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CASES WITH A MESSAGE

Posterior reversible encephalopathy syndrome in Henoch–Schönlein purpura

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To the Editor,

Posterior reversible encephalopathy syndrome is an increasingly recognized clinicoradiological entity characterized by a variable association of headache, nausea or vomiting, altered mental status, seizures, visual abnormalities and motor deficits, classically accompanied by radiological findings showing a posterior-predominant pattern of bilateral gray and especially white matter edema [1]. It results from an endothelial and blood–brain barrier dysfunction with leakage of fluid into the interstitium, leading to cerebral edema.

Between 70 and 80 % of the cases develop after severe rapid increase in blood pressure that overcomes the cerebral vessels' autoregulatory mechanism with resulting hyperperfusion, capillary injury and edema [1]. Between 20 and 30 % of the cases develop in subjects with only moderately increased or even normal blood pressure. In these patients, endothelial and blood–brain barrier dysfunction develops in association with a widespread range of clinical conditions including systemic inflammatory diseases and infections, uremia, treatment with various drugs and "vascular" diseases such as hemolytic-uremic syndrome and systemic vasculitides [1].

Textbooks and reviews do not refer to the possible occurrence of posterior reversible encephalopathy syndrome in Henoch–Schönlein purpura, the most common systemic vasculitis in childhood. We aimed to systematically review the occurrence of posterior reversible encephalopathy syndrome in the context of this vasculitis. For this purpose, we performed a search of the terms ("posterior reversible encephalopathy syndrome" OR "posterior reversible leukoencephalopathy syndrome") AND ("Henoch–Schönlein purpura" OR "anaphylactoid purpura") in the US National Library of Medicine and Excerpta Medica database. Personal files were also screened. The diagnosis of Henoch–Schönlein purpura and posterior reversible encephalopathy syndrome were reviewed according to recognized criteria [1, 2].

Using this strategy, we found 17 rather well-documented cases of Henoch–Schönlein purpura that were complicated by posterior reversible encephalopathy syndrome [3–19]. The cases, published between 1990 and 2016 in English ($N = 15$), French ($N = 1$) and German ($N = 1$), had been reported from the following countries: Turkey ($N = 4$), Japan ($N = 3$), South Chorea ($N = 2$), Canada ($N = 1$), France ($N = 1$), Germany ($N = 1$), Greece ($N = 1$), India ($N = 1$), Spain ($N = 1$), the UK ($N = 1$) and the USA ($N = 1$). The characteristics of the 17 Henoch–Schönlein cases (9 ♂ and 8 ♀) associated with posterior reversible encephalopathy syndrome are listed in Table 1. Patients'

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Table 1 Characteristics of the 17 Henoch–Schönlein patients with posterior reversible encephalopathy syndrome

Characteristics of Henoch–Schönlein syndrome	
Cutaneous involvement	
Mild, *N* (%)	12 (70)
Moderate, *N* (%)	3 (18)
Severe, *N* (%)	2 (12)
Abdominal involvement	
Absent, *N* (%)	2 (12)
Mild, *N* (%)	4 (24)
Moderate, *N* (%)	5 (29)
Severe, *N* (%)	6 (35)
Articular involvement	
Absent	7 (41)
Mild, *N* (%)	5 (29)
Moderate, *N* (%)	4 (24)
Severe, *N* (%)	1 (6)
Renal involvement	
Absent, *N* (%)	10 (59)
Mild, *N* (%)	3 (18)
Moderate, *N* (%)	2 (12)
Severe, *N* (%)	2 (12)
Rash preceded by encephalopathy, *N* (%)	4[a] (24)
Blood pressure	
Normal, *N* (%)	5 (29)
Mild to moderate hypertension[b], *N* (%)	6 (35)
Severe hypertension[c], *N* (%)	6 (35)
Presentation of encephalopathy	
Seizures, *N* (%)	17 (100)
Altered level of consciousness, *N* (%)	12 (71)
Visual disturbances, *N* (%)	10 (59)
Nausea, *N* (%)	5 (29)
Palsy, *N* (%)	2 (12)
Full neurologic recovery, *N* (%)	16 (94)

The classification was performed according to the CAAR (cutaneous, abdominal, articular and renal involvement) grading scale [2]

[a] By 2, 4, 8 and 12 days

[b] Blood pressure > 95th percentile and ≤ 99th percentile + 5 mmHg

[c] Blood pressure > 99th percentile + 5 mmHg

age ranged from 5 to 38 years (median age 8 years; 2 patients aged >18 years). Six patients had been managed with oral prednisone (from 1 to 2 mg/kg day). None of the remaining patients had been treated with drugs that have been causally linked with this condition. Intriguingly, blood pressure was normal or moderately elevated in 11 out of the 17 patients (65 %). Finally, the encephalopathy sometimes preceded (≈20 %) the distinctive purpuric rash.

It is recognized that Henoch–Schönlein purpura may cause vascular lesions secondary to vessel wall proliferation with resultant luminal obliteration (or thrombotic occlusion) or hemorrhages secondary to vessel wall proliferation with rupture of necrotic walls [20]. The results of the present analysis indicate that this systemic vasculitis is sometimes associated also with posterior reversible encephalopathy syndrome. In Henoch–Schönlein purpura, this peculiar form of endothelial and blood–brain barrier dysfunction may result from severe rapid increase in blood pressure, poor renal function, drug management or directly from cerebral vasculitis.

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Compliance with ethical standards

Conflict of interest The authors declare no conflicts of interest.

Ethical approval This article is a systematic review of the literature. Therefore, it does not contain any studies with human participants performed by any of the author.

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