Acute Scrotum in 7 Cases of Schoenlein-Henoch Syndrome

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Schoenlein-Henoch syndrome (SHS), one of the manifestations of systemic vasculitis, usually involves the skin, gastrointestinal tract, joints and kidney. Since the involvement of male genitalia is very rare and there is little mention of it in textbooks, doctors have a tendency to neglect this finding in SHS. Unless there is a confirming diagnosis, it is easily mistaken for testicular torsion and the patients undergo unnecessary operations because they complain of unbearable scrotal pain. SHS is not uncommon in Korea, but hardly any cases of scrotal involvement are found. We have experienced 7 cases of acute scrotum associated with SHS admitted to Severance Hospital, Yonsei University College of Medicine during the last 20 years; 2 underwent operation and 5 received conservative treatment only.

Key Words: Acute scrotum, Schoenlein-Henoch syndrome

Schoenlein-Henoch syndrome (SHS), also known as anaphylactoid purpura or allergic purpura, is one of the manifestations of acute systemic vasculitis related to circulating immune complexes including IgA (Hall et al. 1980). SHS usually involves the kidney, gastrointestinal tract, joints and skin. Except for rare cases that progress to renal failure, it is a disease which heals without complications.

The acute scrotum is a clinical syndrome with diverse etiological factors, including but not limited to epididymitis, abscess, tumor, torsion of testicular appendages, acute idiopathic scrotal edema and torsion of the spermatic cord. The latter is of the utmost concern since it requires immediate surgical treatment. The extreme tenderness over the affected area does not allow an adequate examination of the scrotum, making differential diagnosis difficult to establish and causing a poor correlation between clinical diagnosis and the findings at operation (Riley et al. 1976).

SHS may involve male genital structures and rarely, as a genitourinary symptom, painful edema of the scrotum, epididymis, spermatic cord and testis may be present (Counahan et al. 1977; Kher et al. 1983).

After the first report of scrotal involvement by Allen et al. (1960), rare though it may be, there have been reports of similar cases worldwide; but in Korea there have been hardly any cases reported of scrotal involvement. We are reporting our experience of 7 cases of acute scrotum manifested in SHS patients who were treated at the Department of Pediatrics of Severance Hospital, Yonsei University College of Medicine over the past 20 years.

CASE REPORT

Case 1

A 4 1/2-year-old boy was admitted for purpuric skin rashes on both lower extremities and buttocks,
and pain in both knee joints for 1 day.

At the time of admission, there was no abdominal pain, hematuria or any other physical findings except the rashes.

On blood analysis, WBC was 11,400/mm³, hemoglobin 12.4 g/dl, hematocrit 38.9%, platelet 306,000/mm³, and urinalysis showed no proteinuria or hematuria. The serum immunoglobulin data were IgG 1,220 mg/dl, IgA 185 mg/dl, IgM 167 mg/dl, and the ASO titer was 960 IU/ml. The serum electrolytes and blood chemistry data were within normal limits.

The child had abdominal pain from the third day of admission, and the stool occult blood was positive. On the 12th day, the left scrotum became edematous and tender with severe pain of the testis. Under the impression of testicular torsion, testicular exploration and orchiopexy was performed. The surgical findings showed no testicular torsion, but edema and enlargement of the epididymis suggestive of epididymitis. On the third postoperative day, scrotal pain had subsided. Dexamethasone and prednisolone were given and the child was discharged without any symptoms on the 19th day of admission.

**Case 2**

A 4-year-old boy was admitted for purpuric erythematous skin rashes on both lower extremities and buttocks for 17 days and abdominal pain and scrotal edema for 14 days. He was transferred from another hospital because of unresolving erythematous skin rashes and suspicion of testicular torsion.

The surgical findings did not show testicular torsion, but instead indicated signs of epididymitis. At the time of admission there were no specific findings other than purpuric skin rashes.

On blood analysis, WBC was 13,800/mm³, hemoglobin 14.6 g/dl, hematocrit 41.5%, platelet 593,000/mm³ and there was no proteinuria or hematuria in urinalysis. The serum immunoglobulin data were IgG 793 mg/dl, IgA 137 mg/dl, and IgM 177 mg/dl.

**Case 3**

An 8-year and 9-month-old boy had purpuric skin rashes on both lower extremities and right testicular pain for 9 days. He was admitted to the urology department. The symptoms subsided but he was transferred to the pediatric department because of abdominal pain which soon developed. There were no symptoms other than abdominal pain.

On blood analysis, WBC was 16,100/mm³, hemoglobin 12.3 g/dl, hematocrit 34.5%, platelet 521,000/mm³ and urinalysis showed no proteinuria or hematuria. The serum immunoglobulin were IgG 1,730 mg/dl, IgA 238 mg/dl, IgM 171 mg/dl, and the ASO titer was 480 IU/ml.

Testicular pain was the initial symptom presented along with the skin rashes. The pain and edema of the testicular inflammation subsided after 6 days of conservative treatment only.

