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CASE-BASED UPDATE

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Cerebral hemorrhage in Henoch-Schoenlein syndrome

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Abstract *Background:* Henoch-Schoenlein syndrome (HSS) is the most common form of vasculitis seen in childhood. The clinical diagnosis is based on the association of nonthrombocytopenic purpura, arthritis and abdominal pain. Nephropathy is the most common complication. Hemorrhages can occur in the respiratory, gastrointestinal and urinary tracts. Neurological complications are rare, though they may be particularly severe. *Case report and discussion:* Intracranial hemorrhage is an extremely rare complication of the disease; we report the case of a child with cerebral hemorrhage in HSS and review the literature.

Keywords Henoch-Schoenlein syndrome · Intracranial hemorrhage · Pediatric vasculitis

Background

Henoch-Schoenlein syndrome (HSS) is one of the most common forms of vasculitis seen in childhood. It occurs primarily in children aged 2–8 years, with a male-to-female ratio of 2 : 1 [12].

The clinical diagnosis is based on the association of nonthrombocytopenic purpura, arthritis and abdominal pain; abdominal hemorrhage and nephropathy can be associated features [12]. Nephropathy is the commonest and most dangerous complication. The histological findings are necessary to confirm the diagnosis and consist in necrotizing vasculitis of the small vessels with immunocomplexes and IgA deposits [3]. In rare cases HSS vasculitis leads to serious hemorrhages in the respiratory [15] and gastrointestinal [16] tracts, testicles [13], bladder [1] and skin [10].

Neurological complications

Neurological complications are rare, though they may be particularly severe [14]. CNS involvement, first described by Osler [8] in 1914, is characterized by mild symptoms such as headache, irritability, peripheral neuropathy, behavioral alterations and reduction in the level of consciousness [3]. These symptoms, although rare, are often underestimated: in their series, Ostergard and Storm showed behavioral alterations in 31% of patients and EEG anomalies in 46% [9]. Mild symptoms may be caused by intracranial hypertension, cerebral edema and metabolic disorders, all of which often occur in HSS. The neurological symptoms vary with the area involved and the severity of the cerebral vasculitis, which can easily be detected by MRI [6]. Intracranial hemorrhage (Table 1) is an extremely rare complication of the disease; we have only been able to find five cases reported in the literature [2, 4, 5, 7, 11]. Histologically the cerebral vasculitis is characterized by inflammation, induced by circulating IgA immunocomplexes, as seen in the intestinal, renal and skin lesions [3]. According to these few cases reported in the literature, the site of the hemorrhage is most likely to be intraparenchymal, with parieto-temporal hemorrhage the most prevalent.

Table 1 Demographic and clinical characteristics of children with Henoch-Schoenlein syndrome and intracerebral hemorrhage described in literature

Reference	Sex	Age (years)	Hemorrhage site	Neurological signs and symptoms	Associated coagulative test anomalies	Associated noncutaneous hemorrhages	Treatment	Outcome
[11]	Male	7	Right occipital lobe	Drowsiness, blood hypertension	No	Gastrointestinal hemorrhage	Medical (steroids, diuretics)	Left hemianopsia
[5]	Male	8	Left parieto-occipital region	Unconsciousness, headache, seizures	>PT and PTT	Gastrointestinal hemorrhage	Medical (fluid restriction, steroids, phenobarbital)	Right hemianopsia
[2]	Male	9	Right temporo-parietal-occipital region	Unconsciousness, left hemiplegia, mydriasis, positive Babinski sign	No	Gastrointestinal and renal hemorrhage	Surgical (hematoma evacuation)	Complete recovery
[4]	Female	8	Right fronto-parietal region	Headache, seizures, diplopia, lower left limb paresis	No	Renal hemorrhage	Medical (fluid restriction, steroids)	Complete recovery
[7]	Female	5	Left parietal region and subarachnoid space	Vomiting, drowsiness headache	No	Gastrointestinal hemorrhage	Medical (not specified)	Complete recovery

Diagnosis

The diagnosis is usually made following CT and MR examination carried out in children showing behavioral alterations, EEG anomalies, focal neurological deficits and signs of increased intracranial pressure.

Management

In rare cases, cerebral intraparenchymal hemorrhage in HSS may require urgent neurosurgical treatment [2]. In most cases, medical management (steroids, fluid restriction) is sufficient to control intracranial hypertension and to reduce the evidence and severity of late neurological sequelae, as shown by the favorable outcomes recorded in nearly all cases described in the literature [2, 4, 5, 7, 11].

Illustrative case

This 8-year-old girl was admitted to a district hospital for worsening abdominal pain. Blood tests and abdominal echography were performed, giving results within the normal ranges (white blood cells: 13,000/mm^3; red blood cells: 4,500,000/mm^3; platelet count: 450,000/mm^3). As the abdominal pain persisted the child underwent an explorative laparoscopy, which showed only hypertrophy of the lymph nodes. Twenty-four hours after surgery the child developed a purpura localized to the lower limbs and glutei. Hematuria (10–12 red blood cells/high power field) and proteinuria (350 mg/dl) led to the diagnosis of Henoch-Schoenlein syndrome. Thirty-six hours after the appearance of the purpura, the child developed an intense headache followed by tonic-clonic seizures, which required i.v. diazepam. After resolution of the seizures the child was in a comatose state and was referred to our Pediatric Intensive Care Unit (PICU). Physical examination on arrival revealed a reduced the level of consciousness and the presence of purpuric lesions on the lower limbs and elbows. A second episode of generalized tonic-clonic seizure occurred, which lasted for 2 min after a bolus of i.v. lorazepam (0.1 mg/kg). The Glasgow Coma Score (GCS) after the crisis was 9. On physical examination

Fig. 1 CT scan showing a deep hemorrhagic lesion in the right fronto-parietal region with perilesional edema in an 8-year-old girl with Henoch-Schoenlein syndrome (HSS)

paresis of the left lower limb was noted. An infusion of phenobarbital (5 mg kg^{-1} day^{-1}) was started and a cerebral CT scan performed. A very wide and deeply located hemorrhagic area was shown in the right fronto-parietal region with perilesional edema (Fig. 1). Dexamethasone (0.3 mg^{-1} day^{-1} i.v.), subdivided into four daily doses, was started. In order to exclude a vascular anomaly we performed a cerebral angiography, which was negative. Blood counts and coagulation tests were normal. An EEG was carried out, showing diffuse slow activity, preeminently in the right fronto-parietal region. Proteins and blood cells were still present in the urine. After 5 days of dexamethasone therapy a further CT scan was performed, which showed reabsorption of the hemorrhagic area and reduction of the perilesional edema. Therapy was progressively tapered off as the child continued to recover, though she developed arthritis in one knee. At discharge her physical examination revealed only mild paresis of the left lower limb. The child improved further and was discharged on day 10 with a mild deficit in the left lower limb. Steroids were continued for persistent proteinuria and hematuria. At a cerebral examination 20 days after discharge there was no neurological deficit. As proteinuria persisted, after 2 months of steroid treatment the child underwent a renal biopsy, which showed a focal grade-2 sclerosis.

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