MRI-findings in idiopathic intracranial hypertension (Pseudotumor cerebri)

MRT-Befunde bei der idiopathischen intrakraniellen Hypertension (Pseudotumor cerebri)

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ABSTRACT

Background Pseudotumor cerebri, also known as idiopathic intracranial hypertension, is a disorder of increased intracranial pressure of unknown etiology. Main symptom is headache which has a characteristic similar to other headache diseases, therefore the idiopathic intracranial hypertension often remains undetected. The incidence is 1/100 000 with the number of unreported cases being much higher. This article highlights the essential role of MRI in the differential diagnosis.

Methods A literature search was carried out on idiopathic intracranial hypertension and Pseudotumor cerebri in English and German. Original and Review articles as well as case reports were taken into account. Since the main focus of the article is MRI diagnostics, some images were selected to illustrate the characteristic MRI morphological features.

Results and Conclusion The diagnosis of idiopathic intracranial hypertension is based on an exclusion procedure. Most common syndromes are headache, transient visual obscurations, pulsatile tinnitus and nausea. In the presence of an underlying cause for the increased intracranial pressure one speaks of a secondary intracranial hypertension. The diagnostics include a detailed medical history, neurological and ophthalmic examination, lumbar puncture, and neuroradiological imaging procedures. MRI, in particular, has become increasingly important in recent years, since signs for changes in cerebrospinal fluid pressure are now detectable and well-defined. The therapeutic approaches are symptom-oriented and aim to lower the pressure. With a precise diagnosis and timely start of therapy, idiopathic intracranial hypertension has a good prognosis, especially with regard to the preservation of eyesight.

Key Points:

- The idiopathic intracranial hypertension is an important differential diagnosis for unspecific headache and impaired vision
- Overweight women in childbearing age are particularly affected
- The most important component in diagnostics is MRI

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ZUSAMMENFASSUNG

Hintergrund Der Pseudotumor cerebri, auch idiopathische intrakranielle Hypertension genannt, ist eine Erkrankung mit erhöhtem Liquordruck unklarer Genese. Leitsymptom sind Kopfschmerzen, dessen Charakteristiken anderen Kopfschmerzerkrankungen ähneln, weshalb die idiopathische intrakranielle Hypertension oft unerkannt bleibt. Die Inzidenz wird mit 1/100 000 angegeben, wobei die Dunkelziffer wesentlich höher liegt. Dieser Artikel weist auf die essenzielle Rolle der MRT-Bildgebung in der Differenzialdiagnostik hin.

Methode Es wurde eine Literaturrecherche zu dem Syndrom IIH und Pseudotumor cerebri in englischer und deutscher Sprache durchgeführt. Berücksichtigt wurden sowohl Original- und Übersichtsarbeiten als auch Fallbeispiele. Da der Schwerpunkt des Artikels die MRT-Diagnostik ist, wurde eine Bilderauswahl getroffen, um die charakteristischen MR-morphologischen Merkmale zu veranschaulichen.

Ergebnisse und Schlussfolgerung Die Diagnose der idiopathischen intrakraniellen Hypertension beruht auf einem Ausschlussverfahren. Die typischen klinischen Symptome sind Kopfschmerzen, Sehstörung, pulsatiler Tinnitus und Übelkeit. Findet man eine zugrunde liegende Ursache für den erhöhten intrakraniellen Druck, spricht man von einer sekundären intrakraniellen Hypertension. Die Diagnostik beinhaltet eine ausführliche Anamnese, neurologische und ophthalmologische Untersuchungen, Lumbalpunktion und neuroradiologische bildgebende Verfahren. Insbesondere die MRT hat in den letzten Jahren an Bedeutung gewonnen, da mittlerweile gut erkennbare Zeichen einer Liquordruckänderung definiert sind. Die Therapieansätze sind symptomorientiert und haben das Ziel der Drucksenkung. Bei genauer Diagnostik und rechtzeitigem Therapiebeginn hat die idiopathische intrakranielle Hypertension eine gute Prognose, besonders was die Erhaltung der Sehfähigkeit betrifft.

