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Urological Manifestations of Henoch-Schonlein Purpura: A Review

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Key Words
Henoch-Schonlein purpura • Kidney • Nephritis • Ureter • Bladder • Prostate • Penis • Scrotum

Abstract
Henoch-Schonlein purpura (HSP) is an immune-mediated systemic vasculitis generally found in children. The standard manifestations of HSP are palpable purpura, arthritis, abdominal pain, and renal complications. Although less common, there are significant urological manifestations associated with HSP. The primary objective of this review is to encourage better understanding and management of HSP by emphasizing the common and rare manifestations of HSP, how they are diagnosed, and the latest treatment options for mild to severe complications. Medline searches of HSP and its urological manifestations were conducted along with searches on current diagnostic and treatment methods. Urological manifestations of HSP involve the kidney, ureter, bladder, prostate, scrotum, testicle, and penis. Diagnosis and management of HSP are not always clear due to differential diagnosis and diversity of symptom presentation. Treatment for HSP is mainly supportive and includes use of nonsteroidal anti-inflammatory drugs for pain relief. In more severe cases, glucocorticoids, methylprednisolone, plasmapheresis, and peritoneal and hemodialysis are reported successful. It is important to note different symptoms of HSP in order to distinguish HSP from other diseases. Early diagnosis may prevent severe complications. Treatment options vary from conservative to invasive depending on the severity of the disease and time frame of diagnosis.

Introduction

Henoch-Schonlein purpura (HSP), also referred to as immunoglobulin A (IgA) vasculitis, is an immune-mediated systemic vasculitis commonly found in males between the ages of two and eleven [1, 2]. HSP is rarely reported in adults. Clinical manifestations of HSP include palpable purpura, arthritis, abdominal pain, and renal complications [3]. HSP is diagnosed by its clinical manifestations and the presence of IgA deposits in walls of arterioles, capillaries, and venules of afflicted organs [4, 5]. Diagnosis of HSP is not clear in many cases due to differential diagnosis and diversity of symptom presentation. It is important to review symptoms of HSP in order to prevent severe complications associated with delayed diagnosis. Urological involvement in HSP is rare, but is reported in several cases. The purpose of this paper is to discuss the urological manifestations associated with HSP that involve the kidney, ureter, bladder, prostate, scrotum, testicle, and penis.

Historical Perspectives

Herberden described the first case of HSP in 1801 in his book, On Cutaneous Diseases. Herberden described a young boy with a rash, abdominal pain, hematuria, and subcutaneous edema. Later, the disease was named Henoch-Schonlein purpura after Johann Schonlein and Eduard Henoch due to their role in establishing the clinical manifestations of the disease. Johann Schonlein identified purpura and joint pain as symptoms of HSP, which

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at the time he referred to as purpura rheumatica. Eduard Henoch later linked the disease to gastrointestinal and renal complications.

Pathogenesis

The cause of HSP remains undetermined and symptom severity and presentation vary with each case. Possible contributing factors that are associated with the onset of HSP include bacterial infections of the respiratory and urinary tracts [6, 7], infectious agents, drugs, food, chemicals, and insect bites [8]. Studies are still being conducted to determine whether genes influence HSP [9].

Urological Manifestations

Kidney

In approximately 50% of patients with HSP, renal function is compromised [10]. One common renal manifestation of HSP is nephritis, which is a rare type of kidney disease that can result in chronic kidney disease. Patients with HSP nephritis may experience hematuria and proteinuria and exhibit glomerular fibrin deposition with endocapillary/exocapillary inflammation. HSP nephritis is similar to IgA nephropathy, therefore, it is important to make distinctions between the 2 diseases. In general IgA nephropathy follows a more chronic course resulting in significant decline in renal function, whereas HSP nephritis is acute and kidney function normally returns to the patient's baseline [11].

