Idiopathic intracranial hypertension (IIH) associated with hormonal therapy in a male patient: a case report.

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Background

Idiopathic intracranial hypertension (IIH) is characterized by signs and symptoms of raised intracranial pressure (ICP) with not established pathogenesis. The disorder occurs more commonly in obese women and typically within reproductive age, but about 9% of cases occur in men. Obesity and hormonal abnormalities have been proposed as risk factors for the development of this pathology in male patients. Common symptoms of IIH include headaches, visual loss or pulsatile tinnitus; papilledema is the hallmark of this disorder. Diagnosis is usually based on the modified Dandy criteria, which require the presence of raised ICP and no identifiable secondary cause.

Case Report

A **39-years-old man** presented with a persistent headache in the last year. The headache was constrictive, starting from the occipital regions and extending to the frontal regions bilaterally; it was not present upon awakening but appeared during the day with a gradual increase in intensity. He also reported sporadic episodes of nausea, rarely accompanied by vomiting and photophobia. The patient did not take any regular medication and had no relevant medical history, except for an hormonal therapy with follicle-stimulating hormone (FSH) and human chorionic gonadotropin (hCG) for idiopathic hypogonadotropic hypogonadism that he had started two years before and that he had discontinued after a year for the worsening his headache.

At admission, he showed a BMI of 28.4 kg/m^2; the neurological examination was normal and there was no papilledema bilaterally. **Contrast-enhanced brain MRI** showed an empty-sella and right transverse sinus hypoplasia with contralateral transverse sinus stenosis. The CT angiography did not show signs of cerebral venous thrombosis. A **lumbar puncture** documented an elevated CSF pressure (800 mmH2O), with a normal CSF profile. The withdrawal of CSF produced a transient improvement for his headache for several hours.

We made a diagnosis of IIH: acetozolamide was prescribed and the patient began a diet therapy with an improvement in his headache.

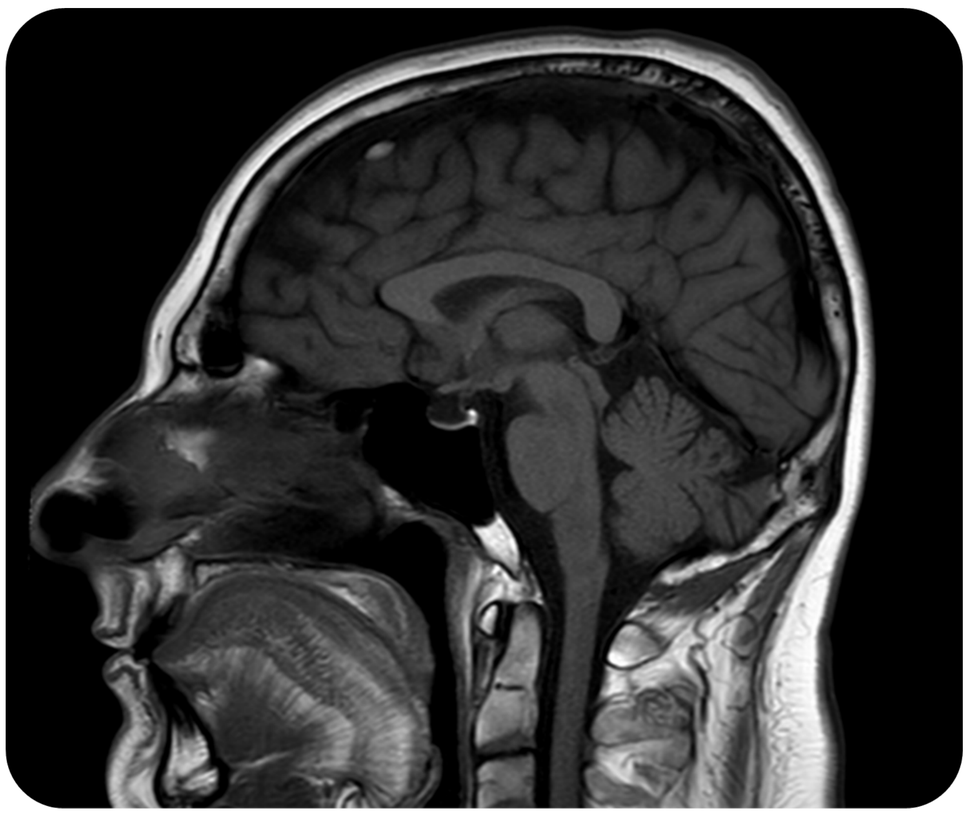


Figure 1. The "empty sella" sign on sagittal T1-weighted MR imaging.

