DEMENTIA AND OLIVOPONTOCEREBELLAR ATROPHY: AN UNUSUAL PRESENTATION OF DURAL ARTERIOVENOUS FISTULA

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BACKGROUND: INTRACRANIAL DURAL ARTERIOVENOUS FISTULAS (DAVFS)

DAVFs are pathologic dural-based shunts, account for 10%–15% of all intracranial arteriovenous malformations.
Highly variable natural history and symptomatology, depending on the lesion location and its venous drainage pattern.
➔ DEMENTIA, isolated or associated to other neurological symptoms, is a rare but possible manifestation of DAVFs, and is a potentially reversible condition.

HEMODYNAMICALLY, DAVFs can determine **widespread venous hypertension** causing diffuse ischemia and/or progressive dysfunction of brain parenchyma, with well known neuroradiological and pathological patterns.

CASE PRESENTATION

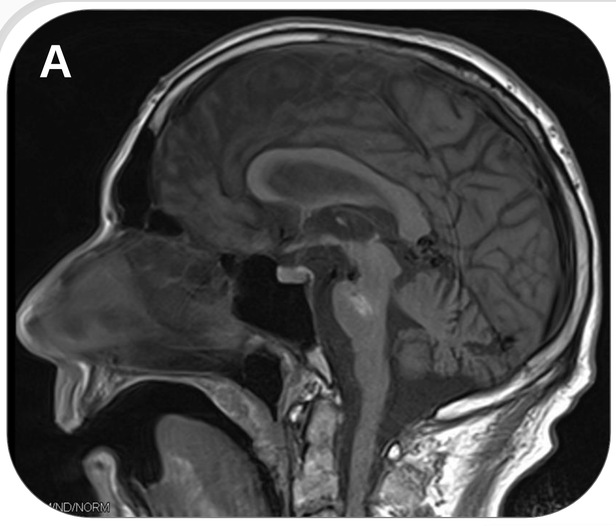
A 63 years old man developed **progressive multidomain cognitive decline, personality changes, urinary incontinence about two years before admission**.
The condition was initially diagnosed as cerebellar multisystem atrophy (c-MSA) for the relief, at a previous brain MRI, of olivopontocerebellar atrophy (a vascular malformation, already visible, was not diagnosed). The patient medical history was unremarkable except for arterial hypertension and prostatic benign hypertrophy.
For progressive worsening of his symptoms, he was admitted at our neurology unit.
AT OUR VALUTATION: the patient showed clear ideomotor slowdown, a partial temporo-spatial disorientation, severe reduction in language fluency, perseveration, attention deficit, motor impersistence , frontal ataxic gait, presence od subcortical reflexes, ideomotor, ideative and constructive apraxia, dysgraphia, right-left confusion and digital agnosia

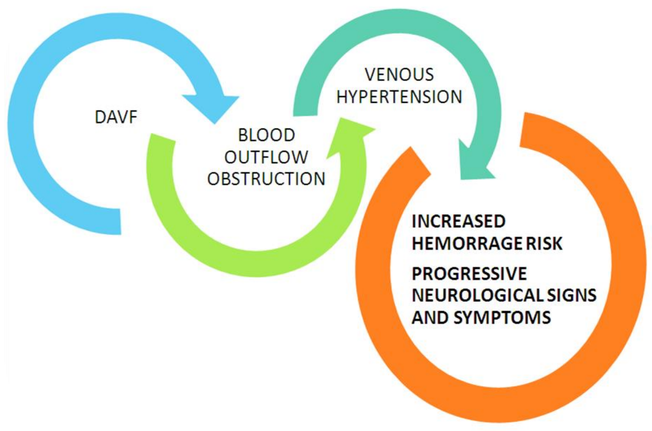
NEURORADIOLOGICAL AND LABORATORY FINDIGS

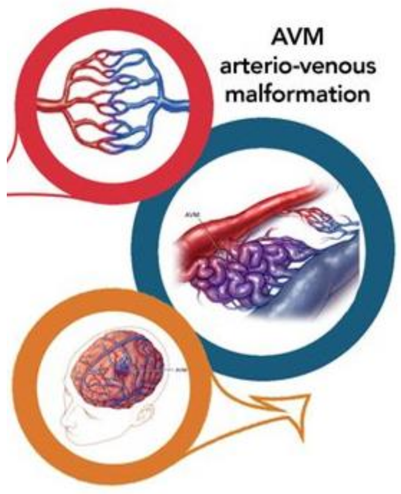
MRI ANGIOGRAM, **followed by** DIAGNOSTIC DIGITAL SUBTRACTION CENTRAL ANGIOGRAPHY (*Figures A-B-C-D*), revealed a III Cognard degree DAVF sited between the splenium of callous body, the quadrigeminal lamina and tentorium. This DAVF determined a severe, predominantly subcortical and left-sided, venous hypertension even involving left hippocampal and insular veins. It was moreover appreciable a consistent subcortical venous ectasia (left Rosenthal basal vein mostly) responsible for the involutive signs of pons, cerebellar hemispheres and peduncles (calcifications and atrophy).

