Sudden death in a case of lateral medullary syndrome

Sir

We report a case of sudden death in a case of lateral medullary syndrome (LMS). Sudden unexpected death is an unusual event in LMS.[1–2] A 39-year-old male, a chronic smoker, presented with vertigo, dysphagia, hoarseness of voice, and imbalance while walking of 6 hours duration. On examination, his blood pressure was 140/90 at admission and the neurological examination revealed sensory loss on the right side of the face and left half of the body with right-sided cerebellar signs, conforming to right LMS. A non contrast computed tomography (CT) scan showed an infarct in the right inferior cerebellum and a CT angiography showed non visualisation of the right vertebral with a thrombus extending into the proximal basilar artery [Figure 1]. The proximal right vertebral artery was well visualised in the CT angiography. Considering the risk of progression to complete basilar artery occlusion, the patient was taken in for an intra-arterial (IA) thrombolysis after full informed consent was obtained. A selective right vertebral artery catheterisation was done. A 5 mg bolus of r-tPA was injected over a period of 1 min. This was followed by a slow infusion of 20 mg r-tPA. However, after an infusion of 4 mg of r-tPA after the bolus, the patient had a sudden cardiorespiratory arrest. He succumbed despite resuscitative measures. A non contrast cranial CT scan was repeated, which did not show any hemorrhagic transformation.

Recent reports have described unexpected sudden cardiorespiratory arrest in lateral medullary infarction during convalescence after a stroke with minimal motor disability.[3–5] Various mechanisms have been postulated for the sudden cardiorespiratory arrest in LMS including cardiac arrhythmia[3] and ischemic penumbra affecting the cardiac and respiratory centers of the medulla. [4] A recent neuropathological study of ßve patients disclosed ischemic lesions in the solitary tract nuclei of the medulla after subacute hypoperfusion of the brain during acute heart failure.[6] It was speculated that these medullary lesions caused autonomic instability, which precipitated death. In our case, a sudden unexpected cardiorespiratory arrest occurred during IA thrombolysis in an otherwise haemodynamically stable patient. The mechanism of the arrest could not be ascertained since monitoring could not be done during the procedure. Sudden death in the presence in LMS is often puzzling and mandates close monitoring of the cardiac and respiratory functions in patients with LMS.

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Parenchymal brain cysts in Schimmelpenning–Feuerstein–Mims syndrome

Sir,

A 10-year-old girl presented with verrucous epidermal
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nevus over right half of the face and neck [Figure 1], recurrent complex-partial seizures from six years of age, and poor scholastic performance. The skin lesions evolved with age from pink to yellowish-brown color. She also had limbal dermoid and the rest of the examination including skeletal system was normal. Magnetic resonance imaging (MRI) of the brain revealed right-sided hemiatrophy with parenchymal cysts in the deep parietal white matter [Figure 2]. The combination of characteristic verrucous skin lesions, ocular findings and the hemiatrophy on MRI are diagnostic of Schimmelpenning–Feuerstein–Mims syndrome (SFM) syndrome.

SFM syndrome is a rare neurocutaneous disorder with clinical heterogeneity. The disorder consists of a triad of linear sebaceous nevus, seizures and mental retardation. Central nervous system (CNS) involvement occurs in the form of hemimegalencephaly with ipsilateral gyral malformations, seizures, hemiparesis, mental retardation, cranial nerve palsies and hydrocephalus. Neuroimaging findings in SFM syndrome include the primary lesions: Malformations of cortical development like hemimegalencephaly, polymicrogyria; gliomatosis, hemiatrophy with or without parenchymal cysts, vascular malformations and intracranial/intraspinal lipomas and secondary features: Porencephaly, infarcts and atrophy. Other rare findings include enlarged ventricles, cortical hypoplasia, intracerebral calcification, arachnoid cysts, intracranial tumors and skull asymmetry. CNS complications are more likely to be associated with epidermal nevi on the head and the CNS abnormalities are most often ipsilateral to the skin lesions. Though hemiatrophy has been reported before in SFM, parenchymal cysts in brain have rarely been reported in SFM syndrome. There is mention of porencephalic cysts in the literature but no report of parenchymal cysts like the ones seen in our case.

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