Adult post-varicella small vessel vaculopathy mimicking hypertensive basal ganglia hemorrhage with coexisting infarcts

Sir,

Virus induced vasculopathies are an uncommon but increasingly recognised causative factor of stroke and transient ischemic attacks (TIA) in children and adults. Varicella-zoster, human immunodeficiency virus (HIV) and cytomegalovirus are commonly associated viruses with stroke. While primary varicella infection is a common cause of neurological manifestations in children, secondary reactivation of varicella-zoster is common in immunocompromised adults. Both small and large vessels are involved often resulting in cerebral ischemia/infarct. Rarely, a subarachnoid haemorrhage, with or without an intraparenchymal component, has been reported. We report a rare case of adult post-varicella vasculopathy involving
small vessels resulting in a co-existing infarct as well as basal ganglia haemorrhage, mimicking a hypertensive bleed.

A 30-year-old male patient presented with a history of sudden onset of severe headache, left sided weakness, slurring of speech, and facial deviation to the right side. He was taken to a local hospital, a computed tomography (CT) scan was done, and he was diagnosed as a case of right basal ganglion bleed. The initial medical management was performed in a local hospital and then he was referred to our centre for further management. He and his younger sister, aged 25 years, had a history of chickenpox one month prior to the ictus. During the course of the infection, he had not developed any central nervous system manifestations.

His blood pressure was elevated at the time of ictus; however, when he came to our hospital after 20 days of ictus, his blood pressure was normal. He had no past history of hypertension or diabetes mellitus. Both the patient and his sister were having the typical, multiple small scar-associated hyperpigmentation at sites corresponding to the previous varicella rash on the face and trunk. His neurological examination revealed left hemiparesis with left sided mild facial palsy. He had a significant improvement in his limb weakness from the time of his ictus until his presentation to our hospital. His blood investigations revealed a normal total leukocyte count with mild neutrophilia, an erythrocytic sedimentation rate of 25 mm/hr, and his HIV test was negative. His platelet count, prothrombin time, and activated partial thromboplastin time were normal. His magnetic resonance imaging (MRI) showed a subacute intraparenchymal hematoma in the right external capsule-putaminal region [Figure 1 a-d]. An acute infarct was seen involving the right lentiform nucleus and caudate nucleus [Figure 1 e and f]. Contrast MR image with dedicated protocol to look for large vessel wall enhancement was negative for vasculitis [Figure 1 g-i]. Another acute lacunar infarct was also seen in the right cerebral peduncle [Figure 1 j-l]. Time-of-flight MR angiography revealed mild diffuse attenuation of the right M1 segment of middle cerebral artery (MCA) possibly due to mass effect from the haematoma/edema. Rest of the intracranial arteries were normal in calibre. The patient was reluctant to undergo an invasive digital subtraction angiography procedure due his improving clinical condition. He was admitted and medically managed along with neurorehabilitation. A short course of steroids and acyclovir injections were given considering the presumptive diagnosis of small vessel varicella vasculitis. He showed significant improvement in his symptoms and became ambulant over a period of next twenty five days. He showed further improvement in his neurological condition during one month of follow up.

Varicella-zoster vasculopathies are due to virus infection of cerebral arteries either during the primary infection (chickenpox) or during subsequent reactivation. The spectrum of vascular pathologies include ischemic infarctions, aneurysm formation, arterial dissections, subarachnoid haemorrhage or intraparenchymal haemorrhage. Both large and small arteries are commonly involved, and less often, an exclusive small or large vessel disease is seen. Presence of a large artery disease is diagnosed by the characteristic segmental constriction or post stenotic dilatation visualised on the performance of a digital subtraction angiogram (DSA) or an MR/CT angiogram. Demonstration of direct vessel wall enhancement by high quality post contrast MRI can also suggest the diagnosis. However, direct demonstration of small vessel involvement is difficult as an angiogram is often negative in this situation. In such cases, the diagnosis is based on cerebral parenchymal changes on MR imaging.

The common location of the infarction in varicella-zoster vasculopathy is the gray-white matter junction. The infarcts in present case were in the right lentiform nucleus and head of caudate nucleus, in the lenticulostriate artery territory. A lacunar infarct in the right cerebral peduncle was also present. Both the infarcts were suggestive of small vessel vasculopathy. Both these locations are uncommon sites for the varicella small vessel vasculopathy, though they have previously been described in the literature. MR angiogram did not reveal a large vessel vasculopathy. The temporal sequence of the initial varicella infection followed by the appearance of neurological symptoms one month later were in accordance with the typical natural course of varicella vasculopathy.

Subarachnoid haemorrhage (SAH) is more common in the varicella-induced vasculitis than intraparenchymal hemorrhage. The previously reported intraparenchymal hemorrhage was in lobar location and was associated with SAH. However, the reported case had secondary reactivation of the varicella-zoster virus in adult life, and was not due to a primary varicella infection. The other case described by Danchaivijitr et al. was that of a 7-month old child, in whom the location of the bleed was in the interhemispheric region and in the subarachnoid space, without a significant intraparenchymal component. In both the cases, the possible vessel involved was a large artery or a small lobar artery. In the present study, the location of the bleed was in the right putamen-external capsule, which was secondary to lenticulostriate artery involvement. The location of the bleed in our case is typically associated with hypertensive
bleed. Our article is one of the first reports to describe an adult patient with a primary varicella affliction developing an intraparenchymal bleed that was co-existing with an infarct attributable to small vessel vasculopathy. The presumed mechanism of bleed appear to be extensive vessel wall inflammation and aggressive tissue viral invasion associated with necrotizing angitis. It cannot, however, be unequivocally stated that the same pathogenetic process led to one small artery undergoing necrosis and causing a hemorrhage, and the other smaller artery undergoing occlusion and resulting in an infarct.

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Conflicts of interest
There are no conflicts of interest.

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