**Case 4**

An 8-year-old boy was admitted for purpuric skin rashes on both buttocks and lower extremities, abdominal pain and pain in both knee joints for 4 days. At the time of admission, there were no hematuria, scrotal edema, pain or any other symptoms.

On blood analysis, WBC was 6,400/mm³, hemoglobin 11.6 g/dl, hematocrit 34.3%, platelet 299,000/mm³, and urinalysis showed no proteinuria or hematuria. The serum immunoglobulin were IgG, 1,700 mg/dl, IgA 293 mg/dl, IgM 266 mg/dl, and the ASO titer was 681 IU/ml.

On the 14th hospital day, left scrotal edema and testicular pain developed, suggestive of epididymitis, and the symptoms subsided after 3 days of conservative treatment only.

**Case 5**

A 5-year-old boy was admitted for pain in the right ankle and elbow joints for 4 days and abdominal pain and vomiting for 3 days. No purpuric skin rashes or scrotal edema were found at the time of admission.

On blood analysis, WBC was 16,700/mm³, hemoglobin 11.1 g/dl, hematocrit 34.2%, platelet 302,000/mm³, and urinalysis showed 100 mg/dl of proteinuria and many red blood cells. Serum immunoglobulin level was IgG 1,900 mg/dl, IgA 205 mg/dl, IgM 121 mg/dl, and the ASO titer was 960 IU/ml.

On the 3rd hospital day, purpuric skin rashes developed on both lower extremities, and on the
12th hospital day left scrotal pain developed. The increased blood flow to the left testis on the testicular scan strongly indicated the presence of epididymitis (Fig. 1). The symptom was relieved after 5 days of conservative treatment.

Case 6

An 11-year-old boy was admitted for having right testicular pain and abdominal pain for 3 days, and purpuric skin rashes on both lower extremities, right upper extremity and scrotum.

On blood analysis, WBC was 10,280/mm³, hemoglobin 13.3 g/dl, hematocrit 38.3%, platelet 350,000/mm³, and no proteinuria or hematuria were found in urinalysis. Serum immunoglobulin levels were IgG 1,930 mg/dl, IgA 362 mg/dl, IgM 125 mg/dl, and the ASO titer was 340 IU/ml.

Testicular pain was the initial symptom developed, along with abdominal pain. Testicular pain and epididymitis subsided after 4 days of conservative treatment only.

Case 7

A 7-year and 5-month-old boy was transferred from a local hospital for scrotal enlargement and pain. He had purpuric skin rashes on both lower extremities for 2 days and pain and edema on both ankle joints for 1 day. At the time of admission, the size of the left scrotum was increased to a diameter of 4 cm, and the testis was tender. The epididymis was hard and enlarged. Ecchymosis was seen on the scrotal wall (Fig. 2).

On blood analysis, WBC was 12,700/mm³, hemoglobin 12.8 g/dl, hematocrit 39.0%, and platelet 410,000/mm³. Urinalysis showed no proteinuria or hematuria. Stool occult blood was negative.

On the 2nd hospital day the size of enlarged scrotum decreased to 2.5 cm, but pain was still present. On the 4th hospital day the lesions on the scrotal wall almost disappeared. Dexamethasone and prednisolone were given and the patient was discharged on the 7th day with a small amount of skin rash left on both elbows and feet (Table 1).

**DISCUSSION**

SHS, an acute systemic vasculitis, rarely involves
Table 1. Characteristics of Schoenlein-Henoch syndrome patients with acute scrotum

<table>
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<tr>
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<th>Case 1</th>
<th>Case 2</th>
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<tr>
<td>Age (year)</td>
<td>46/12</td>
<td>4</td>
<td>89/12</td>
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<td>5</td>
<td>11</td>
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<td>2\textsuperscript{2}\textsuperscript{3}-3.1-5</td>
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*: 1: skin rash, 2: joint pain, 3: abdominal pain, 4: vomiting, 5: scrotal symptoms
\textsuperscript{*}: 1: knee, 2: knee, ankle and wrist, 3: ankle and elbow, 4: knee, 3: ankle

male genitalia and elicits symptoms of acute scrotum such as severe pain and enlargement (Counahan et al. 1977; Kher et al. 1983). Many cases involving male genitalia in SHS (Fitzsimmons, 1968; Nousias et al. 1969; Löh and Jalan, 1974; Khan et al. 1977; Mikuz et al. 1979; Stein et al. 1980; O’Regan and Robitalle, 1981; Rodriguez et al. 1981; Caldamone et al. 1984) have been reported since the first discovery of scrotal and testicular bleeding by Allen et al. (1960).