Introduction

Physiologically, intracranial pressure in healthy adults is 5–15 cmH2O. Values above 20–25 cmH2O are considered pathologically increased intracranial pressure (intracranial hypertension) [1]. Elevated CSF pressure can lead to symptoms such as headache, vision disturbance, pulsatile tinnitus and nausea [2]. This syndrome was grouped under the name pseudotumor cerebri (PTC) and described by the German neurologist Max Nonne in 1904. His aim was to differentiate this group of patients from those with increased intracranial brain pressures due to cerebral tumors, as they exhibited the same clinical symptoms [3]. Nowadays, this definition is no longer up-to-date, since several pathologies, pathophysiological changes and drugs are now known to lead to an increase in intracranial pressure. If an underlying cause is found, it is referred to as secondary intracranial hypertension (SIH). If the etiology remains unknown, the disease pattern is called idiopathic intracranial hypertension (IIH), a concept introduced in 1989 by Corbett and Thomson.

IIH is a complex condition that is not yet fully understood, with an incidence of 0.9/100 000 in Western countries [4], although it can be assumed that unreported cases are significantly higher. On the one hand, technical developments in recent years have significantly improved diagnostics; on the other hand, the prevalence of obesity has increased worldwide [5].

Large differences are also observed depending on geographical location; the reason for this is not yet fully understood. It seems that genetic predisposition plays a minor role compared to environmental factors [6], although BMI provides the strongest correlation [7]. This explains the higher incidence of 19/100 000 in overweight women (20 % overweight). IIH can occur at any age, but the largest risk group is overweight women of childbearing age [8]. Depending on the study, the female-to-male incidence ratio varies from 4:1 to 15:1 [9]. In children, an incidence of 0.5/100 000 was found without gender and BMI-related differences [10].

Symptomology

Clinical symptoms are caused by increased intracranial pressure and may vary from individual to individual.

Headaches are the most common, occurring in 68–98 % of cases [4]. Headache characteristics may vary widely, can aggravate under certain circumstances such as coughing, body positioning and exertion, and may also show similarities to migraine and tension headache [11]. The latter is also the reason why IIH can easily be misdiagnosed.

The second most common complaint is vision disturbances, which are described in 72 % of cases, occurring on one or both sides and ranging from reduced visual acuity to blindness [12]. Vision disturbances are mainly caused by papillary edema, which can be detected in 95 % of cases during ophthalmological examinations, thus vision disturbances are a good indicator of increased CSF pressure. The remaining cases are referred to as idiopathic intracranial hypertension without papilledema (IIHWOP) [13, 14].

Pulsatile tinnitus is another common symptom at 60 % [12].

In addition, there are other, non-specific symptoms such as nausea, vomiting and olfactory disorders [15, 16].

Pathophysiology

Although aspects of this disease pattern are increasingly becoming known, the exact pathomechanism has not yet been clarified.

The Monro-Kellie doctrine states that in a rigid system such as the cranium, the three components (brain tissue, blood, cerebrospinal fluid) within it form a constant volume, whereby a change in one component necessarily results in a change in the other two [17].

There are several theories regarding the genesis of IIH. The common characteristic is the increased CSF pressure, but it is questionable whether there is a uniform mechanism at all and not rather several pathophysiological events acting on each other, which culminate in a vicious circle and result in the syndrome.

Cerebral spinal fluid

CSF is largely produced by the choroid plexus; its production is pressure-independent and relatively constant, but decreases with age. The theory that the increased intracranial pressure is due to an overproduction of cerebrospinal fluid could not be proven [18].

Dysregulation in water transport may play a role in the pathomechanism, as the water channel protein aquaporin-4 is known to be associated with brain edema and, together with aquaporin-1, forms the therapeutic approach to drug therapy. Thus, downregulation of these channels can lead to symptom improvement, whereas glucocorticoids and retinoids upregulate AQP1 channels and cause symptoms to worsen [19].

