INTRODUCTION:
Moyamoya disease (MD) is a rare genetic condition associated with stroke in younger people.

OBJECTIVE:
Report a clinical case about MD with parkinsonism, altered mental status and bilateral pyramidal syndrome as initial symptoms.

METHOD:
OSS, male, 47 years old, hypertensive, on February 2015 presented sudden onset of hemiparesis, hemiparesis and parkinsonism on the left side, being admitted at the Hospital de Clínicas-Unicamp on March 25th. Initial investigation with non-contrast CT scan showed signs of stroke in the territory of the middle right cerebral artery (MRCA) being treated with AAS and statin. He returned on April 2015 with acute mental status deterioration and previous neurological deficits. On admission, he was disoriented, with bilateral tremors, more evident on the left, and cogwheel rigidity on the left arm. Electrocardiogram, transthoracic and transesophageal echocardiograms, spinal fluid analysis, EEG and other laboratorial exams showed no significant changes. Further investigation, involving a brain MRI and angiography showed bilateral occlusion of internal carotid arteries (ICA), intense collateral circulation of the vertebrobasilar system, and external-internal circulation through middle meningeal artery. Areas of terminal stenosis in ICA, anterior and MRCA, as well as proliferation and enlargement of the lenticulostriate arteries in the basal ganglia region, suggested MD diagnosis.

RESULTS AND DISCUSSION:
MD is a genetic disease defined by occlusive process intracranial arteries with formation of collateral vessels. This is a non-inflammatory and non-atherosclerotic condition and usually preserve the posterior circulation. The typical clinical presentations are: transitory ischemic attack, stroke, headache, seizures, and cognitive deficits. Movement disorders are uncommon initial presentation and can have some response with the steroids use, according to few studies.

CONCLUSION: MD is an uncommon cause of stroke and is related mainly with ischemic and hemorrhagic events in younger people. Movement disorders are uncommon and can be related to silent previous strokes.

BIBLIOGRAFIA