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CASE REPORT - CSF CIRCULATION

JEDI (jugular entrapment, dilated ventricles, intracranial hypertension) syndrome: a new clinical entity? A case report

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Abstract
Patients with idiopathic intracranial hypertension are frequently obese women with normal/slit ventricles. Patients with high-pressure hydrocephalus, instead, present enlarged ventricles. We describe a 63-year-old woman with signs and symptoms of intracranial hypertension. Brain MRI revealed hydrocephalus. Venous Doppler ultrasound showed external compression of the omohyoid muscles on the internal jugular veins. During jugular vein decompression, intracranial pressure dropped from 18 to 6 mmHg. Patient is asymptomatic at 2-year follow-up, with decreased brain ventricles. These findings could represent a novel form of high-pressure hydrocephalus that can be successfully treated without a CSF shunt. We called this syndrome JEDI (jugular entrapment dilated ventricles intracranial hypertension).

Keywords Intracranial hypertension · Hydrocephalus · Jugular veins

Background

Patients with idiopathic intracranial hypertension (IIH) are more often obese women with normal or slit ventricles [6].

While IIH is often correlated with an extracranial increase of venous pressure (venous sinuses thrombosis, obesity, etc.), high-pressure hydrocephalus is acute or subacute and often has an intracranial etiology (like bleeding, infection, and trauma).

Here we present a unique and previously undescribed case that could represent a new nosological entity.

Case

A 63-year-old former nurse came to our attention in June 2016 for clinical and radiological signs of intracranial hypertension. She had been treated since 2002 at the Department of Rheumatology for a history of vasculitis.

In 2013, the patient had headache and mild bilateral papilledema. A brain MRI revealed an increase in size of brain ventricles with respect to a previous MRI (Evans index 0.30 vs 0.27 in 2009) associated with narrowed CSF convexity spaces. No intracranial sites of "CSF circulation" blockage were evident. The patient refused surgical intervention and was therefore treated with acetazolamide at a dosage of 2 g/day. Both headache and visual function improved. As brain ventricle volume did not change, the patient continued to receive acetazolamide at a daily dosage of 1.5 g/day.

In 2016, the patient's headache was initially intermittent and then became almost constant; the patient rated its severity 4 to 10 using numeric rating scale. Pulse synchronous tinnitus occurred daily. The patient also presented an impairment of visual acuity, with a grade 4 Frisen papilledema. Neurological examination revealed normal body mass index (21.8) and a

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mild cognitive slowing. Brain MRI showed a further increase of brain ventricles (Evans index 0.36, Fig. 1a). Brain MRI venography excluded the presence of thrombosis in the sinuses and also revealed an apparent turgor of the brain cortical veins. A baseline FDG positron emission tomography (FDG-PET) was performed, showing a diffuse state of hypometabolism (when compared with atlas of normality normalized for age and gender, Fig. 1c).

We therefore looked for the possible presence of an extracranial obstacle to cerebral venous outflow. The patient underwent intra- and extracranial venous Doppler ultrasound evaluation, normalized to the individual inflow [9]. The echo color Doppler (ECD) imaging was synchronized with the ECG trace.

A bilateral external compression of the omohyoid muscle on the internal jugular veins was apparent at high-resolution B-mode (Fig. 1e). The compression was never modified by changing posture, turning the head, and/or changing from supine to upright, as well as by activation of the thoracic pump. From the hemodynamic point of view, the compression corresponded to blocked flow with scarce collateral compensation. Finally, the ultrasound for jugular venous pulse (US JVP), corresponding to the variation of the cross-sectional area of the vein along a single cardiac cycle, was recorded [7].

After discussing the therapeutic options, the patient refused to be treated with a permanent CSF shunting. We therefore decided for a "possibly etiological" therapy. The patient gave her consent to this treatment and to share scientific information on her disease. Under general anesthesia with the patient in a supine position and head slightly elevated with respect to the heart, a right frontal external ventricular drainage was placed, taking care not to lose a drop of cerebrospinal fluid. Intracranial pressure, systemic blood pressure, and electrocardiogram were continuously monitored and recorded. Baseline