CSF flows from the ventricles into the subarachnoid space, where it is reabsorbed via arachnoid granulations. The resorption

rate depends on the pressure gradient between the subarachnoid space and the veins and sinuses. With increased venous pressure, CSF reabsorption is impeded, increasing cerebrospinal and intracranial pressure [20].

There are also additional CSF drainage pathways, such as the lymphatic and g-lymphatic systems [21], which explains why fetuses and children without arachnoid granulations have functioning CSF circulation.

Venous sinus

We know from invasive studies that intracranial venous pressure is elevated in IIH patients [22]. Whether this is a consequence or cause of increased intracranial pressure is still under discussion today. MR morphological correlates are uni- or bilateral sinus stenoses, which are found in almost all IIH patients [23].

Several publications describe the decrease in sinus stenosis after normalization of CSF pressure, suggesting the primary role of this pressure [24–27].

Other authors have observed that in the vast majority of cases, sinus stenoses persist after CSF pressure relief [28, 29]. Accordingly, sinus stenosis can probably be considered the cause of increased CSF pressure rather than the consequence in some cases.

Current scientific knowledge also widely accepts that sinus stenosis (primary or secondary) plays an important role in increasing intracranial venous and CSF pressure.

Due to the changed pressure gradient between the spinal fluid and veins, CSF resorption is reduced, which maintains or increases the pathological condition [30].

According to another hypothesis, disturbed vascular autoregulation can lead to cerebral hyperemia due to increased intracranial arterial influx even without a relevant venous outflow obstruction, which forms the basis for the increased intracranial pressure [31]. However, this could not be confirmed in a more recent study [32].

Parenchymal edema

The role of cerebral edema in the development of IIH has not been adequately demonstrated to date [33].

Obesity and sex

Associations between increased BMI or recent weight gain, female sex, and IIH are statistically proven, but pathophysiological bases are still lacking [34]. Various hormonal aspects are discussed, such as leptin, glucocorticoids, aldosterone or sex hormones [18, 35].

With certain drugs (various hormones, antibiotics, NSAIDs, vitamin A, lithium), increased intracranial pressures have been observed in some cases, although the causal correlation has not been clearly established [36, 37].

Diagnosis

IIH is a diagnosis of exclusion. The modified Dandy criteria continue to form the basis for diagnosis [15]:

1. Symptoms of intracranial pressure increase (headache, vision disturbance, etc.), papilledema
2. Evidence of elevated cerebrospinal fluid pressure (> 25 cmH20 with lumbar puncture in the supine position)
3. Normal CSF composition
4. Exclusion of differential diagnoses
5. No focal neurological deficit (except unilateral and bilateral abducens palsies)
6. Normal CT or MRI

Technical developments of the last decades, especially in imaging diagnostics, led to the revision of item 6. MRI imaging made it possible to visualize indirect intracranial pressure signs [38, 39].

In addition, the increasing number of publications in recent years has led to expansion of diagnostic criteria. In 2013, the diagnostic criteria were revised and updated, especially for patients with IIHWOP, who often have less elevated CSF pressure than patients with papilledema [14]. In such cases, the diagnosis of IIH should be made on the basis of radiological features in conjunction with other available clinical and diagnostic findings. Thus, the limit of CSF opening pressure between 20–25 cmH20 is also discussed as a diagnostic gray area and considered pathological by some authors from 20 cmH20 onwards [1, 28, 40]. If not all criteria are met, the diagnosis of IIH is an individual decision [40].

MRI findings

MRI is now the most important component in the diagnosis of IIH, used not only to rule out causes of increased intracranial pressure, but also to detect subtle changes in the cranium caused by increased pressure. Due to higher sensitivity and lack of radiation exposure, it is clearly superior to computed tomography.

