Case report

Post-varicella intra cranial haemorrhage presenting as young stroke

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Varicella-zoster vasculitis presenting with ischemic stroke is a known entity and has been described as one of the rare but important causes of stroke, especially in children[1,2]. But primary intracranial haemorrhage has been reported only in few cases. The child reported here had basal ganglia haemorrhage causing acute left hemiplegia two weeks post varicella.

Case report

A previously healthy 16-year-old female presented to our ward with acute left hemiplegia. Two weeks prior admission, the patient has developed an erythematous vesicular rash on her chest and abdomen which spread to her face, arms, and legs. The vesicular lesions on the trunk crusted week after appearance, consistent with varicella.

On examination her vital signs were normal, as is her mental status and cranial nerve examination. Motor strength was 0/5 in right upper and lower extremities, and 5/5 in the left with a mild left hemisensory deficit.

The initial computerized tomography (CT) of the patient's brain, performed few hours after admission, revealed a left basal ganglia haemorrhage without midline shift or mass effect (Fig. 1a). MRI done next day showed the same haemorrhage, but did not show any underlying structural cause for the bleed (Fig. 1b).

Laboratory investigations showed mild anaemia, normal white cell count, normal coagulation profile (prothrombin time, activated partial thromboplastin time and thrombin time), and normal erythrocyte sedimentation rate. But C-reactive protein levels were 14 mg/L (normal range 0-6 mg/L). Laboratory investigations did not show any bleeding diathesis. Her systemic vasculitis workup was negative for anti-nuclear antibodies, anti-double-stranded DNA, and rheumatoid factor. Echocardiography was also normal. Anti-VZV immunoglobulin M (IgM) was detected in blood but VZV immunoglobulin G (IgG) was not present. She did not have a history of any other systemic illness.

The patient was managed conservatively and patient was referred to physiotherapist and occupational therapist. Four weeks after the episode her weakness improved to a level where she was able to perform activities of daily living.

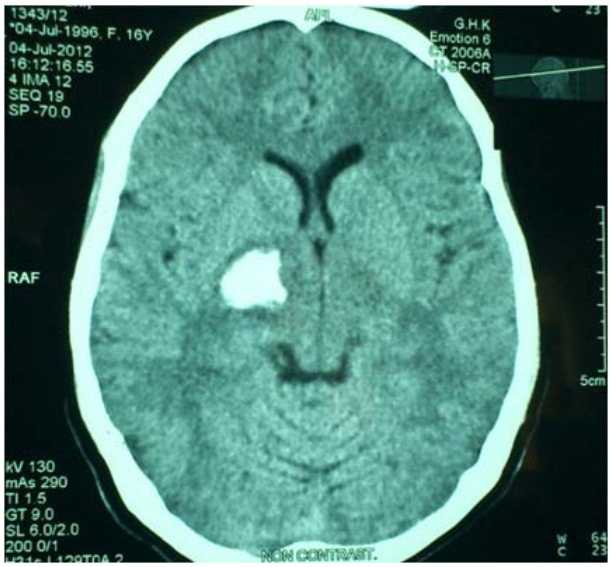

Figure 1(a). Axial non-contrast enhanced brain computerized tomography shows a left thalamic haemorrhage without midline shift or mass effect.

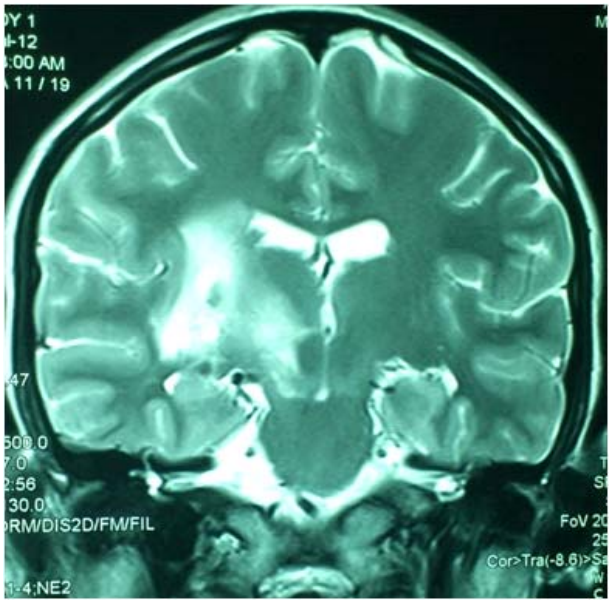

Figure 1(b). Coronal magnetic resonance imaging confirmed computerized tomography findings.

