

Spontaneous intracranial hypotension syndrome with extracranial vein dilatation

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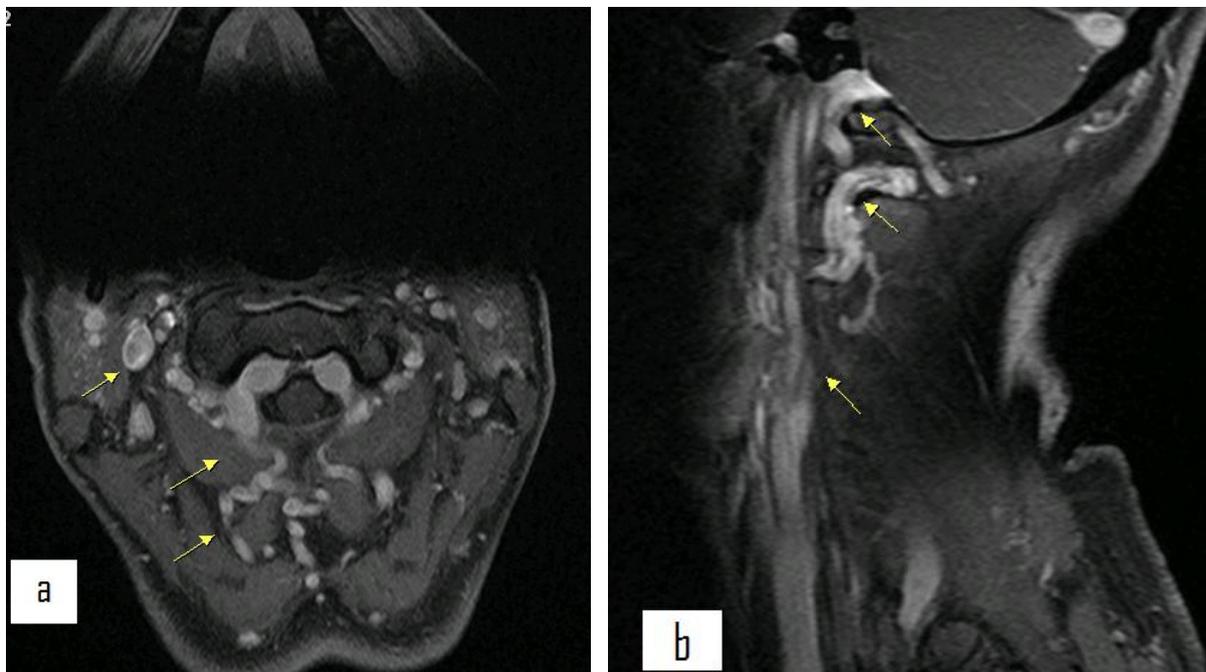
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Introduction

A 46-year old female patient presented with a 2-month history of cervicalgia and predominantly occipital headaches. The symptoms would appear in the upright position and would abate in the supine position. She also reported a symptoms' progression, in that the interval between the alteration of posture and the appearance of headache would become shorter. She also reported that in the beginning the headache would subside promptly, just by lying down, whereas later on the headache would persist for hours, even when recumbent. There was no history of trauma or a spinal procedure.

The neurological examination was unremarkable. The symptoms could be evoked by placing the patient upright, while Valsalva's maneuver led to symptoms' improvement.

An initial MRI scan of the brain and the cervical spine showed dilated epidural veins and tortuously dilated cervical veins (figure 1a-c)



