UNUSUAL PRESENTATION OF HENOCH-SCHONLEIN PURPURA IN CHILDREN - A CASE REPORT

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ABSTRACT
Henoch-Schönlein purpura (HSP) is an acute immunoglobulin A (IgA)–mediated (Coppo et al., 1984; Davin et al., 1987) disorder characterized by a generalized vasculitis involving the small vessels of the skin, the gastrointestinal (GI) tract, the kidneys, the joints and rarely, the lungs and the central nervous system (CNS) (Saulsbury, 2001). It is a subset of necrotizing vasculitis characterized by fibrinoid destruction of blood vessels and leukocytoclasis. Being most common vasculitis (Gardner-Medwin et al., 2002) in children, we are reporting a case of HSP in 8yrs old child with unusual presentation (mono-articular arthritis, epididymitis, skin involvement confined only to legs) requiring high degree of suspicion for diagnosis.

Keywords: HSP, Vasculitis, IgA Immunoglobulin

INTRODUCTION
The disease is named after Eduard Heinrich Henoch (1820–1910), a German pediatrician and his teacher Johann Lukas Schönlein (1793–1864), who described it in the 1860s. Schönlein associated the purpura and arthritis, and Henoch the purpura and gastrointestinal involvement.

The English physician Heberden (1710–1801) and the dermatologist William (1757–1812) had already described the disease in 1802 and 1808, respectively, but the name Heberden–Willam disease has fallen into disuse.

William Osler was the first to recognise the underlying allergic mechanism of HSP. HSP peaks in children aged 3-10 years, but the condition is also seen in adults (Blanco and Martinez-Taboada, 1997; Pillebout, 2002). The male-to-female ratio is 1.5:2:1. The incidence of HSP in children is about 20 per 100,000 children per year, making it the most common vasculitis in children (Gardner-Medwin et al., 2002).

CASES
8years old boy, 2nd issue of non-consanguineous married couple came to emergency department with complaints of rashes over legs with acute onset since 2days and progressive joint pain & swelling in left knee, swelling and pain in right scrotum since 1day. Pain in child was so severe, that he is crabbing and unable to move from single position.

There was no history of fever or cough with expectoration or pain abdomen or hematuria. No history of drug intake. There was history of sore throat 1week prior to presentation for which no medical aid was given and child was treated at home with salt water gargling. No history of asthma or allergy in person or in family.

At presentation, child was conscious and oriented and vitals were stable. On examination he had erythematous maculopapular rash over legs bilaterally, with rashes coalescing to form large lesions looking like ecchymotic patches and had left knee joint swelling, tenderness and severe limitation of joint movements and swelling with severe tenderness of right scrotum. Investigations revealed an elevated ESR and C-reactive protein. USG scrotum revealed b/s/o Right epididymitis. USG & X-ray left knee suggested mild joint effusion. Urine routine and USG
Figures 1: Mono-articular arthritis, scrotal involvement, skin lesions (maculopapular rash) respectively

Abdomen were within normal limits. ASLO titres were positive suggesting previous streptococcal infection. A provisional diagnosis of HSP was made and oral steroids were started keeping severity of presentation in mind. Perilesional Skin biopsy was taken for immunofloresence and was positive for IgA vasculitis conforming the diagnosis. Child was successfully treated and showed dramatic improvement for steroids.

DISCUSSION

The dominant clinical features of HSP include cutaneous purpure (Henoch, 1974), arthritis (Aliyazicioglu et al., 2007; Blanco and Martínez-Taboada, 1997; Gedalia, 2004; Szer, 1994) (75%), abdominal pain (80%), GI bleeding and nephritis (Coppo et al., 1984) (50%). In one half to two thirds of children, an upper respiratory tract infection (URTI) precedes the clinical onset of HSP by 1-3 weeks (Rigante et al., 2013). In general, patients with HSP appear mildly ill. They often have a fever, with a temperature that usually does not exceed 38°C (100.4°F). HSP is typically an acute, self-limited illness, and treatment is primarily supportive. However, few cases present with serious complications like bowel ischemia, renal failure, orchitis which warrants therapy. Here in our case child did not present with pain abdomen (GI symptoms) which is most common presentation of complicated HSP but only with Skin lesions, severe
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mono articular arthritis and epididymitis. By high degree of suspicion provisional diagnosis of HSP was made, skin biopsy for confirmation of diagnosis was done. Child was successfully treated with short course of prednisolone in view of severity of presentation with epididymitis which is rare and arthritis involving single large joint which was limiting child’s daily activities without abdominal involvement.

Conclusion

Although HSP is a common small vessel vasculitis (Gardner-Medwin, 2002) in children below 10yrs of age with skin involvement (Henoch, 1974), abdominal signs, multiple joint involvement (Blanco and Martínez-Taboada, 1997; Gedalia, 2004; Szer, 1994) and orchitis, few cases, like in our case, can also present with rare atypically, like very severe mono-articular arthritis, epididymitis and skin involvement limiting only to legs. Hence high degree of suspicion is needed and skin biopsy helps for confirmation of diagnosis. Steroid therapy is required even in absence of abdominal symptoms and renal involvement to hasten the recovery.

REFERENCES