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## Introduction

Haemoglobinopathies represent a frequent disease in the African continent, as well as the most common recessive inherited disorder in Mediterranean population. It could cause cerebrovascular diseases even in younger subjects. We describe the case of a 43-year-old Nigerian man who came to our observation with a cerebral haemorrhage with a sickle cell trait.

## Case presentation

The patient was admitted to our ED for a loss of consciousness with acute onset. A CT brain scan showed an hemorrhagic stroke in left nucleo-capsular region (fig. 1a), while a CT angiography of intracranial vessels was unremarkable. Since the arrival in ED, the patient revealed extremely high blood pressure levels, and a multiple antihypertensive therapy was set, with poor response. At the admission to our Stroke Unit, the neurological examination showed a right facial droop with a right hemiparesis; there was no sensory deficit, plantars were downgoing bilaterally. We performed a CT scan above 24 hours from the onset, that showed a left frontal-basal hypodensity, not present at the previous images, compatible with a cerebral ischemia (fig. 1b).

Blood pressure continued to be unresponsiveness to therapies, so the 24-hour urine metanephrines test was performed, and revealed an high normetanephrine level (2.835 mcg/24h, n.v. 88-444). An abdominal MRI was performed (fig. 2). Doxazosin at the dosage of 2 mg twice daily led to a drastic reduction in pressure values, stabilized at 120/80 mmHg.

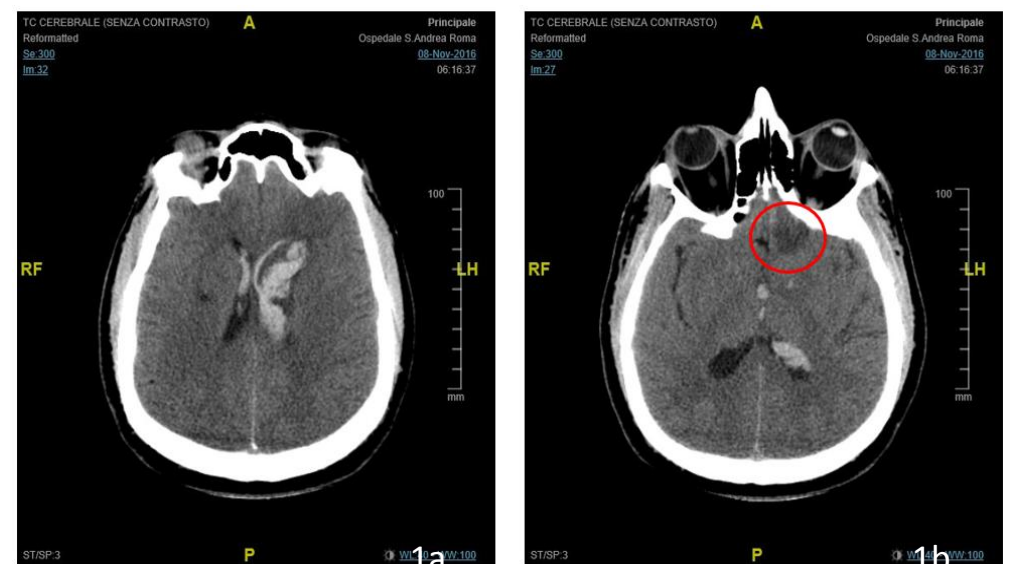


Fig. 1a Haemorrhagic stroke in left nucleo-capsular region; 1b Hypodensity in the fronto-basal region

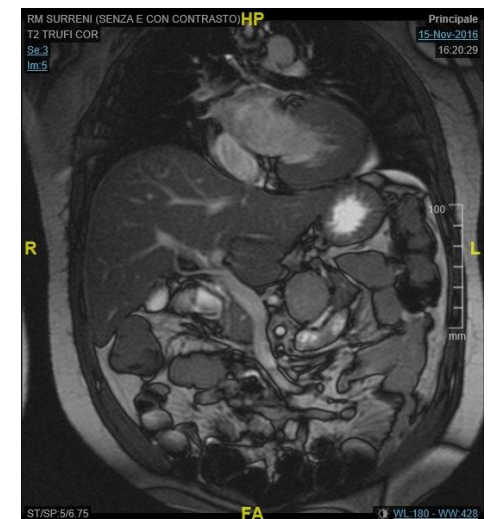


Fig. 2 Abdominal MRI showing in the left adrenal loggia the presence of a nodular formation of 53x38mm.

- The patient showed a worsening anaemia, associated to a diffuse limb pain and high d-dimer level (39.534 ng/ml, n.v. <250). A CT chest scan showed a thrombotic pulmonary embolism, in absence of lower limb doppler ultrasound alteration. A LMW heparin therapy was started with improving of symptomatology.
- The electrophoresis of haemoglobin showed the presence of **Hb S** with a value of **30.6%**. Considering the clinical status and the previous events, we decided to treat the patient with hydroxyurea at the dosage of 500 mg daily. One month later, the patient could undergo to a laparoscopic surgical operation with mass removal. The histology showed an isolated and diffused extrasurrenal abdominal paraganglioma, positive for chromogranin and synaptophysin, with no characteristics of malignancy. The patient was dismissed few days later, with no neurological deficits and a normalized blood pressure.

## Conclusion

The study of pathogenic variants of haemoglobin should be routine in both juvenile ischemic and haemorrhagic cerebrovascular diseases.