An appropriate standard MRI protocol to rule out secondary intracranial pressure elevation includes:

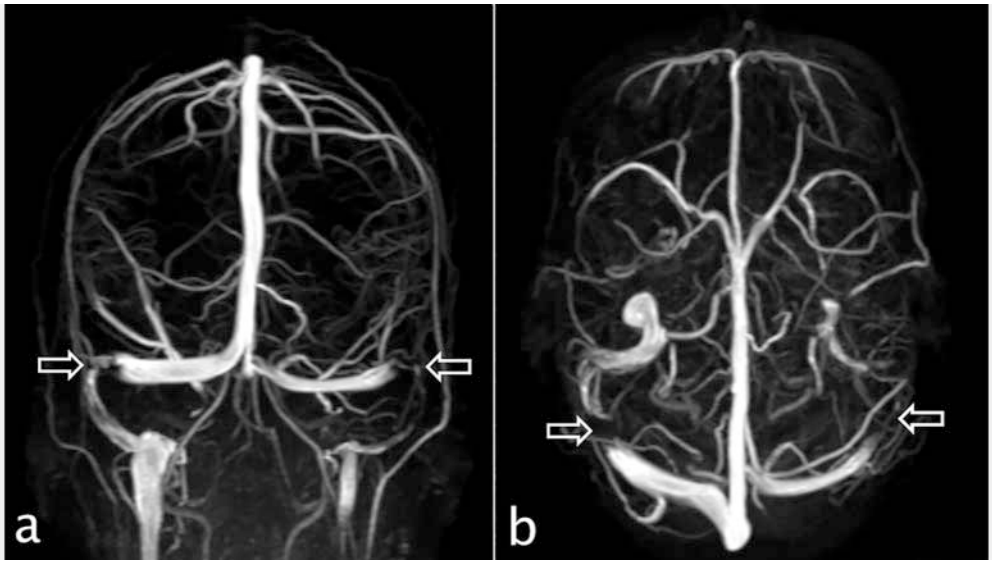
- Axial DWI (3 mm),
- Axial T2w (3 mm),
- FLAIR (fluid-attenuated inversion recovery, 4 mm),
- MR venography, e. g. PCA (venous phase contrast angiography, velocity encoding VENC 15 cm/s) for non-contrast imaging of the venous vessels,
- Coronal STIR (fat-suppressed inversion recovery sequence) of the orbita and sella (3 mm).

There are now numerous published MR morphologically detectable signs of intracranial pressure elevation that are reliably detected with the MR protocol listed above, although good quality MR venography is of critical importance [41].

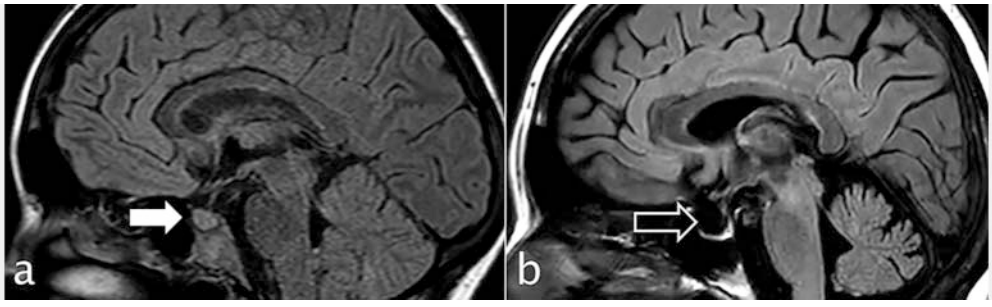
Typical pitfalls of venous phase-contrast angiography include Paccioni granulations, which can mimic stenosis. In most cases, these can be identified as such based on their strong T2w hyperintense signal. “Genuine” sinus stenoses do not show high signals in T2w images. In case of ambiguous findings, contrast-enhanced MR angiography in combination with a 3 D T1 data set can also be performed as a supplement.

The most important MRI signs in descending diagnostic weighting are:

1. Uni- or bilateral sinus stenoses (▶ **Fig. 1**),
2. Excavation of the pituitary gland to the “empty sella” (▶ **Fig. 2**),



▶ **Fig. 1** Venous PCA shows bilateral sinus stenoses at the junction of the transverse sinus and the sigmoid sinus (arrows), coronal **a**, axial **b**.