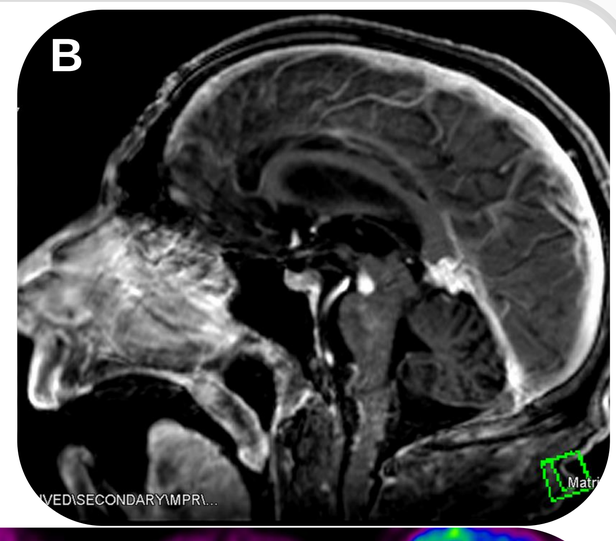
LUMBAR PUNCTURE revealed elevated proteins rate in CSF (0.99 mg/dL) as an indirect sign of the altered mechanism in venous and CSF reabsorption

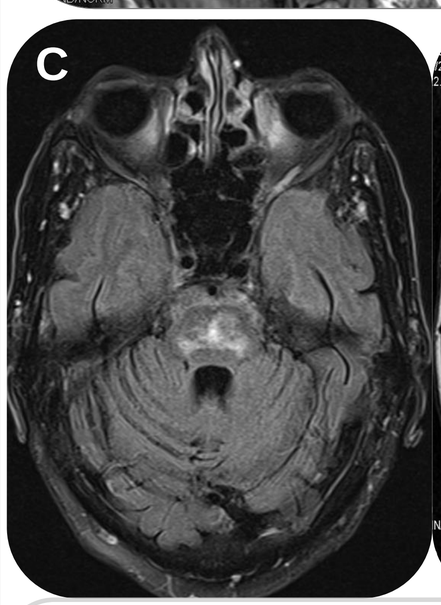
18-FDG CEREBRAL PET IMAGING (*figure F*) showed diffuse cortical-subcortical mostly left sided hypometabolism, with a strict connection with the areas involved in the venous hypertension circuit.

