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HENOCH–SCHONLEIN PURPURA - A CASE REPORT OF A NINE YEAR OLD GIRL

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ABSTRACT

Henoch-Schonlein purpura is a disease which causes irritation and swelling of the small blood vessels in the skin resulting in a rash. The illness usually affects children from 2 to 10 years of age, but it may affect people of any age. It is characterized by purple spots on the skin, joint pain, digestive problems, and glomerulonephritis. It develops primarily as an immune response to an infection however the cause is not fully understood. Most cases relieve without initiation of therapy, but in caseswhich require treatment the main aim is to relieve symptoms such as joint pain, abdominal pain, or swelling. We describe a case report of a pediatric patient who presented with reddish purpuric spots and erythematous rashes over both lower limbs.

KEYWORDS: Henoch-Schonlein purpura, Immunoglobulin vasculitis, Purpura, Rash.

INTRODUCTION

Henoch-Schonlein pupura (HSP), also known as immunoglobulin vasculitis[1] is referred to as a systemic vasculitis with a prominent cutaneous component characterized by the tissue deposition of IgA containing immune complexes.[2] The disease is characterized by its hallmark symptoms of a tetrad of clinical manifestations that are palpable purpura in patients neither with thrombocytopenia nor coagulopathy, arthritis/arthralgia, abdominal pain and renal disease. It is the most commonly observed form of vasculitis in children with an annual incidence of 140 cases per million persons. The mean age of 5.9 years is seen in patients affected with this disease.[3]

The symptoms of HSP include rash, digestive tract problems such as vomiting and abdominal pain, arthritis and kidney involvement (hematuria being the most common sign). Symptoms affecting the central nervous system such as seizures, affecting lung such as pneumonia are seen in very rare cases. HSP is diagnosed with the presence of a characteristic rash. Antibody deposits on the skin can confirm HSP. Kidney biopsy and urine tests are also used to investigate the presence of hematuria and proteinuria.

There is no known cure for people affected with Henoch-Schonlein purpura (HSP). Aim of the treatment is to relieve the symptoms of this condition. Non-steroidal anti-inflammatory drugs (NSAIDs) or corticosteroids (such as prednisone) can be used to in certain cases to relieve pain. If the kidneys are severely damaged in an individual with HSP, immunosuppressive medications, such as cyclophosphamide, may be prescribed.[4][5] In rare cases, individuals with HSP may hospitalization if they experience severe abdominal pain, bleeding from the digestive tract, or kidney problems.

CASE REPORT

A 9 year old female child was admitted in the IP department with reddish spots and erythematous rashes over both the lower limbs. The patient experienced difficulty in walking. The patient had high grade intermittent fever (102 F) and complaints of mild upper abdominal pain. General examination revealed that the patient was dull looking, had tenderness over both the ankle joints and experienced bilateral pedal edema. Her vitals were within the normal range. Laboratory workup was as in the table below. Urine analysis reported of proteinuria (urine protein-1227 mg/dl). Pus cells (32/HPF), RBC (20/HPF), epithelial cells (108/HPF) were also elevated in the urine analysis.

Table No. 1: Laboratory investigations.

Sr. No.	Component	Patients Level(Reference Range)
1	Hemoglobin(g/dl)	10.4 (12-15)
2	RBC(*10^{12} /L)	3.75 (3.5-5.5)
3	ESR(mm/hr)	105 (Up to 20)
4	C reactive protein (mg/L)	34.3(<3)
5	Serum urea (mg/Dl)	64
6	Serum creatinine(mg/dl)	1.8(0.6-1.2)
7	Platelet (*10^{9} /L)	316(100-300)
8	WBC(*10^{9} /L)	9.2(4-10)

Ultrasound of the abdomen and pelvis showed raised corticalechoes of both kidneys and minimal free fluid pelvis. A renal biopsy was planned for the patient but as she had

hypocomplimentemia, it was deffered. She was treated with oral diuretics (T.Dytor 5 mg), IV abtibiotcs (Inj. Ceftriaxone 1 gm) and other supportive. The child improved spontaneously.

DISCUSSION

Henoch-Schönlein purpura is a disease that causes small blood vessels in the body to become inflamed and leak. The primary symptom is an erythramatous rash. It can occur at any time of the life time, but it predominantly affects children under the age of 11. A London physician, Dr. William Heberden, described the first cases of Henoch-Schönlein purpura (HSP) in 1801. Decades after Heberden, additional cases regarding the disease was reported by Edouard Henoch (1874) and Johann Schönlein (1837).

Henoch-Schonlein purpura is an abnormal immune system response disease in which the body's immune system attacks the body's own cells and organs. the immune system generally makes antibodies, or proteins, to protect the body from foreign pathogens such as bacteria or virus. in hsp, these antibodies target and destroy the blood vessels. the factors that cause the immune response is mostly unknown. in more than 30 percent of patients an upper respiratory tract infection before getting HSP is precipitated.[6] Other factors related to HSP are infectious agents such as chickenpox, measles, hepatitis, and HIV virus's medications, foods, insect bites, exposure to cold weather and trauma.

HSP usually produce vomiting and abdominal pain that ranges from mild to severe. Pain and swelling of the joints are mains symptoms of HSP that mainly affects the knees and rarely the elbows and wrists. Hematuria— blood in the urine—is a common sign that HSP has affected the kidneys. Proteinuria—large amounts of protein in the urine—or development of high blood pressure suggests more severe kidney problems. The major complication of HSP in children is kidney failure. Up to 40 percent of adults with HSP will have CKD or kidney failure within 15 years of diagnosis.[7] HSP is mainly diagnosed with the presence of a typical rash, arthritis and abdominal pain. Occasionally, when the diagnosis is uncertain, skin and kidney biopsies are performed for the diagnosis.[8] Physicians may also recommend an ultrasound to rule out other causes of abdominal pain and to check for possible complications. HSP is usually self-limited. Therefore, treatment is not indicated in all cases, and full recovery is the rule. It usually goes away on its own within a month with no lasting ill effects.[9]

CONCLUSION

The main aim of treatment is to relieve symptoms such as pain in the joints and abdomen and swelling. People with kidney damage may receive treatment aimed at preventing long-term kidney disease.

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