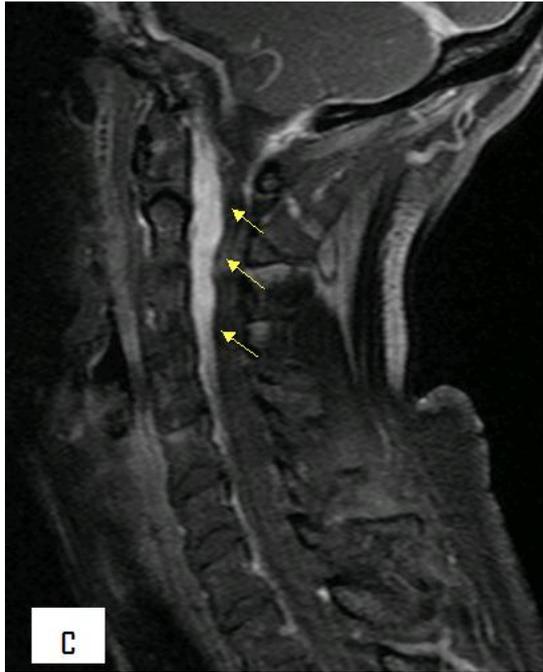


Figure 1: axial (a) and saggital (b) T1-Weighted images showing dilated extracranial veins. (c): Saggital T1-Weighted image showing dilated epidural veins.

Questions

1. What is the differential diagnosis?

Space-occupying intracranial lesions must be excluded in every patient with a sudden onset of persistent headaches. Migraine, meningitis, Chiari malformation and spontaneous intracranial hypotension have to be included in the differential diagnosis. Due to the presence of the venous anomaly, other less common causes – such as a dural fistula- have to be excluded.

2. What would be the next diagnostic step?

We performed a Digital Subtraction Angiography (DSA) to further estimate the morphology and dynamics of the intracranial and extracranial vessels. The DSA was unremarkable for any other vascular abnormalities, aside from the known pathology of dilated venous collaterals (Figure 2).

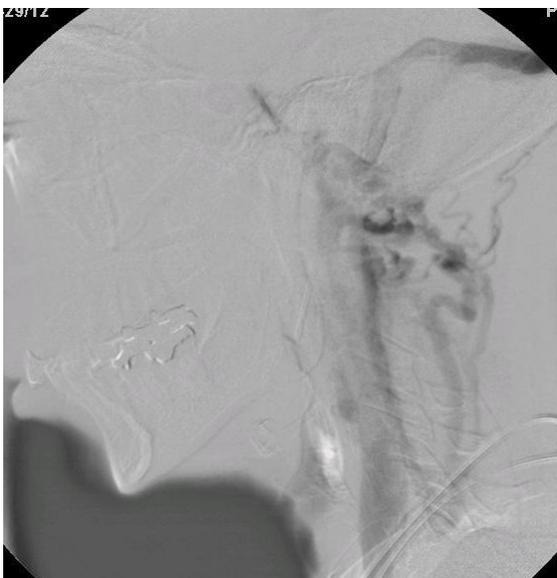


Figure 2.

3. What could be the cause of the patient's symptoms? What would be your next diagnostic step?

The patient's symptoms, especially the postural headaches, are very suggestive of intracranial hypotension. In such cases one should exclude a cerebrospinal fluid leak which can arise spontaneously, even in the absence of previous trauma or iatrogenic causes. In cases of intracranial hypotension, contrast-enhanced MRI scans may show basal pachymeningeal enhancement a finding that supports this diagnosis [1] (Figure 3),