▶ **Fig. 2** Left **a** a healthy young female patient (filled arrow), right **b** female patient with increased CSF pressure and excavated pituitary gland (empty arrow) in the sagittal FLAIR sequence.

3. Bilateral dilatation of the optic sheaths (▶ **Fig. 3**),
4. Flattening of posterior sclera (▶ **Fig. 4a**),
5. Tortuosity of the optic nerve (▶ **Fig. 4b**),
6. Preliminary enhancement of the optic nerve,
7. Dilatation of the Meckel cavity,
8. Expansion of the superior ophthalmic vein,
9. Dilatation of the foramina of the skull base, such as the foramen ovale [42].

Uni- or bilateral sinus stenoses are found in almost all IIH patients. Bilateral sinus stenoses are especially pathognomonic for this disease [11]. These are found predominantly at the transition from the transverse sinus to the sigmoid sinus (▶ **Fig. 1**), but also in the course of the transverse sinus and in the distal sigmoid sinus above the confluence of sinuses [43]. The extent of stenosis ranges from mild constriction to flow interruption. In some cases, reversibility of the stenoses is then seen after successful pressure reduction via lumbar puncture [24, 44], in other cases, however, a persistence of the stenoses can be observed, which makes them appear as fixed stenoses [28, 29].

In a healthy state, the pituitary gland has a roundish configuration, especially in younger patients. Under increased CSF pressure, however, it shows flattening or excavation to the point of an "empty" sella (▶ **Fig. 2**). After normalization of CSF pressure, there may be an increase in pituitary height again. The normal values for pituitary height differ between the sexes; in addition, pituitary volume decreases with age, which must be taken into account when assessing this pressure phenomenon [45].

The width of the liquid-filled optic sheaths shows no age-related variance, which is why it is a good marker for intracranial pressure conditions [45]. The optic sheaths are barely detectable when there is negative pressure; they are usually slender, and when the pressure is positive they expand (▶ **Fig. 3**). In the latter case, one speaks of an optic sheath hydrops, which can also be visualized sonographically.

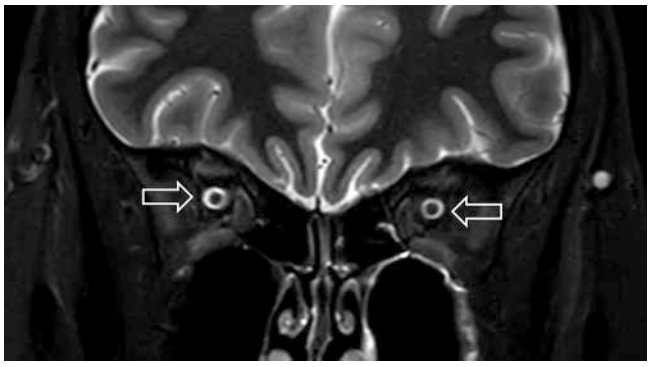
Although congestion papillae are not an MRI phenomenon, they are still visible MR morphologically in pronounced cases (▶ **Fig. 4a**).

Unlike other forms of CSF hypertension (hydrocephalus), the ventricles in IIH are normally wide.

Differential diagnosis includes all diseases that may lead to or be associated with increased intracranial pressure. The clinical work-up includes a detailed neurological examination with laboratory and cerebrospinal fluid diagnostics as well as ophthalmologi-

cal examinations with special regard to congestion papillae. It should be noted that a pathologically altered CSF status rules out the diagnosis of IIH, but absent congestive papillae does not (IIHWOP).

Radiologically relevant differential diagnoses are primarily sinus or venous thromboses (▶ **Fig. 5**), intracranial and spinal lesions, various forms of hydrocephalus, vascular malformations (dAVF), meningeosis, meningitis, and encephalitis.