Symptoms of vasculitis in the genital organ include pain, edema, enlargement and ecchymosis. The involved sites are the scrotal wall (Fitzsimmons, 1968; Haahr and Sparrevoehn, 1971), testis (Fitzsimmons, 1968; Loh and Jalan, 1974; Turkish et al. 1976), testicular appendage (Fitzsimmons, 1968), spermatic cord (Eadie and Higgins, 1964; Counahan et al. 1977; Mikuz et al. 1979) and the epididymis (Fitzsimmons, 1968; Haahr and Sparrevoehn, 1971; Turkish et al. 1976). As well, testicular infarction (Mikuz et al. 1979) and torsion (Loh and Jalan, 1974) have been reported. By 1986 there were reports of 5 cases in which severe testicular symptoms were the first and only symptoms in SHS (Eadie and Higgins, 1964; Haahr and Sparrevoehn, 1971; Mikuz et al. 1979; O’Regan and Robitalle, 1981; Clark and Kramer, 1986).

The frequency of scrotal and testicular involvement in SHS has been reported as 2~38% (Byrn et al. 1976; Khan et al. 1977; Clark and Kramer, 1986). Khan et al. found 15% (9/59 cases) of SHS having scrotal edema, and among them 4 patients had symptoms similar to testicular torsion (Loh and Jalan, 1974; Khan et al. 1977). Caldamone et al. found 4 cases of acute scrotum among 150 children with SHS(Caldamone et al. 1984). O’Regan and Robitalle found 3 patients of SHS among 89 patients who were surgically operated on under the impression of testicular torsion (O’Regan and Robitalle, 1981).

The diagnosis of testicular torsion was confirmed by surgery in 1 case (Loh and Jalan, 1974) in which scrotal enlargement, bleeding and edema on the spermatic cord and testis, vasculitis on the spermatic cord and testicular torsion were found on the third day of onset of the disease.

Haahr and Sparrevoehn reported that among 6 cases of epididymitis in children, 2 cases occurred in conjunction with SHS, and the surgical findings showed severe edema and enlargement of the epididymitis. The testis had a normal external appearance (Haahr and Sparrevoehn, 1971).
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Epididymitis usually occurs about age 5, and is accompanied by pain and enlargement of the scrotum, tenderness of the testis, and color changes of the scrotal skin. Under the impression of testicular torsion, there have been a number of cases undergoing surgery, but the findings have been otherwise normal, except for purpura and petechia over the epididymis. Epididymitis does not require surgery. The scrotal pain and enlargement usually disappears in 3–5 days (Fitzsimmons, 1968; Haahr and Sparrevoorn, 1971; Turkish et al. 1976).

All of our 7 patients also had scrotal pain and enlargement, and testicular tenderness. Two underwent exploratory incision. The testes were found to be normal and the epididymis was enlarged and edematous. The 5 remaining patients were improved in 3–6 days with only conservative treatment.

Mikuz et al. reported a case of SHS in a 6-year-old boy who had an appendectomy and orchietomy and later developed purpura on the skin (Mikuz et al. 1979). The boy had a hemorrhagic infarction on the testis and spermatic cord. Just as vasculitis becomes the predisposing factor of intussusception in the gastrointestinal tract, vasculitis of the testis with bleeding and edema may be the predisposing factor of testicular torsion (Loh and Jalan, 1974).

In evaluating an SHS child, imaging helps the diagnosis and prevents testicular exploration (Stein et al. 1980). Rodriguez et al. reported that in evaluation of an acute scrotum, compared with the surgical findings, the diagnostic probability of the testicular scan was 100%, and the Doppler ultrasound showed a 79% positive correlation (Rodriguez et al. 1981). But the Doppler ultrasound flow study frequently shows false-positive results in differentiating epididymitis and acute scrotal torsion. This is mainly due to the severe edema of the surrounding tissues. Case 5 showed increased blood flow in the left scrotum in the testicular scan, which helped us differentiate from testicular torsion.

The acute scrotum is frequently misdiagnosed as testicular torsion and requires unnecessary surgical procedures (Nousias et al. 1969). Since there was only one confirmed case reported as testicular torsion, when testicular torsion is suspected, careful observation of other SHS symptoms should be made and conservative treatment should be carried out (Fitzsimmons, 1968; O'Regan and Robitaille, 1981).

On the other hand, there was a report of Loh and Jalan (1974) that testicular torsion should be differentiated by surgery. Turkish et al. reported that testicular and scrotal bleeding is rare in SHS and torsion is difficult to diagnose when it is a complication of the testicular vasculitis; and these two conditions may exist at the same time (Turkish et al. 1976). If there is no pain and enlargement of the testis, there is a greater probability that only the scrotum may be involved, and therefore conservative treatment should be done.

Generally, epididymis, testicular and scrotal lesions in SHS are not mentioned in pediatric textbooks. Since there is not much emphasis of this in the literature, doctors neglect these findings in SHS. Unless there is a confirmative diagnosis, it is easily mistaken for testicular torsion (Fitzsimmons, 1968; Khan et al. 1977).

There have been reports of both the uncertain efficacy and effectiveness of oral prednisolone therapy in the treatment of acute scrotum.

REFERENCES


Number 1