Case report

An unusual case of transient cortical blindness with sagittal sinus thrombosis in a case of Henoch-Schonlein purpura

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Abstract

Introduction: Henoch-Schonlein purpura (HSP) is one of the most common causes of small vessel vasculitis in children, but sometimes may have an atypical presentation. **Objective**: To report an unusual case of transient cortical blindness in a patient with Henoch-Schonlein purpura. **Case**: A 3-year-old female child was brought with the complaint of diffuse abdominal pain and hematochezia, which was preceded by high grade fever and cough. Three days later she developed hematuria, hematemesis, melena and hemoptysis along with palpable purpura. Four days later she became irritable and developed a few episodes of generelized tonic clonic seizure, followed by cortical blindness. The CT scan of the brain showed bilateral non-enhancing occipital hypodensity. The magnetic resonance venography showed thrombosis in transverse and sigmoid sinus. She was treated with corticosteroids and her mental status and vision improved. **Conclusion**: The HSP can cause transient cortical blindness, and recovery is good if therapy is initiated at the appropriate time.

Key-words: Henoch-Schonlein purpura, cortical blindness, magnetic resonance venography

Introduction

The HSP is one of the most common vasculitides of childhood (Cassidy et al, 2005). A diagnostic triad of purpuric rash, arthritis, and abnormalities of the urinary sediment was proposed by Schonlein in 1837, and Henoch described the association of purpuric rash,abdominal pain with bloody diarrhea, and proteinuria in1874 (Cassidy et al, 2005). The etiology is unknown, but HSP often follows an upper respiratory tract infection. Males are affected twice as frequently as females (Cassidy et al, 2005). HSP is an IgA-mediated vasculitis of small vessels. Immunofluorescence techniques show deposition of IgA and C3 in the small vessels of the skin and the renal glomeruli; the role of complement activation is controversial. To fulfil classification criteria for HSP by the existing ACR criteria, two of the following were required: age less than 20 years, palpable purpura, abdominal pain, and vessel wall granulocytes on biopsy (Cassidy et al, 2005). The EULAR-PReS consensus criteria, for HSP are palpable purpura (mandatory criterion) in the presence of at least one of the following four criteria: diffuse abdomonal pain, any biopsy showing predominent IgG deposition, athritis (acute, any joint) or arthralgia, renal involvement (any hematuria

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and/or proteinuria) are required (Miller et al, 2007). CNS manifestations of HSP are rare and produce significant morbidity. Anticardiolipin or antiphospholipid antibodies may be present and contribute to the intravascular coagulopathy.

Case report

A 6-year-old female child presented with diffuse abdominal pain and haematochezia for 3 days with a history of high grade fever and cough for 5 days before. Abdominal pain was severe and only partially relieved by antispasmodics. There were also slightly erythematous painful swellings over the right sole and forehead. Three days later, palpable purpuraed over the knee joint and ankle, which later spread to involve thigh, trunk and forearm. At the same time, she develops hematuria, hematemesis, melena and hemoptysis. Seven days after admission, she developed generalized tonic clonic seizures, which recurred 2-3 times thereafter. Following this, she completely lost her vision and became very irritable. Her menace reflex was lost, but light reflex was present. There was no involvement of other cranial nerves and no focal neurological deficit was recorded. Her Blood Pressure was within the 50th percentile. Her speech was intact. Her hemoglobin was 12.4 gm/dl, platelet-435×10^3/cm, RAF, ANA and anticardiolipin antibody was negative. The prothrombin time and activated partial thromboplastin time was 13.2 and 34 respectively. The CT scan of head showed "bilateral occipital patchy non-enhancing hypodensity", the MRI showed "mild diffuse brain shrinkage" and the MR venography showed "cerebral venous sinus thrombosis in left transverse and sigmoid sinuses with extension into left juglar vein". She was commenced on corticosteroids, following which her hematuria and hemoptysis disappeared, mental status improved, and the melena and purpuric spots diminished. Low dose aspirin was started on the basis of the MR venogram. She improved rapidly thereafter.

