Acta Neurologica Belgica (2024) 124:1397–1399
https://doi.org/10.1007/s13760-024-02490-4

NEURO-IMAGES

Unilateral proptosis and jugular thrombosis following ventriculoperitoneal shunt placement

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Received: 29 November 2023 / Accepted: 22 January 2024 / Published online: 1 March 2024

Keywords Cranial dural arteriovenous fistulas · Shunting

Clinical history and imaging findings

A 57-year-old patient presented to the emergency department for progressive decreased visual acuity over the past 10 months, losing the ability to read. The patient also complained of retro-ocular headaches, worse during concentration, without positional component noticed. Complementary examinations revealed a bilateral papilledema, a bilateral optic atrophy, a bilateral perioptic nerve sheath distention and a posterior flattening of the eye globes. The lumbar puncture opening pressure was 37 cm H20. Both CSF chemistry and cellularity were normal. No signs of cerebral venous thrombosis were noticed. In the hypothesis of pseudotumor cerebri, acetazolamide was initiated. The patient, however, developed severe hypokalemia (Liddle syndrome). The medical treatment was, therefore, suspended and a ventriculoperitoneal shunt (VPS) was surgically placed. The patient immediately reported an improvement in the visual function of his left eye after surgery.

Twenty days after surgery, the patient experienced again a deterioration of visual acuity despite the regression of the papilledema. It was associated with a 22 mm proptosis of the right eye. The patient also reported dizziness and a change in his headache symptomatology (i.e., now Valsalva related). A second brain MRI was performed, with particular focus on orbits and vascular structures. The MRI revealed a dural arteriovenous fistula, involving the right sigmoid sinus, associated with signs of right jugular thrombosis (Fig. 1). Digital subtraction angiography (DSA) confirmed a Cognard II a + b, Borden II fistula with direct communication between the right occipital artery and the right sigmoid sinus (Fig. 2) [1, 2].

The fistula was retrospectively visible on preoperative MRI and significantly increased in volume after the shunting surgery. According to the Monro–Kellie doctrine, the substantial drainage of the homolateral ventricle led to an increase of the shunt effect and of the vascular back-flow in the right sigmoid sinus, decreasing the flow in the right jugular gulf leading to its thrombosis.

Discussion

Cranial dural arteriovenous fistulas (dAVFs) are rare vascular malformations composed of abnormal vascular connections between meningeal feeding arteries and the intradural venous system [1, 3, 4].

Symptomatology of dAVFs depends on the arterial supply, location and venous drainage pattern [4]. Unilateral

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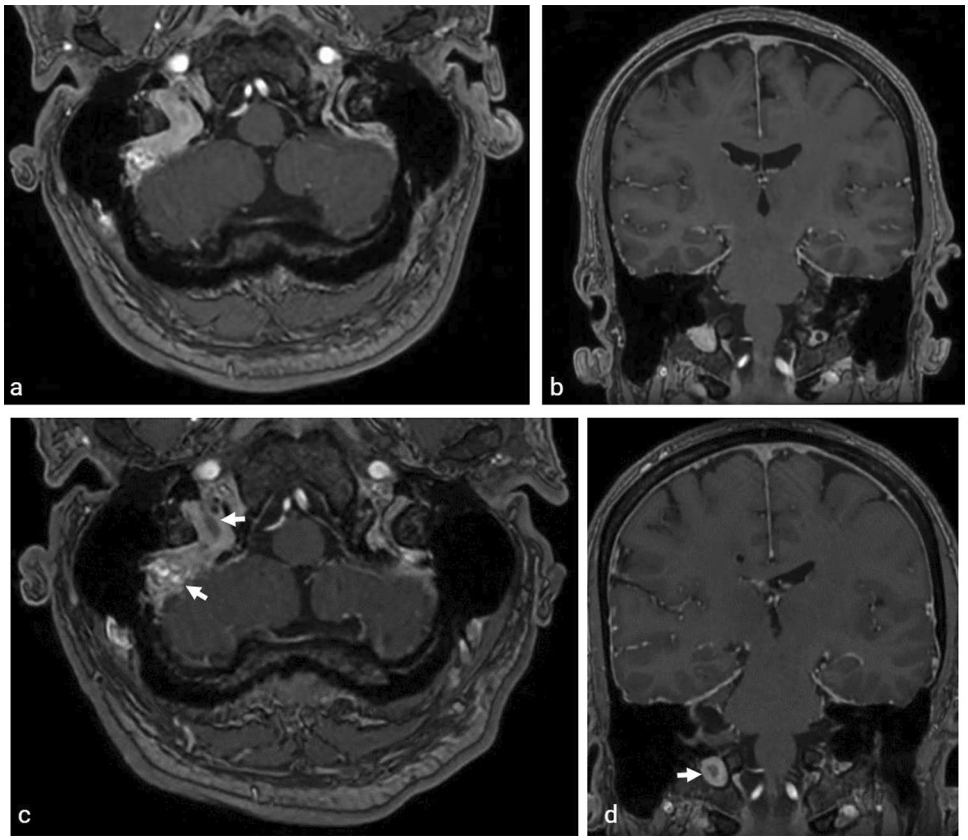

