CASE OF THE MONTH

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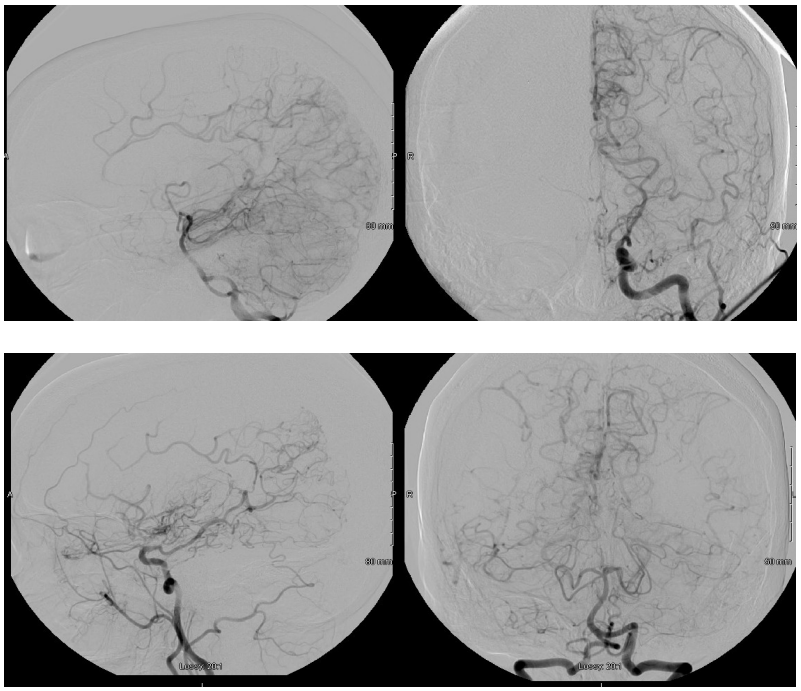
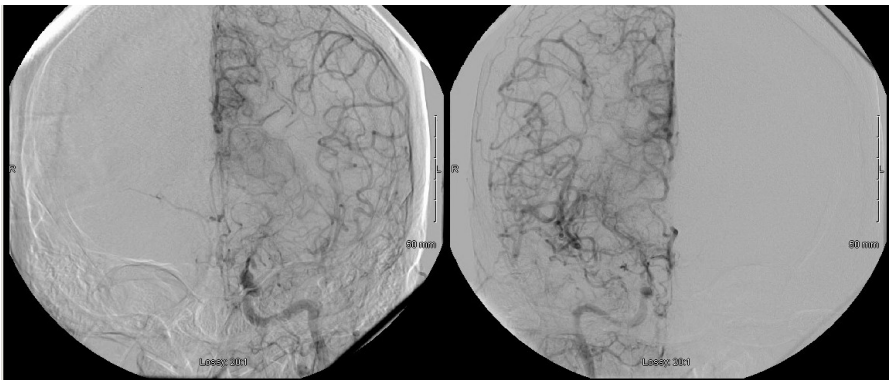
COCAINE INDUCED MOYAMOYA

Moyamoya is a disease that was first reported in Japan, in the 1960s. It is a rare and progressive disorder that affects the arteries at the base of the brain, leading to the occlusion of the distal internal carotids and their major branches, including the proximal segments of the middle and anterior cerebral arteries. The stenosis that occurs leads to poor blood flow to those areas that each artery serves. A "hazy puff of smoke" is the characteristic appearance on imaging studies, resulting from the irregular perforating vascular networks found near these stenotic vessels.

CASE PRESENTATION:

A 36 year old African American female presented to Truman Medial Hospital with complaints of aphasia, right-sided weakness, dizziness and nausea over the past 24 hours. Her past medical history was significant for hypertension, depression, alcohol abuse, polysubstance abuse and noncompliance. Medications included albuterol, hydrochlorothiazide, lisinopril, ibuprofen and promethazine. Family history was unremarkable. Physical exam revealed a BP of 183/120. Neurological examination was remarkable for a right-sided facial droop, 0/5 strength in both right upper and right lower extremities and an expressive aphasia. A urine drug screen was positive for cocaine and marijuana. A non-contrast CT of the head showed multiple ill-defined hypodense areas in the frontal lobe; a followup MRI of the head and neck revealed acute infarcts in the left precentral gyrus cortex and scattered throughout the left centrum semi ovale. MRA of the head showed markedly small bilateral anterior cerebral arteries and middle cerebral arteries past the bilateral cavernous carotids, along with irregularity and drop out of signal in the proximal bilateral posterior cerebral arteries. A transthoracic echo, obtained to rule out an embolic stroke, was normal. A hypercoagulable workup did not reveal any abnormality. A 4-vessel cerebral angiogram (images below), performed at St. Luke's Hospital, in Kansas City, was consistent with Moyamoya Disease; the latter was thought to be secondary to chronic cocaine use. The patient was started on ASA 162 mg by mouth daily and was transferred to an outpatient rehabilitation center.





DISCUSSION:

Cases of Moyamoya have been described throughout the world, including Africa, Europe, Australia and the United States. Recent studies showed that this disease is more common in the Japanese population, with a prevalence of 10.5 per 100,000 patients and a bimodal age distribution in the first and fourth decades. Four categories of Moymoya have been described, including ischemic, hemorrhagic, epileptic and "other." Ischemic symptoms predominate in the young while hemorrhagic symptoms predominate in the elderly. The most common cause of death in Moyamoya patients, demonstrated in autopsies, is intracerebral hematoma. The classic presentation of this disease is multiple attacks of weakness, paralysis or seizures. Less common manifestations include visual disturbances, altered consciousness and abnormal speech.

It is unclear what factors predispose to this condition. One theory postulated that there is a genetic susceptibility that results from mutations in chromosome 17. As in our case, some reports have linked the disease to chronic cocaine abuse; cocaine induces spasm of the cerebral vessels which, over time, leads to the formation of irregular collateral networks that characterize Moyamoya. In addition, prolonged cocaine use may cause endothelial injury, exposing damaged vessels to enhanced platelet activity with subsequent acute thrombosis.

While milder cases of Moyamoya may be treated conservatively, patients who experience severe symptoms may need surgical correction. There are three types of surgical procedures utilized in treating this condition: indirect, direct and combined bypass. In the indirect procedure, healthy, new vasculature from adjacent tissue are redirected to ischemic zones. The direct procedure involves the creation of a middle cerebral artery-superficial temporal artery bypass. If chronic ischemia persists after either procedure, a combination of these interventions may be attempted.

CONCLUSION: This case highlights one of the less common complications of cocaine abuse. It also emphasizes the importance of considering Moyamoya Disease in young patients who have recurrent strokes. Moreover, it demonstrates the diagnostic value of cerebral angiography in Moyamoya disease, revealing the stenotic vessels and new collaterals that characterize this condition. As technology progresses and imaging modalities become more sensitive, more patients might be diagnosed with this disease.

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