stroke, dementia, and hearing loss before the diagnosis and the introduction of ERT. Three FPs (one male, two females) have asymptomatic multiple small white matter lesions on MRI scans. Two FPs (one male, one female) have increased cIMT. Normal, tortuosity-free cerebral arteries were detected by MRA.

Conclusion: Neurological complications in FD are common, complex and may be devastating, as ischaemic stroke. Early diagnosis is very important to introduce ERT as soon as possible. The long-term follow-up and treatment of patients are elementary in FD to prevent complications.

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Cerebral venous thrombosis in a patient with localised scleroderma

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Introduction: Localized scleroderma has a prevalence of less than 3/100,000. It is characterized by sclerotic lesions of skin and tissue underneath. It distinguishes from systemic forms by its limited development and it was thought that it had lack of internal organ involvement. Today it is known that, even in localized presentations, neurologic involvement is possible.

Clinical case: Twenty-nine years old woman, with history relevant for migraine-like headache with 4 years evolution and one generalized tonic-clonic seizure the year before. She had been admitted after headache worsening in the past 3 weeks and brain MR revealed several bilateral T2 hyperintense lesions with contrast captation. She was transferred to our department after developing a focal status epilepticus (left motor seizures). She was started on valproate. A new brain MR showed images compatible with sagittal sinus venous thrombosis, and initiated IV heparin with progressive clinical and image improvement, and no further epileptic seizures. Her EEG (performed after seizure cessation had a centro-temporal right dysfunction. Her physical examination was relevant for a limitation of right eye adduction and referred diplopia since 1 year before. There were a right frontotemporal and a parietal cutaneous-muscular atrophy regions with 7 years evolution. A en-coup-de-sabre form of localized scleroderma was diagnosed by Dermatology Department. The CSF analysis was irrelevant for infection or inflammation. No thrombotic disturbance or risk factor was found. After steroid treatment, brain inflammatory lesions regressed.

Conclusion: The inflammatory lesions found on MRI, the history of chronic headache and seizures and oculomotricity limitations could be associated with neurologic manifestations of localized scleroderma described in literature. No reference to cerebral venous thrombosis with localized scleroderma was found, although, questions about a possible manifestation of a cerebral vasculopathy, documented in histologic studies could be raised.

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Ischaemic stroke associated to acute cocaine and amphetamine abuse: a case with angiographic documented intracranial artery stenosis

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Objective: Cocaine and Ecstasy have been associated to ischemic stroke, but this relationship is best established in the case of cocaine. However the exact mechanism of cocaine related ischemic stroke is not completely understood. We aim to report a case of an ischemic stroke after cocaine and ecstasy consumption with a symptomatic posterior cerebral artery (PCA) stenosis.

Case report: A 28-year-old, male, with sporadic drug use, was admitted due to right visual field visual loss, noticed immediately after cocaine and ecstasy consumption. Observation confirmed a right homonymous hemianopsia. Brain MRI and angioMRI disclosed an ischemic stroke in the left posterior cerebral artery territory and irregularity of P1 and P2 segment of same artery. The transcranial Doppler (TCD) showed a marked increase in flow velocity in the left PCA. Cerebral angiography confirmed slight irregularity at the P1 segment and progressive reduction in the size of the P2 segment of left PCA with occlusion of the distal arteries. TCD performed 2 months after the event showed a persistent left PCA stenosis. Cardiological evaluation disclosed a small patent foramen ovale. Prothrombotic, vasculitis and infection screening was unremarkable.

Discussion: In this case temporal coincidence between ischemic stroke and cocaine and ecstasy consumption suggests a causal relation between the two events. Besides an irregular stenosis of left PCA was documented by angiography. This localized arterial lesion could be related to vasospasm or direct injury, induced by drug abuse. Cocaine related cerebral vasospasm has been documented. However there are only few report documenting intracerebral arteries abnormalities, by cerebral angiography, in patients with acute drug-induced ischemic stroke.

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Traumatic isolated basilar artery dissection

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Objectives: Primary or isolated dissection of basilar artery, spontaneous or traumatic, is a rare finding in stroke, and in most reports extracranial or vertebral arteries are involved. Magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) increased the recognition of dissections of intracranial arteries.

Methods: Case report.

Results: We report a 46-year-old man with locked in syndrome due to multiple ischemic infarcts in the brain stem, pons, thalamus, cerebellum and occipital lobes, due to traumatic dissection of the distal basilar artery from a minor head trauma that caused a linear stress fracture of the posterior wall of the sphenoid sinus, in close vicinity to the basilar artery. Diagnosis of dissection was made on the basis of MRI and MRA findings, and was confirmed by exclusion of other risk factors, as well as the documentation of the resolution of the basilar artery flow abnormalities with repeat MRA, 30 days after ictus.

Conclusion: This case is remarkable for the absence of dislocation of anatomic structures, the lack of relevant cranial injuries and the fact that extracranial or intracranial vertebral arteries were not involved.

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Right internal carotid artery occlusion in a previously healthy young woman: cryptogenic stroke?

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