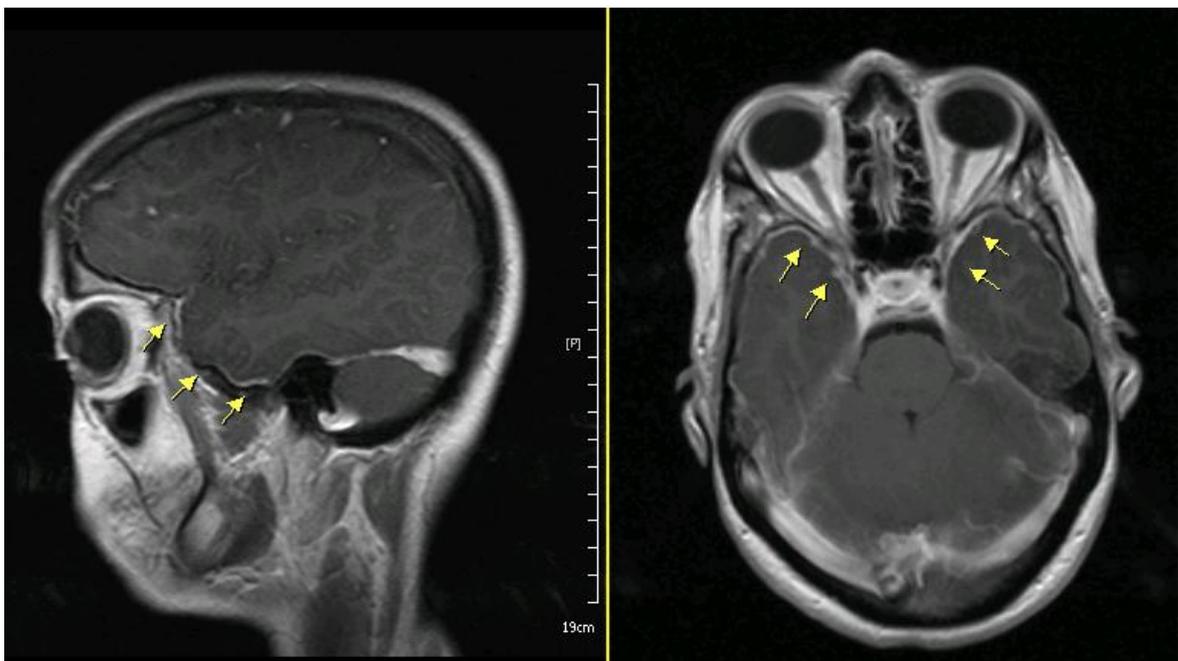


Figure 3.

The diagnosis of a spontaneous intracranial hypotension syndrome now seemed very probable.

4. What would be your next step?

In many cases, bedrest and hydration are sufficient for symptoms' resolution. A higher success rate, however, is achieved through epidural blood patches after identification of a spinal dural CSF-leak [2-3]. Spinal CT-myelographies or conventional myelographies are the best means to identify these leaks [4]. Indeed, myelography showed an epidural contrast enhancement between T2 and T10, thereby proving a CSF-leak at that height (figure 4).

