



# Hemiplegic migraine as presenting symptom of Evans syndrome: a case report

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## Background

**Hemiplegic migraine (HM)** represents a rare form of migraine with aura presenting with episodes of transient motor weakness/hemiparesis<sup>1</sup>.

**Evans syndrome<sup>2</sup> (ES)** is an autoimmune condition that presents with two or more cytopenias, which include autoimmune haemolytic anaemia and thrombocytopenia.

**Aim:** We report a case of ES with episodes of HM as symptoms of onset.

## Case presentation

14-year-old woman, non-contributory personal and family history for hemiplegic migraine

**Three episodes** occurring in one week:

gradual development of **scintillating scotomas**, **paraesthesia on left arm**, **left hemiparesis** and **speech disturbance**, followed by a **migrainous headache**.

Each symptom lasted approximately 10 min and was fully reversible.

## **First-line diagnostic investigations**

- CT brain excluded ischemic/haemorrhagic stroke
- Haematochemical tests revealed anaemia (haemoglobin 6,2 g/dL), with increased LDH (1148 U/L), thrombocytopenia (136000/mm<sup>3</sup>) and reticulocytosis

## **Clinical findings**

A fourth episode of transitory left hemiparesis during admission to the hematology unit. Symptoms fully recovered (2h) and neurological examination was normal (NIHSS=0).

## **Second-line diagnostic investigations**

**Urgent multimodal brain MRI during neurological symptoms** excluded acute stroke, showing on the right hemisphere reduced flow signal representation at distal arterial branches A2/A3, M2/M3/M4, P3/P4 associated with perisylvian cortical venous congestion, and a concomitant hypoperfusion on the entire right hemisphere compared to contralateral hemisphere, as seen on pCASL sequences (**fig.1**).

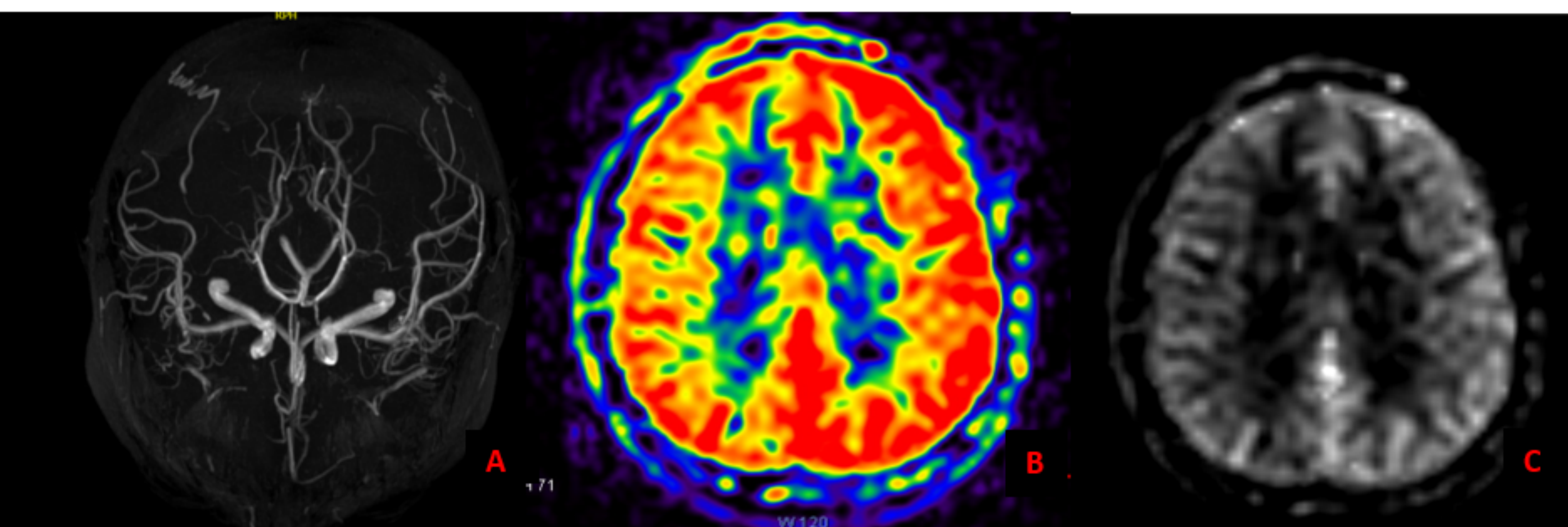


fig.1 MRA (A), pCASL(B) and SWI sequences

## **Therapeutic intervention**

On suspicion of an immune-mediated haemolytic process (Coombs test positive) it was started therapy with IV immunoglobulin (0,5g/kg/die for five days), IV methylprednisolone (2,5mg/kg/die) and blood transfusions.

## **Follow-up and outcome**

Causes of secondary ES were ruled out. After starting specific therapy for ES, the patient no longer presented episodes of HM.

## Conclusions

At best of our knowledge, it is the first case of ES with episodes of HM as presenting symptoms. ES should be considered a cause of new-onset HM.

## **References:**

<sup>1</sup> Headache Classification Committee of the International Headache Society (IHS) The International Classification of Headache Disorders, 3rd edition. Cephalalgia. 2018; 38: 1–211.

<sup>2</sup> Shaikh H, Mewawalla P. Evans Syndrome. In: *StatPearls*. Treasure Island (FL): StatPearls Publishing; June 12, 2023