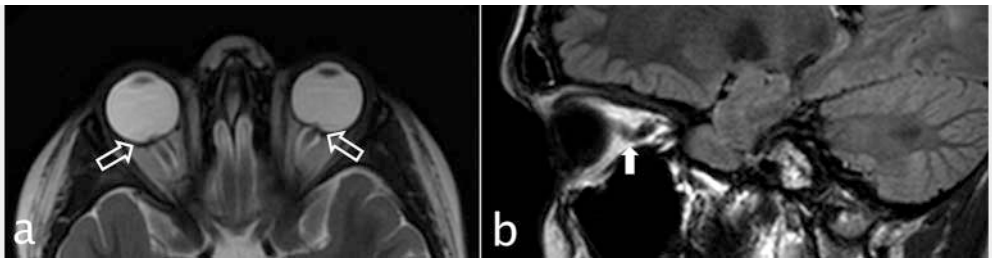
▶ **Fig. 3** Expanded optic sheaths (arrows) in the coronal STIR sequence.

Therapy

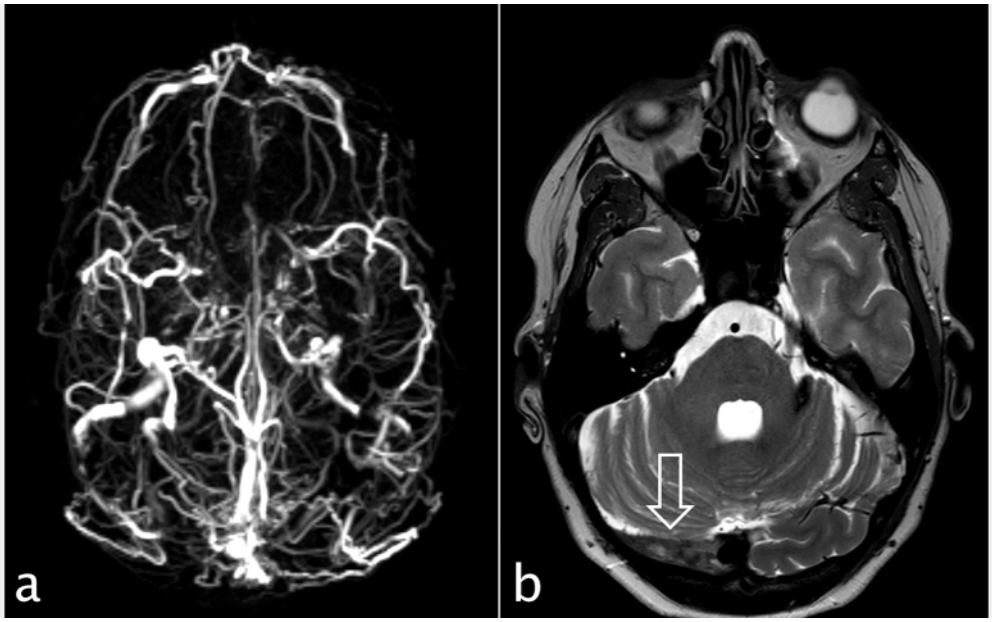
To date, there are no evidence-based treatment guidelines [46]. The therapeutic decision is individual and depends on the severity of the symptoms. The primary goal is to preserve vision and relieve headache symptoms. The desired pressure reduction can be achieved by means of invasive and non-invasive forms of therapy.

Unless acute visual disturbances (loss of visual acuity) force an immediate therapy, treatment follows an escalation strategy:

