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C. C. Ng
S. C. Huang
L. T. Huang
S. C. Huang

Henoch-Schönlein purpura with intracerebral hemorrhage: case report

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C. C. Ng · S. C. Huang (✉) · L. T. Huang · S. C. Huang
Department of Pediatrics, Chang-Gung Memorial Hospital, Kaohsiung 123, Ta-Pei Road, Niao-Sung Hsiang, Kaohsiung Hsien, Taiwan, ROC

Abstract We report the case of a 5-year-old patient with Henoch-Schönlein purpura (HSP) who had a large intracerebral hematoma (ICH) in the left parietal area. HSP complicated by ICH is rare, with only three cases reported in the literature. Conservative treatment with steroid therapy in this patient was successful and no further major neurologic sequelae occured.

Introduction

Henoch-Schönlein purpura (HSP) is a systemic necrotizing vasculitis which is characterized by three major symptoms and signs: purpuric skin lesions, intermittent abdominal colicky pain, and large joint pain and/or arthritis. Bleeding from the gastrointestinal tract is common. However, massive intracerebral hemorrhage (ICH) is rare and we believe that this is the first case with cranial MRI [1–3]. We wish to report this case of HSP with ICH studied by cranial CT, MRI, and cerebral angiography.

Case report

A 5-year-old girl presented with a 2-day history of vomiting with coffee grounds and passing tarry/fresh bloody stools. Some ecchymoses were noted on the ears, eyelids, neck, and the lower legs in a symmetrical distribution. The patient's and her family's past histories were unremarkable. She complained of intermittent abdominal pain and bilateral knee joint pain. The results of coagulation studies were within normal limits. After admission, abdominal sonography was performed which revealed thickening of the bowel wall. Panendoscopy was done immediately, showing marked swelling and hyperemia with multiple areas of mucosal necrosis in the whole stomach and the duodenal bulb. The diagnosis of HSP complicated by erosive gastritis and duodenitis was made.

The patient received treatment with dexamethasone, histamine$_2$-blocker and antacids. However, she became progressively drowsy, spoke non-coherently and vomited. The neurologic examination showed muscle power of grade 1/5 in the right-sided extremities. The Babinski test on the right side was positive. An emergency brain CT was done which showed a large ICH in the left parietal area with rupture into the left lateral ventricle and the subarachnoid space (Fig. 1 a). Brain MRI (Fig. 1 b), as well as magnetic resonance angiography (MRA) and cerebral angiography were done on the 3rd and 5th day after admission, respectively. No evidence of an arteriovenous malformation was found. Considering the cortical involvement over the motor area and the rupture into the ventricle, which may have partly released the intracranial pressure, no surgical intervention was made.

The patient received conservative treatment and improved gradually during the 19-day hospitalization period. She was discharged with improved right-sided weakness (muscle power grade 3/5), total recovery of clear consciousness and no obvious cognitive deficit. She showed a minimally limping gait and full recovery of muscle power 2 months later. However, microhematuria and mild proteinuria developed gradually 1 month later.

Discussion

In HSP, massive bleeding is not uncommon, mostly in the form of gastrointestinal bleeding. However, massive intracerebral hemorrhage was rarely reported and the incidence is unknown. Clark and Fitzgerald reported one case of HSP with a large ICH where abnormal results were obtained in coagulation studies [1]. They attributed the coagulopathy to vitamin K deficiency. Scattarella et al. also reported one case of HSP with a large ICH, emphasizing the rapid diagnostic value of brain