HSP nephritis is associated with glomerulonephritis with IgA deposition, frequently seen after kidney transplants. Symptoms in these cases are similar to HSP nephritis in that patients may exhibit proteinuria, hematuria, abdominal pain, and elevated levels of serum creatinine. Shimizu et al. [12] reported a 46-year-old Japanese male diagnosed with HSP nephritis following kidney transplantation. The patient exhibited crescentic glomerulonephritis in his kidney graft after transplantation. Diagnosis of glomerulonephritis is determined by the presence of crescents and mesangial hypercellularity on the glomeruli, which are detected via biopsy of the kidney graft. In severe cases, allograft function deteriorates and hemodialysis is required. In order to prevent severe outcomes, HSP nephritis associated with glomerulonephritis with IgA deposition should be considered as a possible indicator of HSP.

HSP nephritis is considered the most common secondary kidney disease in pediatrics [13]; however, it affects adults more severely than children [14]. Pillebout et al. [2] reported 250 adults (average age of 50) suffering from HSP nephritis. Within 4 months after the onset of HSP nephritis, 32% of the patients experienced some decline in renal function, with the majority of patients experiencing hematuria and proteinuria. In general, 10–30% of adult patients with HSP nephritis were diagnosed with end-stage renal disease after approximately 15 years. In this study 13% of the adults had a creatinine clearance lower than 30 ml/min, and 14% had a creatinine clearance between 30 and 50 ml/min. However, 20% of patients showed no evidence of hematuria, proteinuria, or elevated serum creatinine. Due to the average age of participants in this study, it is essential to note that this study may not reflect prognosis of HSP nephritis in young adults. Though symptom severity varies among patients, elder patients are more likely to develop renal complications and should therefore be monitored in order to prevent severe outcomes.

Ureter

Although rare, HSP can affect the ureter resulting in ureter obstruction and ureteritis. These conditions may occur with HSP or after HSP has resolved. Obstruction is either unilateral or bilateral and may be partial or complete depending on the severity of the case. Ureter obstruction occurring alongside HSP may result from peri-ureteral vasculitis, which may lead to ureteral ischemia. Bruce et al. [15] reported a case in which a female patient with HSP exhibited bilateral ureteral obstruction as a result of peri-ureteral vasculitis. This patient was also diagnosed with ureteral ischemia. The diagnosis was made after discovering hydronephrosis on the contrast urography. In cases where obstruction occurs after HSP has resolved, ureter symptoms may be seen as late as 6 weeks after HSP symptoms disappear. Powell et al. [16] described an adult male with delayed unilateral, recurrent ureteric obstruction as a result of HSP. The patient did not exhibit symptoms of obstruction until 6 weeks after his HSP symptoms had resolved.

In many cases, identification of the stricture or mass causing obstruction is necessary for diagnosis and treatment. Radiographs are used to show strictures of the ureter, but further testing is required if pain progresses following stricture repair. Retrograde ureterograms are used to identify complete obstruction of the ureter, which may be due to an edematous ureteric epithelial mass. Removal of the mass typically results in recovery [16].

Bilateral and unilateral ureteritis are other complications associated with HSP. Symptoms of ureteritis typically appear about 1–2 months after the onset of HSP. For example, Corbett et al. [17] reported a 7-year-old African American boy with HSP who experienced bilateral sclerosing ureteritis, developing 4 weeks after the onset of HSP. Siomou et al. [18] reported a 3-year-old boy diagnosed with unilateral stenosing ureteritis 8 weeks after the onset of HSP. In some cases, as in the case described by Siomou et al. [18], ureteritis may be accompanied by nephritis, which may conceal underlying symptoms of ureteritis. This may result in delayed diagnosis of ureteritis and increased risk of complications. The necessity of nephrectomy and bilateral ureterocalycostomies increases as the risk for complications increases. In order to reduce this risk, routine ultrasounds of the urinary tract should be performed throughout the natural history of HSP.