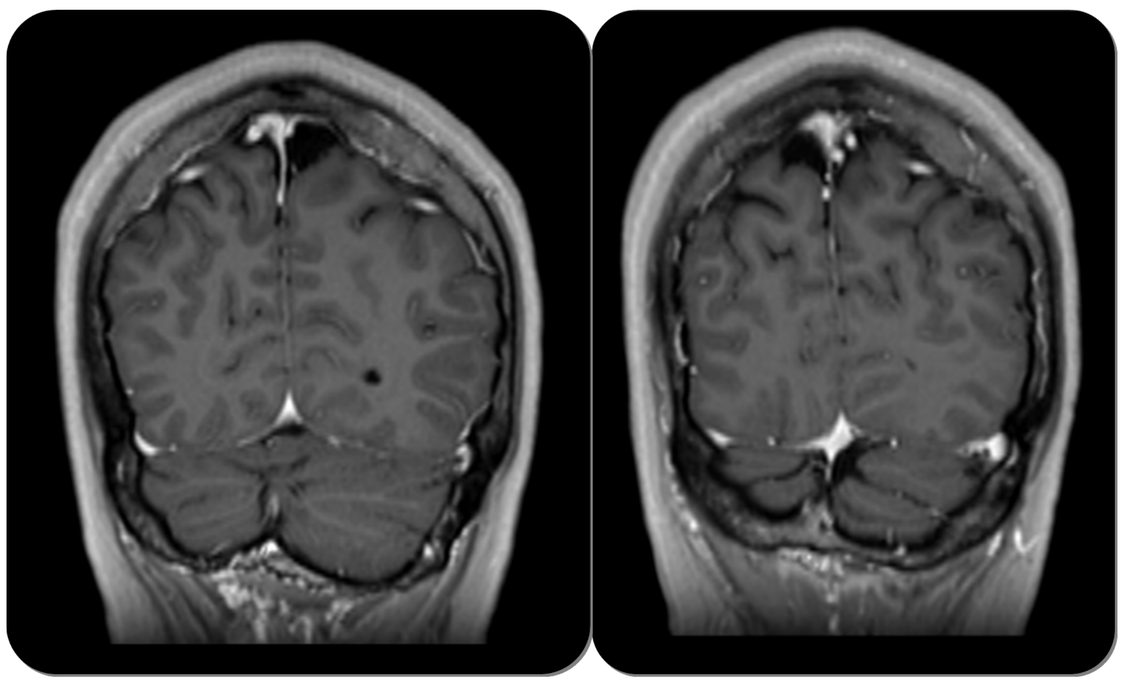


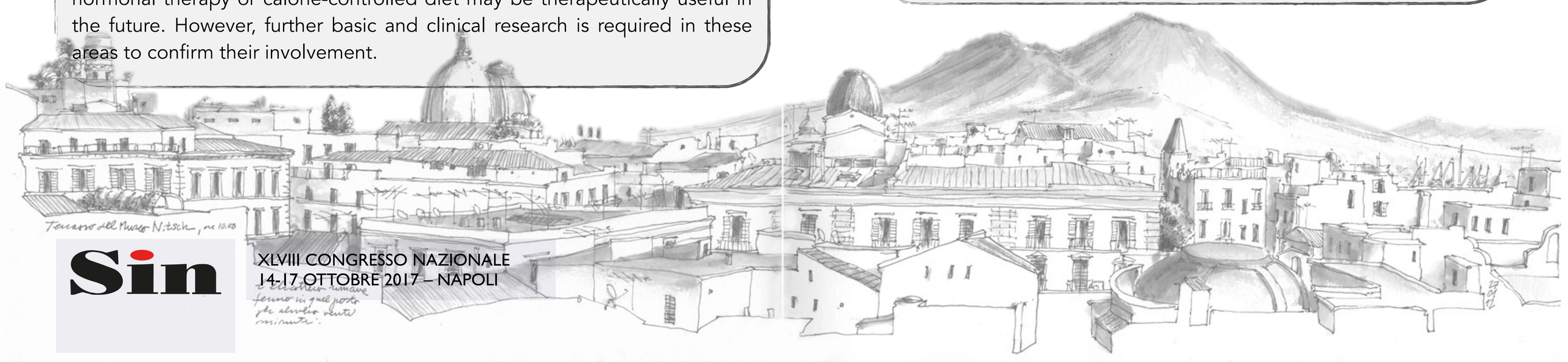
Figure 2. A narrowed left transverse sinus (figure on the left) with contralateral transverse sinus hypoplasia (figure on the right), seen on MR venography.

Discussion

The pathogenesis of IIH remains poorly understood. However, obesity and hypogonadism can emerge as causal mechanisms in the development of IIH in men, perhaps mediated through sex hormones and distribution of body fat. Our case can prove a **possible role of hormones like hCG and FSH in the pathogenesis of IIH**: the importance of an intervention in the form of hormonal therapy or calorie-controlled diet may be therapeutically useful in the future. However, further basic and clinical research is required in these areas to confirm their involvement.

References

1) J.A. Fraser, B.B. Bruce, J. Rucker, L.A. Fraser, E.J. Atkins, N.J. Newman and V. Biousse. Risk factors for idiopathic intracranial hypertension in men: A case-control study. J Neurol Sci. 2010 March 15; 290(1-2): 86.
2) K.A. Markey, S.P. Mollan, R.H. Jensen, A.J. Sinclair. Understanding idiopathic intracranial hypertension: mechanisms, management, and future directions. Lancet Neurol 2016; 15: 78-91.
3) Michael Wall. Update on idiopathic intracranial hypertension. Neurol Clin 35 (2017) 45-57.



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