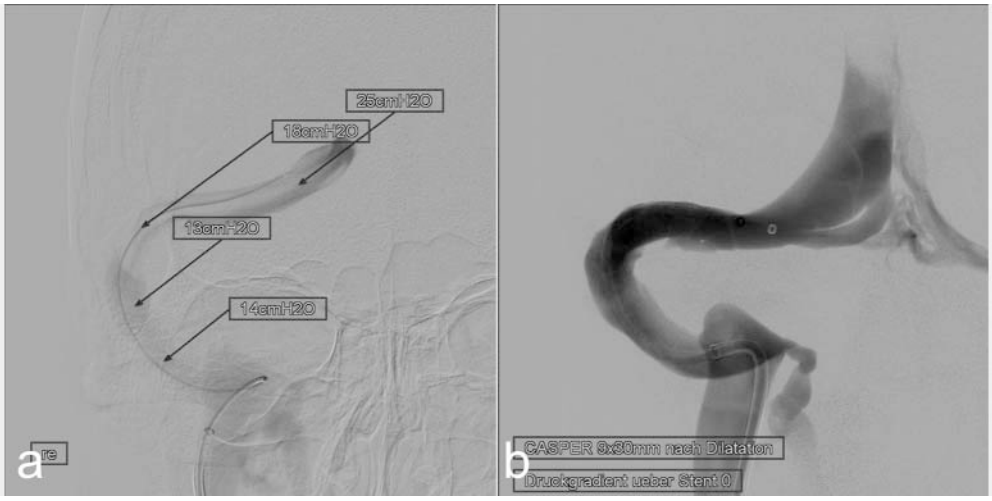
1. Non-invasive options include medications and weight loss. The most effective drugs are acetazolamides from the group of carbonic anhydrase inhibitors, which reduce CSF production by up to 57 % [46]. Other medications include the diuretic furosemide and the antiepileptic topiramate [47, 48]; the latter is



▶ **Fig. 4** Congestive papillae and flattening of the posterior sclera (empty arrows) in the axial T2w sequence **a** and tortuosity of the optic nerve (white arrow) in the sagittal FLAIR image **b**.



▶ **Fig. 5** Thrombosis of the transverse and sigmoid sinuses on both sides and part of the superior sagittal sinus in the venous PCA **a**. Correlated to this is the lack of a flow signal in the right transverse sinus (arrow) in the axial T2-weighted sequence **b**.



▶ **Fig. 6** Diagnostic angiography of a 21-year-old female IIH patient with bilaterally fixed sinus stenoses as demonstrated by MRI. Digital subtraction angiography shows a pressure gradient of 11 cmH20 **a** over the sinus stenosis on the right at the transverse-sigmoid junction. After successful stent angioplasty of this stenosis, normalization of the pressure gradient to 0 cmH20 above the stent **b**.

also used in migraine prophylaxis. Corticosteroids are now no longer recommended in therapy [49].
Numerous studies have shown a positive correlation between BMI and CSF pressure [50]. Congestive papillae in particular react sensitively to weight reduction, their reversibility is a good indicator of successful therapy [51].

2. Invasive methods include lumbar puncture (LP), which is also considered the diagnostic gold standard. Lumbar puncture is used to determine opening pressure, obtain CSF for analysis, and provide therapeutic pressure relief. In a few cases, a single LP is sufficient to produce longer-term clinical improvement [27], but most often the procedure must be repeated several times. On average, approximately 30 ml of CSF is collected.
3. Stent angioplasty of the sinuses: Endovascular therapy has become the first invasive treatment option of choice, especially in specialty centers. For a long time, this was mainly true for MR-morphologically fixed sinus stenoses after lumbar puncture, but is now also valid for non-fixed stenoses. Regardless of the direction of the causality between increased CSF pressure and venous pressure, there is positive interaction between the two components [52]. Implantation of a stent in the stenosis not only reduces the venous but also the CSF pressure, which leads to an improvement in the clinical symptoms [53, 54] (▶ **Fig. 6**).
4. Other surgical measures to reduce the CSF pressure should be mentioned here, but apart from bariatric approaches, they are becoming more and more of a secondary importance: lumboperitoneal and ventriculoperitoneal shunt [55], optic nerve fenestration [56] as well as bariatric surgery for weight reduction [57].

Summary

IIH is an often unrecognized condition and predominantly affects young overweight women. Patients often present to the clinic with nonspecific headaches. Because headaches occur as a concomitant symptom in many diseases, they often go unrecognized although they are much more common than assumed. In these patients, MRI should be performed during the diagnostic workup to exclude secondary causes of headache. Since IIH is associated with increased CSF pressure, it is imperative to know the pressure signs on MRI and choose an appropriate related protocol. In particular, a good MR phlebography technique is of great relevance, since bilateral sinus stenoses are pathognomonic for this pathology.

Conflict of Interest

The authors declare that they have no conflict of interest.

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