Bladder

HSP has been reported to affect the bladder in rare cases. Occasionally, patients are diagnosed with HSP while undergoing treatment for bladder cancer. HSP symptoms may appear during or after treatment and typically follow clinical manifestations of HSP. Hirayama et al. [19] described an 83-year-old man who developed HSP while undergoing treatment for superficial bladder tumors. The patient underwent multiple transurethral resections and Bacillus Calmette-Guerin (BCG) intravesical therapy. Following the third course of therapy, purpura appeared on the lower region of the patient's legs, an uncommon side effect of BCG. The diagnosis of HSP due to BCG therapy was made. It is important to note that intravesical BCG is usually considered safe and not many cases have been reported with HSP side effects [20].

A second case reported by Ishigaki et al. [21] described a 70-year-old male with bladder cancer diagnosed with HSP following neoadjuvant chemotherapy and a radical cystectomy. After radical cystectomy, an abscess developed below the rectus abdominis muscle and the patient was placed on antibiotics. Twenty-three days after radical cystectomy, the patient presented with purpura, and the diagnosis of HSP was made based on a skin biopsy. The patient was treated with diaphenylsufone and prednisolone and was symptom free after 3 weeks. Because both cases were diagnosed within a short time frame, the patients did not experience severe complications. It is important that physicians continue to check patients for possible complications or chronic diseases that can result from HSP.

Prostate

HSP has been linked to prostate cancer. HSP patients with prostate cancer typically exhibit all or some of the following HSP symptoms: purpura, polyarthralgia, monocmonal IgA, leukocytoclastic vasculitis with IgA and C3 deposits, abdominal pain, diarrhea, and edema of the joints [22, 23]. Most reported cases involving the prostate describe the onset of HSP as occurring approximately 1 month following cancer diagnosis [24]. Garcias et al. [6] reported a 60-year-old male who began showing symptoms of HSP 4 weeks after undergoing surgery for Stage B2 prostate cancer. The patient developed arthlagias, myalgias, abdominal pain, and purpura on his extremities following a bilateral pelvic lymphadenectomy and I-125 implant. The relationship between the development of HSP and prostate cancer remains unknown, but Couzi et al. [22] postulated that tumor antigens or irregular IgA production may be contributing factors. In most cases, HSP symptoms resolve with conservative treatment, but rare cases have been reported in which prostate cancer patients with HSP do not respond well to immunosuppressive therapies in the absence of treatment for the malignancy, resulting in fatality [25].

Scrotum and Testicle

Scrotal and testicular manifestations associated with HSP are rare, however, a significant number of cases have been reported. Common testicular manifestations of HSP include acute scrotum, epididymitis, orchitis, and spermatic cord complications. Symptoms of scrotal involvement associated with HSP typically include clinical manifestations of HSP as well as scrotal pain, redness, and swelling. In a study of 120 Korean boys with HSP by Ha et al. [26], 26 boys presented with scrotal symptoms. Out of these 26 boys, 23 had scrotal swelling, 18 suffered from pain and soreness in the scrotum, and 7 had bilateral involvement. Thirteen of the boys showed epididymitis and 2 showed orchitis. Although a significant number of patients suffer from scrotal and testicular complications associated with HSP, diagnosis is often difficult.

Scrotal involvement is not easily linked to HSP because, in some cases, the typical HSP symptoms are not present or scrotal complications are seen after HSP symptoms have resolved. Misdiagnosis is still frequent in cases where scrotal symptoms occur alongside HSP symptoms due to the large volume of diseases that share similar symptoms with scrotal and testicular manifestations of HSP. Table 1 illustrates the anatomical organization of the differential diagnosis associated with scrotal and testicular pain.

