteritis are other complications reported in IgAV, and they appear about 1–2 months after the onset of the disease [69]. In some cases, ureteritis is accompanied by nephritis, which results in delayed diagnosis and an increased risk of complications [70].

IgAV could also affect the urinary bladder. Hirayama et al. described an 83-year-old man who developed IgAV in the course of *Bacillus Calmette–Guerin* (BCG) intravesical therapy. Following the BCG maintenance, purpura appeared on the lower region of the patient's legs [71]. Ishigaki et al. described a 70-year-old male with bladder cancer



diagnosed with IgAV following neoadjuvant chemotherapy and a radical cystectomy. Twenty-three days after radical cystectomy, the patient presented with purpura, and the diagnosis of IgAV was made based on a skin biopsy [72].

In adults, IgAV has been linked to PCa. The relationship between the development of IgAV and PCa remains unknown, but it is postulated that tumor antigens or irregular IgA production may be contributing factors [73].

#### Other Small Vessels Vasculitides Associated with Immune Complex Deposition

Other small vessel vasculitides associated with immune complex deposition are anti-glomerular basement membrane antibody (Anti-GBM disease, former Goodpasture's Syndrome), cryoglobulinemic vasculitis, and Hypocomplementemic urticarial Vasculitis (HUV, anti-C1q vasculitis). Urinary system involvement in these diseases is reported exceptionally. No lower urinary tract manifestations of the anti-GBM disease have been presented in the literature. Glomerulonephritis with non-visible hematuria appears to be the only reported urological manifestation of cryoglobulinemic vasculitis [74]. It is estimated that approximately 50% of all HUV patients will have renal involvement [75].

#### 3.4. Variable Vessels Vasculitides

##### Behçet's Disease (BD)

The most specific urological manifestations for BD are genital ulcerations, occurring in approximately 75% of patients with BD. Genital ulcers are most commonly found on the scrotum in men and the vulva in women. The ulcers are similar in appearance to the oral aphthae, and they are usually painful. An unusual reported localization of genital ulcers in BD is external urethral meatus [76]. The recurrence rate is typically less frequent compared to oral ulcerations. However, genital lesions are usually deeper and last longer than oral ulcers [76]. Due to the clinical heterogeneity of BD and the unpredictable disease course, the systemic treatment is tailored to the severity of symptoms and organ involvement. For mild oral and genital ulcerations, the current recommendation as first-line therapy is application of topical agents [76].

Other urological involvements are encountered scarcely, usually in the form of epididymitis [77]. There is a single case report of bilateral epididymitis in a 24-year-old man with ulcerous lesions, erythema nodosum, folliculitis-like exanthema, and multiple oral aphthae, eventually diagnosed as BD [78].

Another reported urological manifestation of BD is deep dorsal penile vein thrombosis (DDPVT). It requires an early etiological and symptomatic approach to preserve erectile function and prevent recurrences. Beddouche et al. presented a case of dorsal penile vein thrombosis revealed by spontaneous priapism that didn't resolve adequately. After the management of priapism and DDPVT, the etiological investigation revealed BD. The patient underwent etiological treatment with good clinical evolution and preservation of erectile function [79].

The urethrovaginal fistula formation may occur during the course of the BD. The pathogenesis of fistula is related to inflammation, and finally, necrosis caused by severe vasculitis. A few case reports are available in the literature [80]. Surgical treatment may be performed for repairing the urethrovaginal fistula, and in the mid-term, it may be successful. Long-term follow-up is necessary because of the high recurrence rate.

The incidence of bladder involvement in BD is considered to be 0.1% [81]. Direct involvement of the bladder is possible in the form of ulceration, nodules, or recurrent cystitis. Storage symptoms could also be observed in patients with BD as a result of neuro-Beçet's disease, direct bladder wall infiltration, or their combination [82]. Urge incontinence, emptying symptoms, and urinary retention may also occur. Sphincter function can be normal. However, detrusor sphincter dyssynergia or sphincter deficiency could also be observed [82]. Gross hematuria is very rare. Treatment of bladder dysfunction is individualized for each patient according to their urodynamic and imaging findings. Emptying failure is usually managed by clean intermittent catheterization while storage

problems have been treated by anticholinergics. However, augmentation cystoplasty becomes mandatory occasionally.

#### 4. Conclusions

Urological manifestations of PSV are infrequent. However, almost all organs of the genitourinary system can be affected. In general, the presence of vasculitis should be considered in patients who present with systemic or constitutional symptoms in combination with evidence of single and/or multiorgan dysfunction and especially with some key manifestations. Proper diagnosis requires careful assessment of all available clinical, laboratory, radiologic, and pathologic examinations. Fast diagnosis of PSV in patients with urological involvement is crucial in implementing appropriate therapy, and it allows to avoid unnecessary and invasive surgery in some cases.

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