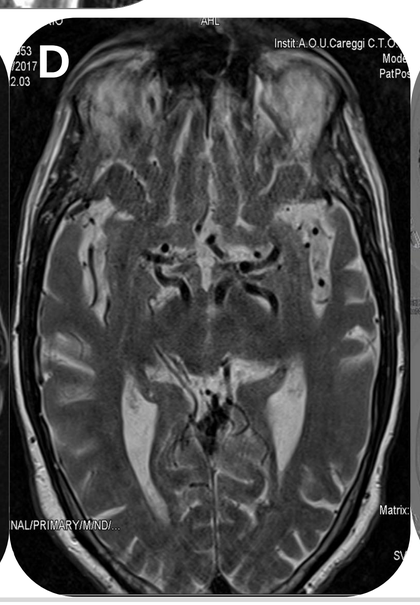


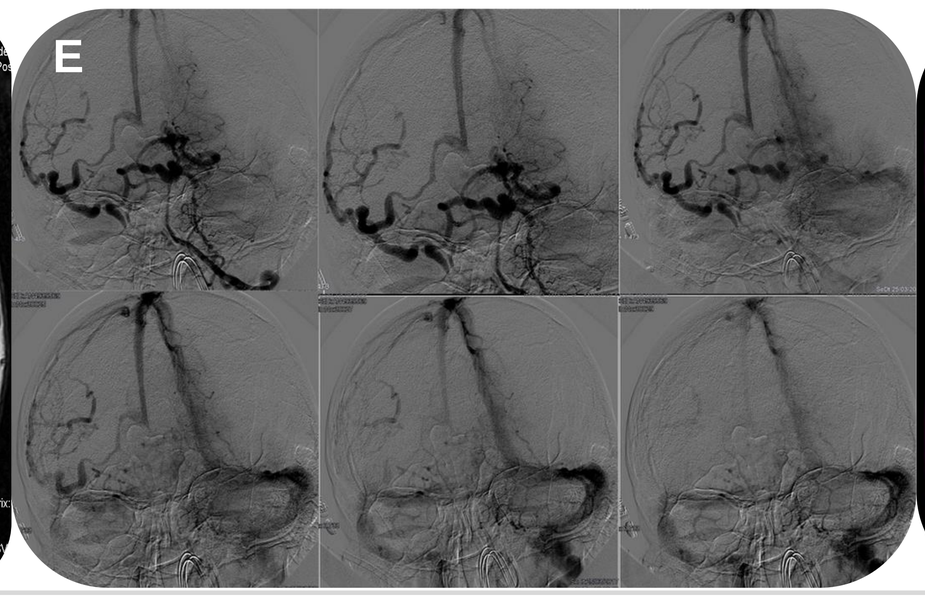


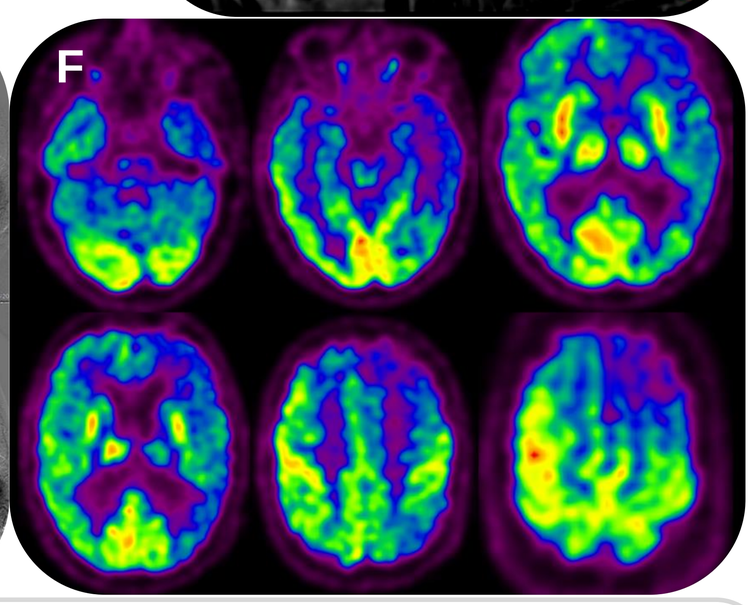












FIGURES: *A* **(T1 MR IMAGING)** *and C* **(T2 FLAIR MR IMAGING)** showing involutive pontine lesions and enlarged cerebellar cortical *sulci; B)* **T1 MR IMAGING WITH GADOLINUM** enhancement highlights the DAVF (between the splenium of callous body, the quadrigeminal lamina and tentorium) and the ectasic left Rosenthal vein; **D) T2* MR IMAGING**: ectasic aspect of basal veins, in particular of left Rosenthal vein on mesencephalon, the AV nidus is visible too. **E) DIGITALSUBTRACTION CENTRAL ANGIOGRAPHY**: this sequence shows the AV shunt between the left posterior cerebral artery (*feeder)* and the venous deep and superficial system. **F) 18-FDG PET**: a severe diffuse cortical-subcortical mostly left sided hypometabolism is evident

TREATMENT

Patient underwent to endovascular DAVF embolization with complete closing of the AV shunt. After postsurgical stabilization he was admitted to a neurorehabilitation structure showing a mild improvement in his conditions.

DISCUSSION

Our patient showed an anatomical olivopontocerebellar atrophy without extrapyramidal or cerebellar symptoms but with a severe multidomain cognitive impairment, probably linked to a diaschisis o cortical-subcortical connections caused by the high grade subcortical venous hypertension, according with PET and CSF reliefs . Despite DAVF was detectable even at previous brain MRI, it wasn't recognized leading to a diagnostic and therapeutic delay.

DEMENTIA LINKED TO DAVFS AND ITS CONSEQUENT CEREBRAL VENOUS HYPERTENSION IS A POTENTIALLY REVERSIBLE CONDITION BUT OFTEN MISDIAGNOSED AND, THEREFORE, RECOGNIZED WITH DELETERIOUS DELAY. A CORRECT DIAGNOSTIC STRATEGY IS RELEVANT TO ALLOW, IF POSSIBLE, AN ENDOVASCULAR/SURGICAL TREATMENT BEFORE INVOLUTIVE DAMAGE OCCURS TO BRAIN STRUCTURES.

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