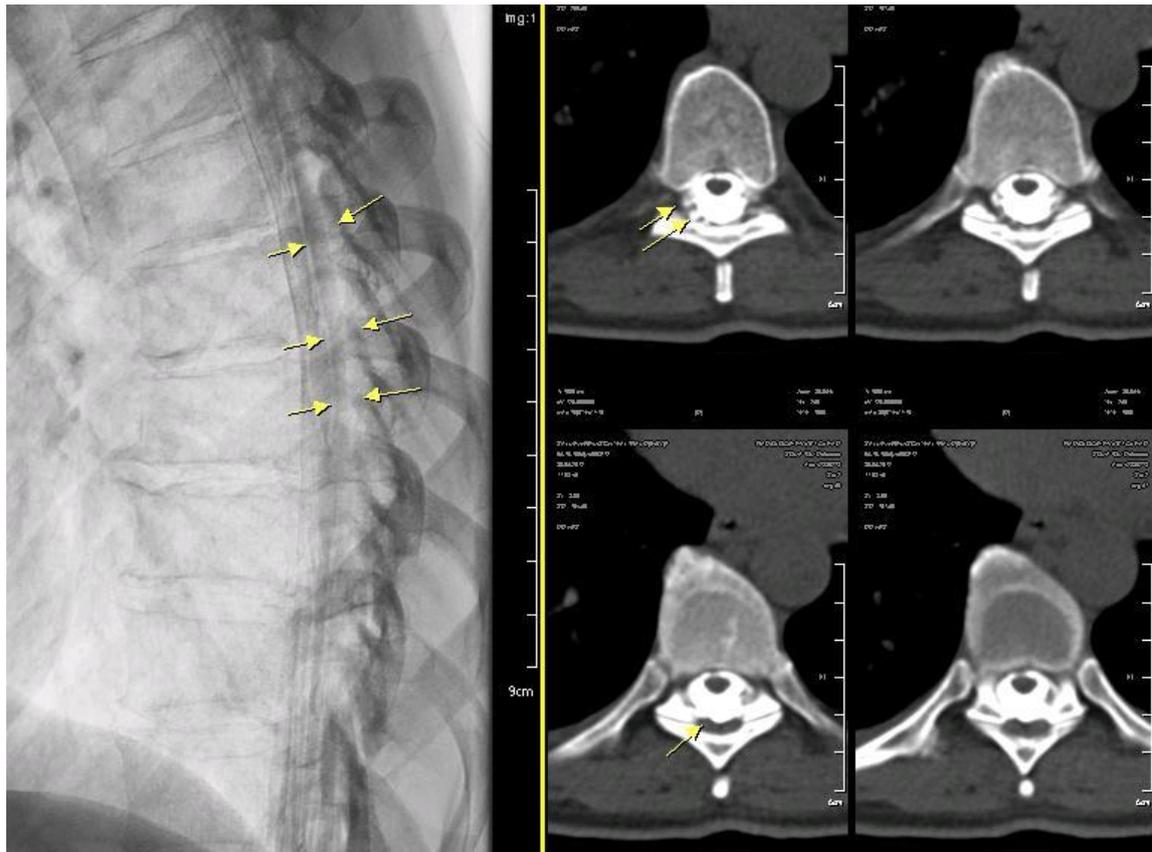


Figure 4

5. What would be your treatment plan?

Various techniques have been described for sealing spinal CSF-leaks, with epidural blood patches being the treatment of choice [5,6]. These can be applied transcutaneously with an epidural lumbar puncture or surgically through a small interlaminar fenestration, especially when the site of the dural tear has been identified [8]. Alternatively, fibrin glue sealant or direct neurosurgical repair can be performed for cases in which an epidural blood patch has failed [89]. We performed an interlaminar fenestration at level T7/T8. During the resection of the yellow ligament cerebrospinal fluid purged spontaneously. A silicon catheter was advanced epidurally for approximately 7cm and the distal end externalized. The day following the procedure, 2ml blood was applied via the catheter. The conscious patient showed no neurological deficits. After 2 days of bedrest and subsequent mobilization, the patient showed a remarkable improvement of her headache attacks up to the point of complete symptoms' resolution within 2 weeks.

6. Is there an association between the initial finding of venous dilation and the presence of intracranial hypotension? What clinical impact could this have?

The association between the spontaneous intracranial hypotension syndrome owing to CSF-leak and the dilated *extracranial/extraspinal* veins can only be hypothesized. Although there are numerous reports in the literature of distended epidural spinal veins after chronic CSF loss, we could not identify any case similar to our own in the literature. We postulate, however, that the pathophysiology of the epidural and extraspinal vein dilatation is similar and can

be explained by venous entrapment as hypothesized by Sood et al (2004) [9]; chronic low CSF-pressures lead to a decrease of the CSF-pressure and subsequent increase in the transmural vein pressure. This in turn expedites vein dilatation. Chronic vein distension leads to loss of vessel tone and thus increased vein collapsibility. In the recumbent position, where the vein pressure (P_V) equals CSF pressure (P_{CSF}), the now flaccid veins totally collapse, leading to intracranial blood entrapment, rapid raise of the cerebral blood volume and consequently increased intracranial pressure (ICP). The ICP crisis resolves slowly when $P_V = P_{CSF}$ and veins reopen. This theory cannot only explain the presence of the dilated vessels, but also the progression of the patient's symptoms; initially, the recumbent position would suffice to relieve the headache, just by raising the P_{CSF} . Later on, however, the chronic vein dilatation would cause a collapse of the capacity veins and hence increased cerebral blood volume at the prone position sustaining the headache. Addressing the problem of decreased CSF pressure caused eventually a normalization of the vein/CSF dynamics.

References

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