Table 1. Anatomical differential diagnosis of scrotal/testicular pain

Spermatic cord	Testicle/epididymis	Tunica vaginalis	Scrotal skin
Spermatic cord thrombosis	extravaginal and intravaginal torsion of testicle	hematoma and hematocele	scrotal cellulitis
Entrapment syndrome of inguinal nerves and spermatic cord	intermittent torsion of testicle	hernia of a hydrocele	insect bites of scrotum
Thrombosis of pampiniform plexus	torsion of testicular appendages	hydrocele of hernia	acute idiopathic scrotal edema
Vasculitis of spermatic cord and testicular vessels	epididymitis and epididymorchitis		scrotal fat necrosis
Thrombosis of varicocele	testicular and scrotal abscess		Fournier's gangrene
Vasculitis	mumps orchitis		pneumoscrotum
Incarcerated inguinal hernia	Henoch-Schonlein purpura		pyocele
Inguinoscrotal hernias	testicular fracture/rupture		folliculitis
Filariae and schistosomiasis funiculitis	penetrating and blunt injury of scrotum/testicle		infected sebaceous cysts
Sclerotherapy/embolization	blast injury of scrotum and testicle global or partial testicular infarction orchialgia retractile testicle testicular compartment syndrome Postoperative testicular infarction secondary to sickle cell disease testicular tumors accessory ectopic spleen cysts of epididymis partial or incomplete torsion		

In cases where diagnosis is unclear, surgical exploration is required to exclude testicular torsion, orchitis, and other related disorders. Unlike torsion where the blood flow to the testicle is absent, HSP exhibits hyperemia or normal blood flow [27]. Orchitis is usually gradual in nature with associated nausea and vomiting, whereas scrotal involvement due to HSP presents with instantaneous pain, without nausea or vomiting [28]. Through use of imaging and other tests such as Duplex Doppler, an early diagnosis of orchitis or scrotal complications associated with HSP can be made and surgical exploration can be avoided [29]. In order to prevent delayed diagnosis, it is important to consider different disorders that mimic scrotal and testicular manifestations of HSP.

Hematomas and edema of the spermatic cord are complications of HSP. These complications are rare and may be confused with testicular torsion and acute scrotum, so it is important to make distinctions [30, 31]. Some HSP patients with spermatic cord involvement suffer from thrombosis of the spermatic veins. Diana et al. [31] described a pediatric case in which a young boy experienced thrombosis of the spermatic veins, a complication not typically seen in children. In this particular case, low-molecular-weight heparin treatment was effective in treating symptoms.

Penis

Penile manifestations of HSP include thrombosis and priapism. Thrombosis and priapism are rarely reported secondary to HSP. Sari et al. [32] described a patient that presented with thrombosis and priapism after being diagnosed with HSP. The case was a 37-year-old male who experienced penile thrombosis repeatedly after developing HSP, resulting in priapism. The patient did not have coagulopathies, therefore the timing of the initial HSP symptoms and onset of thrombosis imply a possible causational relationship.

Another penile manifestation of HSP is purpuric lesions on the penis. Such lesions may appear on the scrotum, glans penis, and shaft of the penis before or after the onset of HSP. Adhesions and swelling of the prepuce have been reported. One case described a 4-year-old boy exhibiting purpuric lesions on the scrotum, glans penis, and shaft of the penis shortly after being diagnosed with HSP. Thorough examination showed a lesion that spread from the middle of the penile shaft to the glans penis. The prepuce exhibited edema and adhesions, and the scrotum exhibited purple lesions. The base of the penis and testes appeared normal [33]. In this case and other similar cases, symptoms subside with supportive care. Burrows et al. reported a 19-year-old male who exhibited purpuric

Table 2. Differential diagnosis of HSP

Disease	Differentiating factors
Acute hemorrhagic edema of infancy (AHEI)	may not be considered a distinct disease
Hypersensitivity vasculitis	absence of IgA deposition
Microscopic polyangiitis, eosinophilic granulomatosis with polyangiitis, and systemic lupus erythematosus	not typically found in children
Juvenile idiopathic arthritis, and rheumatic fever	tests for serum complement, antinuclear and anti-dsDNA antibodies, and rheumatoid factor can help with differentiation

lesions on the penile shaft prior to the development HSP. This case was particularly difficult to diagnose because this patient did not initially present with classical signs of purpura. Palpable purpura did not appear until 4 weeks after HSP onset, though penile lesions persisted [34]. Because the timing of penile lesions in relation to the onset of HSP is not absolute, it is important for physicians to keep such complications in mind when making a diagnosis.