Fig. 1 MR-images: **a** transverse and **b** coronal T1 weighted + gadolinium before VPS surgery, **c** transverse and **d** coronal T1 weighted + gadolinium after VPS surgery showing a jugular thrombosis and a fistula enlargement

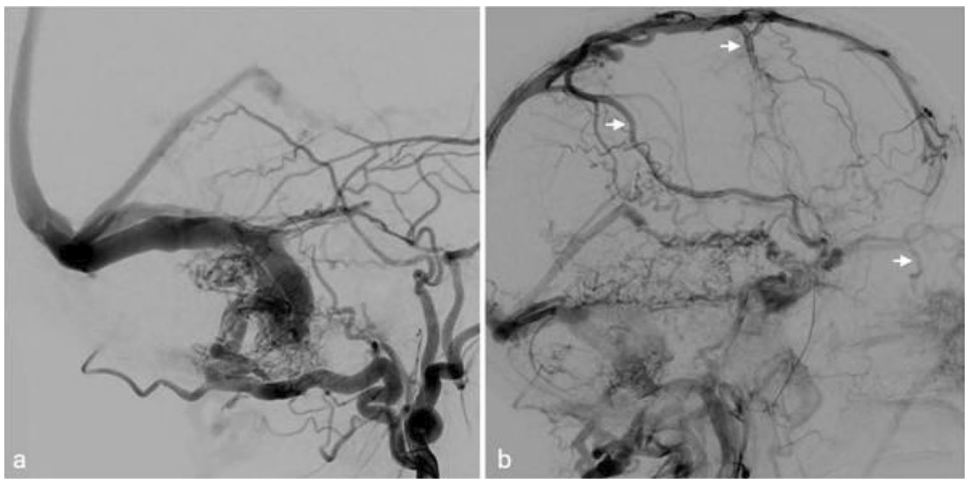

Fig. 2 DSA images: Cognard II a + b dural fistula involving right external carotid branches and the right sigmoid sinus, with a retrograde flow into cortical and ophthalmic veins

ocular manifestations, including proptosis and chemosis, are unusual but possible in non-carotid cavernous arteriovenous fistulae, depending on the venous drainage route, collaterals and fluid dynamics of the fistula. As an example, an illustrative case report of Ghorbani et al. details the history of two patients who experienced unilateral ocular symptoms resulting from direct fistulas between branches of the external carotid artery and the sigmoid sinus, with retrograde venous drainage to the ophthalmic vein [3].

Intracranial hypertension is frequent in dAVFs (20% for Cognard type II) [2]. In patients meeting pseudotumor cerebri criteria [5], we preconize specific consideration for stenosis and vascular abnormalities (especially dAVFs) involving venous sinuses, considering the risk of a possible clinical deterioration after ventriculoperitoneal shunt placement.

Conclusion

dAVFs are unfamiliar causes of intracranial hypertension. A close look for vascular anomalies before a cerebrospinal fluid diversion surgery is essential, since following the Monro–Kellie doctrine may lead to a worsened condition due to a vascular compartment expansion.

Author contributions Eric Vigneul: conception of the draft, conception of the figures, and clinical management of the patient. Elisabeth Van Boxstael: conception of the draft and clinical management of the patient. Pierre Goffette: conception of the figures and clinical management of the patient. Andre Peeters: substantial contributions to the conception and design of the work and clinical management of the patient. Edward Fomekong: substantial contributions to the conception and design of the work and clinical management of the patient. Ethics approval: no financial support was received for this work. The manuscript is an original contribution, not submitted simultaneously in another journal or published elsewhere, partially or in full in any form or language. Results are presented clearly, honestly, and without fabrication, falsification or inappropriate data manipulation (including image-based manipulation). The authors adhere to discipline-specific rules for acquiring, selecting and processing data.

Funding No financial support.

Data availability All medical data are available and recorded in a secure computerized medical file.

Declarations

Conflict of interest Nothing to declare.

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