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Discussion

There are many possible causes of non-traumatic intra cranial haemorrhage, and at least one risk factor usually can be identified if a complete evaluation is done. A study (Table 1) of non-traumatic hemorrhage in children found at least one potential cause for the hemorrhage in children. These authors failed to identify risk factors in only 15% children whose evaluation included standard cerebral angiography[3].

The most common risk factors for any type of intra cranial haemorrhage were intracranial vascular anomalies, congenital heart disease, and brain tumors. These were excluded in our patient reported here.

But vasculitis of the intracranial vessels can be difficult to diagnose with confidence. Not all individuals have clinical or laboratory signs of inflammation and the classic angiographic findings of arteritis (arterial beading and alternating areas of constriction and dilatation) are nonspecific. Angiography is also not a freely available investigation as in our case.

Table 1. Nontraumatic brain hemorrhage in children: aetiology and presentation

	No of Children (N=85)				
Risk factor	*ICH*	*SAH*	*SDH*	*Total*	*% of children*
Brain tumor	11	1	1	**13**	15
Congenital heart disease with or without surgery or anticoagulation	6		8	**14**	16
Intra cranial vascular lesion	21	3		**24**	28
AVM	11			**11**	13
Cavernous malformation of the brain	7			**7**	8
Venous angioma	2			**2**	2
Aneurysm		2		**2**	2
Moyamoya syndrome (with sickle cell disease or Down syndrome)					
Infection	3	3		**5**	6
HSV encephalitis		1		**1**	1
Sepsis (thrombocytopenia)	2	1		**3**	4
Endocarditis	1			**1**	1
Leukemia or lymphoma (thrombocytopenia)	5	1		**6**	7
ARF, aplastic anaemia or LCHAD	4	1		**5**	6
Coagulation factor deficiencies	1		3	**4**	5
Genetic (e.g. Menkes syndrome)			1	**1**	1
Unidentified	10	2	1	**13**	15

Abbreviations: ARF, acute renal failure; **AVM,** arteriovenous malformation; **HSV,** Herpes simplex virus; **ICH,** intracerebral hemorrhage; **LCHAD,** long-chain 3-hydroxyacyl coenzyme A dehydrogenase deficiency; **SAH,** subarachnoid haemorrhage; **SDH,** subdural haemorrhage.

Over the past few decades there have been an increasing number of reports of vascular disease after VZV reactivation. Unlike early cases of acute hemiplegia after contralateral zoster caused by large-artery disease, the recognized clinical range of this disease has expanded to include transient ischaemic attacks and protracted illness involving both small and large arteries. In addition to ischaemic infarction, VZV can cause aneurysm, cerebral and subarachnoid haemorrhage, and arterial ectasia, and might be a co-factor, along with trauma, in the pathogenesis of cerebral arterial dissection. Furthermore, VZV can also cause peripheral arterial disease. In adults, the exact incidence of VZV vasculopathy is difficult to estimate, although it is more common in immunocompromised individuals. In children, VZV vasculopathy has been proposed to account for 31% of all arterial ischaemic strokes;[4] moreover, stroke was preceded by chickenpox in 44% of children with transient cerebral arteriopathy[5].

Brain imaging reveals abnormalities in most cases[6,7]. Abnormalities are cortical and deep, and occur in both the grey and white matter and at grey-white matter junctions in particular a clue to the cause of disease[8]. Although lesions of the grey and white matter are commonly seen in patients with other disorders such as metastatic carcinoma and embolic disease, VZV vasculopathy should be included in the differential diagnosis of patients with lesions at grey-white matter junctions. Most lesions are ischaemic, but haemorrhagic lesions also occur; some lesions enhance on MRI with contrast, indicating breakdown of the blood-brain barrier.

When a clinical diagnosis of VZV vasculopathy is suspected and is corroborated by single or multiple characteristic lesions on MRI or CT, virological confirmation is required.

The clinical diagnosis of VZV vasculopathy is strongly suspected when a patient with a recent history of varicella or zoster has a transient ischaemic attack or stroke corroborated by MRI abnormalities, particularly at the grey-white matter junction.

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