Differential Diagnosis

Before definitive diagnosis of HSP can be made other examples of vasculitis should be excluded. These include testicular vasculitis, polyarteritis nodosa, necrotizing granulomatous vasculitis, and spermatic cord vasculitis. Testicular vasculitis occurs in 2 forms: systemic testicular vasculitis and single-organ/isolated testicular vasculitis. Single-organ/isolated testicular vasculitis is identified by the presence of a testicular mass and normal lab results. Systemic testicular vasculitis more frequently presents with musculoskeletal manifestations, increased erythrocyte sedimentation rate, and anemia [35]. Polyarteritis nodosa commonly involves the spermatic cords and epididymis. Manifestations include bilateral scrotal swelling, pain, and epididymitis [35–37]. Necrotizing granulomatous vasculitis typically involves the spermatic cord and epididymis [38]. Spermatic cord vasculitis is associated with testicular infarction [39]. It is important to exclude idiopathic testicular infarction by routinely performing duplex Doppler of the scrotum in order to prevent a delayed diagnosis of testicular infarction. Fukuda et al. [40] reported a 12-year-old boy who was misdiagnosed with epididymo-orchitis secondary to HSP before testicular infarction was identified. Due to the delayed diagnosis, the boy required an orchiectomy. Upon initial diagnosis of HSP there were no signs of infarction hence it is unsafe to assume that normal testicular blood flow on duplex Doppler at the onset of HSP excludes future infarction. Testicular infarction may also develop as a result of Schistosoma mansoni [41]. By ruling out different types of vasculitis, physicians can diagnose HSP with more certainty. The differential diagnosis of HSP is illustrated in table 2.

Diagnosis and Radiology

Frequently the diagnosis of HSP is made by its classical clinical manifestations [42], which include palpable purpura on the lower limbs and buttocks, arthritis of the hips, knees, and ankles, and abdominal pain [43, 44]. Occasionally diagnosis may be less clear, requiring skin biopsies to examine for IgA deposition. A large amount of IgA deposits in the organ being tested favors the diagnosis of HSP [45].

Aside from skin biopsies that detect IgA deposition, there are currently no definitive laboratory tests to diagnosis HSP. Hematological work up should be performed to assess platelet and clotting function [46]. Thrombocytopenia and coagulopathy are not characteristics of HSP; therefore, their presence allows for the elimination of HSP. Urinalysis and chemistries are essential to assess renal function [47].

Scrotal symptoms can be further explored through sonography and ultrasounds with duplex Doppler and radionuclide scans, which exclude testicular torsion. Sonographic and ultrasonographic markers of HSP include hydrocele, swelling in the epididymis and scrotal skin, and normal blood flow to the testes. In patients with gastrointestinal manifestations of HSP presenting with severe abdominal pain, abdominal radiography and ultrasounds are utilized to identify dilated loops of bowel, hematomas, peritoneal fluid, and intussusception. CT scans are used in order to distinguish between different ureter and kidney complications. If renal function is severely impaired and a noninvasive approach is preferred, ultrasound of the kidney is recommended.

Treatment

HSP is a self-limited disease [48]. Treatment is mainly supportive involving rest, hydration, and symptom relief. If patients exhibit significant abdominal pain, gastrointestinal bleeding, joint pain, and renal insufficiency, or if they are unable to tolerate necessary hydration, hospitalization is recommended. The utilization of nonsteroidal anti-inflammatory drugs (NSAIDs) is successful in relieving arthritic symptoms and abdominal pain. Naproxen is the preferred NSAID, although Ibuprofen is also effective.

Glucocorticoids

Treatment with glucocorticoids and other steroids has been reported effective in some cases. Glucocorticoids have been reported to alleviate abdominal pain and lessen the likelihood of intussusception [49, 50]. Ben-Chaim et al. [51] reported immediate relief of scrotal symptoms after the administration of steroids. Prednisone with 1–2 mg/kg of daily has been reported effective in treating abdominal and joint pain [52]. However, glucocorticoids have only shown progress in treating symptoms associated with HSP and not in preventing renal complications associated with HSP [53].