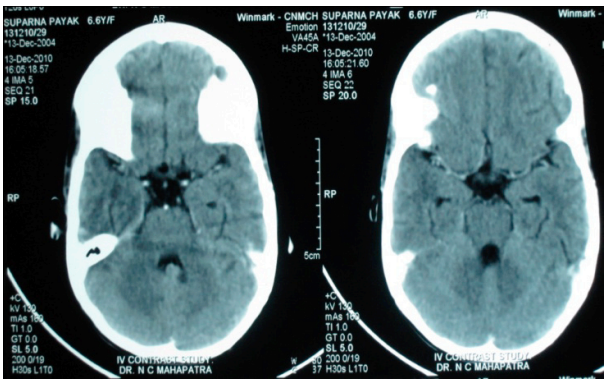

Figure 1: CTS shows bilateral non-enhancing occipital lobe lesion

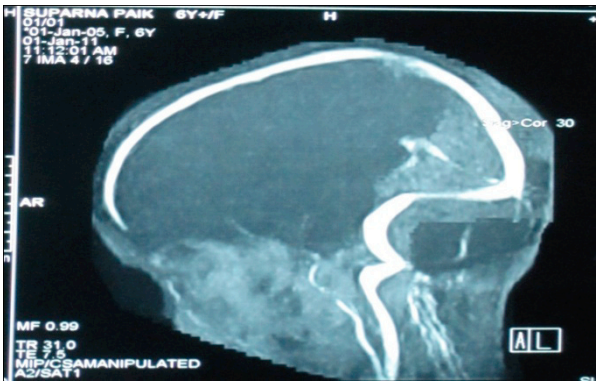

Figure 2: MR venogram showing left transverse sinus thrombosis

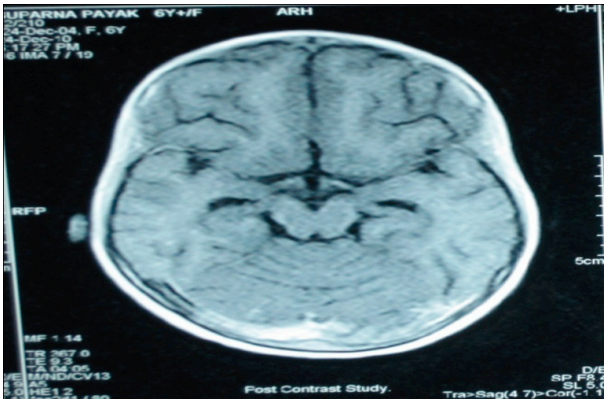

Figure 3: MRI showing diffuse brain shrinkage

Discussion

This case was diagnosed as HSP on the basis of palpable purpura, abdominal pain with bleeding manifestation and normal platelet count. In this particular case, the CNS manifestation in the form of blindness and seizure was also supported by the CT scan, MRI and MR Venography showing

cerebral venous thrombosis. From 1969 to 2009, 54 cases were published with CNS manifestations worldwide, of which 44 cases were aged less than 20 years (Ozen et al, 2006). Similar cases reported from India were very rare (Ozen et al, 2006). The cause of CNS involvement is either cerebral vasculitis or intra-cerebral hemorrage. CNS involvement can present with or without hypertension in rare cases, Henoch-Schoenlein purpura can be associated with seizures, paresis, coma, altered mental status, apathy, hyperactivity, irritability, mood liability, somnolence, seizure, and focal deficits (eg, aphasia, ataxia, chorea, cortical blindness, hemiparesis etc).

Polyradiculoneuropathies and mononeuropathies may also occur. The initial management of patients with suspected cerebral Henoch–Schönlein syndrome includes control of arterial hypertension, seizures and repair of disordered hemostasis. Some trials with plasmapheresis have also been reported (Garzoni et al, 2009; Chen et al, 2000). Anti-coagulation agents are advised in patients with secondary anti-phospholipid syndrome (Abend et al, 2007). Regular follow up is required as the renal manifestations can occur 6 weeks after the appearance of skin rash.

Conclusion

The HSP can present with CNS manifestations including transient cortical blindness. It is potentially reversible if the treatment is given at the appropriate time.

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