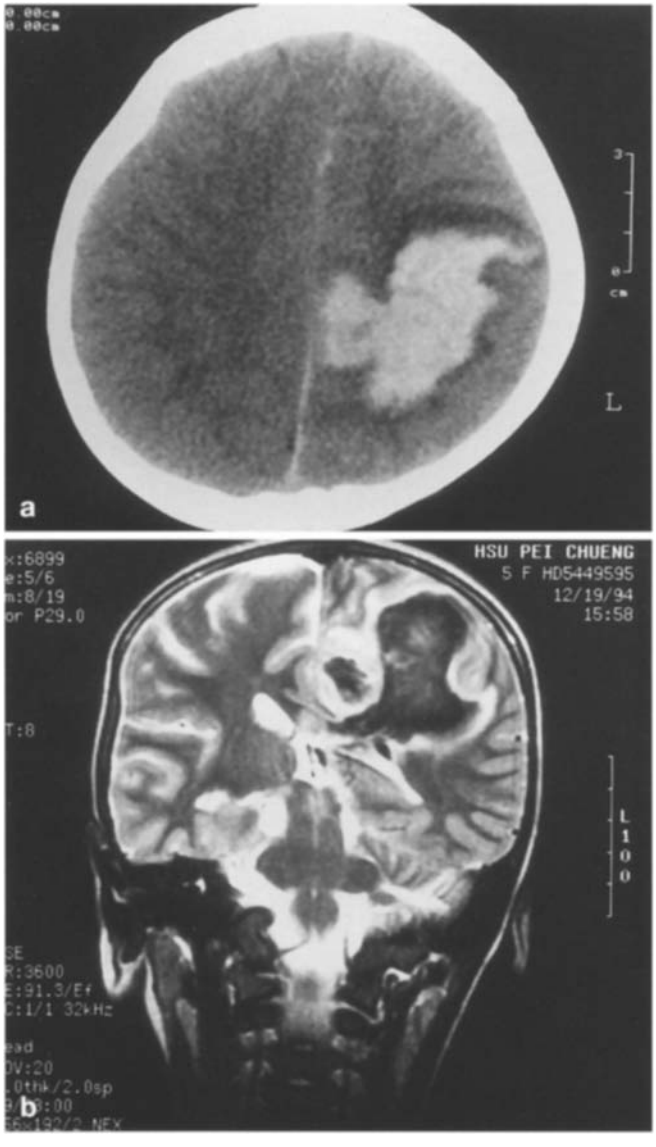

Fig. 1 **a** Non-enhanced axial brain CT scan showing a large hematoma in the left parietal area with rupture into the left ventricle. There was no midline shift (*L*, left; *R*, right). **b** Coronal T2-weighted (TR 3600/TE 91) MRI view of the brain showing a large hematoma in the left parietal area with rupture into the left ventricle and subarachnoid space

CT [2]. In one study of intracranial vasculitis by angiography and MRI, Harris et al. concluded that a negative MRI scan excludes intracranial vasculitis more definitively than a negative cerebral angiogram [4]. Accordingly, ICH shown on brain CT or MRI is one of the diagnostic criteria for intracranial involvement by systemic vasculitis. Therefore, the large hematoma shown in the brain CT of our patient indicates the diagnosis of intracranial vasculitis. To further prove this concept, brain MRI/MRA and cerebral angiography were performed within 2 days. There was no segmental narrowing or paradoxical dilatation of any intracerebral vessel either by MRA or cerebral angiography except for the mass effect over the left middle cerebral artery. Altinors and Cepoglu reported a case of HSP with a large ICH and midline shift which required emergency surgical removal because of rapid neurologic deterioration [3]. In contrast, surgical intervention was avoided in our patient because the rupture of the hematoma into the ventricle provided a route for relieving the pressure. Fortunately, our patient, as in the other three cases of HSP with significant ICH cited above, improved without major sequelae.

We speculate that early diagnosis and thus early steroid treatment might prevent the massive hemorrhagic complication of this immune-mediated necrotizing vasculitis. Since headache and non-specific acute behavioral changes are the most common neurologic manifestations in HSP, although found in only one third of the cases [5], these symptoms could obscure progression to intracerebral hemorrhage given the rare occurrence of this complication. In conclusion, in order to avoid this potentially fatal complication of HSP, we emphasize the importance of early diagnosis of intracerebral involvement of HSP. This is only possible by taking a careful history, monitoring any subtle neurologic manifestations, the use of CT/MRI rather than angiography for diagnosis, prompt treatment with steroids, and correction of any existing coagulopathy.

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