Methylprednisolone and Currently Supported Medications

One study supports a high dosage of methylprednisolone as a successful treatment option for patients with severe HSP nephritis. A study involving 38 children with nephrotic syndrome demonstrates the positive effects of administering intravenous methylprednisolone prior to oral prednisone. By following this course of treatment, 90% of the children avoided end-stage renal disease. Of the 10% who had presented with end-stage renal disease, the majority of patients underwent delayed treatment, hence early intervention may be necessary to prevent severe renal outcomes [54]. Cyclosporin A, rituximab, and mycophenolate mofetil have also been used to treat HSP nephritis [55–57].

Plasmapheresis

Another method of treatment proven successful in some patients with severe crescentic disease and renal failure is plasmapheresis. However, plasmapheresis efficacy and potential side effects are still being evaluated. Chen et al. [58] reported a case in which double filtration plasmapheresis was effective in recovering renal function in a 33-year-old man with HSP associated crescentic glomerulonephritis. Plasmapheresis is known to improve cerebral vasculitis resulting from HSP. A case described a 7-year-old girl with HSP associated seizures, loss of vision, and loss of consciousness. She was given oral steroids and intravenous pulse methylprednisolone to treat her HSP nephritis; however, 10 days after the treatment she developed cerebral vasculitits. Plasmapheresis was performed and her condition noticeably improved [59].

Peritoneal and Hemodialysis

Peritoneal and hemodialysis are other treatment options in patients with severe renal disease. Vonend et al. [60] reported an 18-year-old male who suffered from several incidents of acute renal failure secondary to HSP. The patient underwent hemodialysis for 2 months following the initial episode of renal failure and recovered shortly after the onset of the second episode. Rapid recovery suggests the efficiency of hemodialysis in treating renal complications of HSP. Tsuboi et al. [61] reported a case in which a patient with end-stage renal disease due to HSP showed improvement in skin lesions after hemodialysis treatment but not with continuous ambulatory peritoneal dialysis. In this case, hemodialysis was the preferred, more effective treatment.

Renal Transplantation

HSP patients with severe renal disease are potential candidates for transplantation. However, it should be noted that transplantation is not a cure to HSP, and symptoms may recur. Moroni et al. [62] conducted a study involving 17 patients with HSP who underwent kidney transplantation and 42% of the patients being studied experienced the reappearance of HSP nephritis, which caused graft failure. However, Kanaan et al. [63] claim that rates of HSP nephritis recurrence are not as high as reported in other studies. Based on their study involving 43 kidney transplant patients with HSP nephritis, they predicted a 7.5% chance of graft failure in patients with recurring HSP nephritis at 10 years. The likelihood of graft failure after transplantation is debatable.

Conclusion

HSP is a small-vessel vasculitis generally found in children [4]. Typical manifestations include palpable purpura, arthritis, abdominal pain, and renal complications [3]. However, urological manifestations are reported in several cases. The kidney is more commonly affected by HSP compared to other urological organs [10]. HSP ne-

phritis is usually the presenting disease when the kidneys are involved [11]. Obstruction of the ureter and ureteritis may occur as a result of HSP [17]. Therefore, it is important to perform ultrasounds routinely throughout the course of HSP to assess for possible ureteric complications [18]. Several cases have been reported in which patients undergoing treatment for prostate or bladder cancer develop HSP. Scrotal swelling, orchitis, thrombosis, priapism, and purpuric lesions on the penis have also been observed in patients with HSP.

Treatment for HSP is mainly supportive. NSAIDs such as naproxen are proven to alleviate arthritis and abdominal pain. Glucocorticoids, specifically prednisone, are not recommended in the prevention of renal disease, but have been reported to alleviate abdominal pain and scrotal symptoms [51]. In patients with severe end-stage renal disease, administration of methylprednisolone, plasmapheresis, and hemodialysis may be beneficial.

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