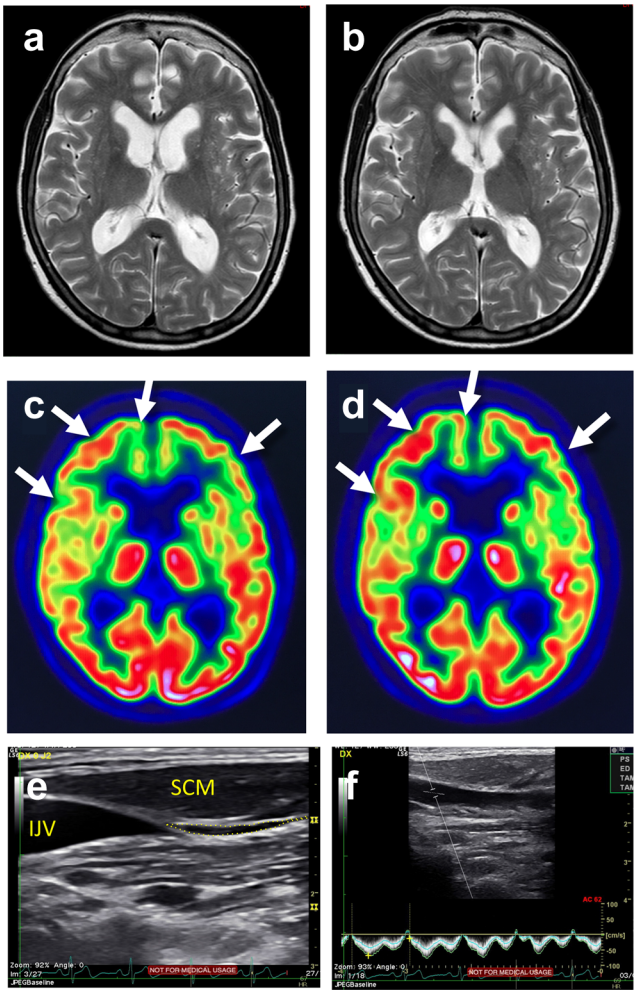

Fig. 1 **a** Preoperative MRI. **b** Postoperative 24-month follow-up MRI. **c** Preoperative FDG-PET showing hydrocephalus with hypometabolism (arrows). **d** 24-month follow-up FDG-PET showing decreased ventricles and improved metabolism (arrows). **e** High-resolution B-mode ultrasound of the longitudinal aspect of the right neck depicting the muscular nutcracker of the internal jugular vein (IJV). The compression of the omohyoid muscle (dotted line, below the sternocleidomastoid muscle (SCM)) on the IJV persists at head rotation and in whatever postural and respiratory condition. The lumen re-appears only when the patient is invited to slowly yawn. The picture was bilaterally symmetric. **f** Ultrasound (US) and Doppler show postoperative restoration of flow within the IJV

intracranial pressure was 16–18 mmHg, with a pathological waveform (Fig. 2). While on intracranial pressure continuous monitoring, the patient underwent also US JVP intraoperative monitoring of the internal jugular veins. A bilateral omohyoid muscle transection was therefore performed, at the places where these muscles determined a marked compression of the internal jugular veins. After the omohyoid muscle transection, a sudden drop of the intracranial pressure was observed (about 6 mmHg), with a normalization of the ICP waveform (Fig. 2). Another ultrasound evaluation of the internal jugular vein areas was therefore recorded (Fig. 1f). No further modifications of the ICP and the ICP waveform were observed in the next 20 min. The right frontal catheter was therefore removed.

Headache and tinnitus disappeared soon after surgery. The patient was discharged 2 days after surgery and acetazolamide was discontinued. Visual acuity improved and funduscopic examination performed at 1-, 6-, 12-, and 24-month follow-up showed no papilledema. Three, 12, and 24 month brain MRI showed a decrease of brain ventricles (Evans index 0.31) with more evident convexity CSF spaces (Fig. 1b). Positron emission tomography also showed an improvement of brain metabolism compared with preoperative findings (Fig. 1d).

At 28-month follow-up, the patient is asymptomatic.

Discussion

The omohyoid muscle consists of the superior and inferior bellies and an intermediate tendon. The muscle runs obliquely in the lateral cervical region connecting both the hyoid and the scapula bones. The intermediate tendon is connected to the clavicle by a fascial sling. The omohyoid muscle is the best landmark for identifying the internal jugular vein because of the direct adhesion of the intermediate tendon to the anterior wall of the vein. Compression of the internal jugular vein by the short omohyoid muscle may lead to flow blockage and modifications of intracerebral venous hemodynamics [8].

High-pressure hydrocephalus of an adult is acute or subacute and mainly occurs after intracranial bleeding, infection, and less frequently after trauma.

Patients with idiopathic intracranial hypertension are more often obese women. Brain MRI usually shows slit ventricles [5].

Clinical signs and symptoms in both cases are related to increased ICP: visual acuity impairment with papilledema, headache, pulsatile tinnitus, and, for acute hydrocephalus, drowsiness and cognitive impairment.

While idiopathic intracranial hypertension is often correlated with an extracranial increase of venous pressure (venous sinuses thrombosis, obesity, etc.), high-pressure hydrocephalus often has an intracranial etiology (like bleeding, infection, and trauma) [2–4]. Our patient presented with signs and symptoms of intracranial hypertension but had hydrocephalus associated with extracranial internal jugular vein entrapment: these findings can represent a new syndrome, a novel form of high-pressure hydrocephalus that can be successfully treated without a CSF shunt. We called this syndrome JEDI (jugular entrapment dilated ventricles intracranial hypertension) syndrome.

Interestingly, as a consequence of the increased intracranial pressure, our patient presented a diffuse state of hypometabolism before surgery that improved after intervention [1]. While our patient returned to normal life after surgery, we do not know if this hypometabolic state could chronically determine an alteration of brain functions. From a pathophysiological point of view, jugular vein entrapment, with significant increase of extracranial venous outflow resistance, could act as a constant Queckenstedt's maneuver, thus determining an increased ICP and an alteration of CSF dynamics, similarly to what happens in cases of intracranial idiopathic hypertension. What is not clear is why this patient had dilated ventricles instead of slit ventricles.

Conclusion

The case we have presented could represent a novel form of high-pressure hydrocephalus that can be successfully treated without a CSF shunt. We called this syndrome JEDI (jugular

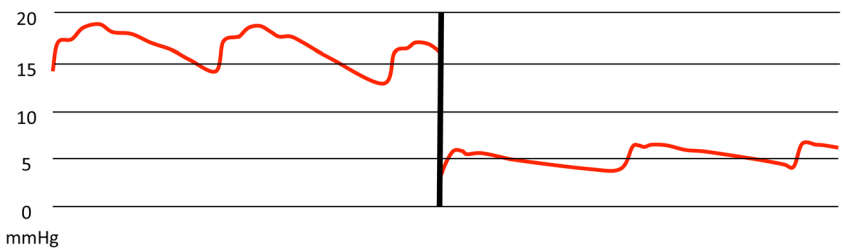

Fig. 2 Left: Baseline ICP, 16–18 mmHg with pathological wave. Right: ICP after omohyoid muscle transection 6 mmHg

entrapment dilated ventricles intracranial hypertension). Further cases and studies are needed.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Patient consent The patient has consented to the submission of the case report for submission to the journal.

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