

It is Not Just a Headache: Atypical Case Presentation of Idiopathic Intracranial Hypertension

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Case Report

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Abstract

Background: Idiopathic intracranial hypertension (IIH) is an uncommon but serious neurological condition that can present with various symptoms. In this report, we present a case of IIH that initially presented with left-sided ptosis and an inferior altitudinal visual field defect, along with a review of the relevant literature.

Case presentation: A 64-year-old Egyptian man presented with a 1-week history of acute onset of left eye ptosis and a transient episode of complete visual loss in the same eye, lasting a few minutes. Vision recovered spontaneously, but he noted a persistent inferior altitudinal visual field defect. Brain MRI showed bilateral frontal and parietal subcortical ischemic foci, lacunar infarcts, and subcortical arteriosclerotic encephalopathy. A computed tomographic angiography was performed to assess the cerebral arteries. Stenosis of both transverse sinuses was accidentally discovered in the venous phase of CTA. A high opening pressure was found. Diagnostic central venous catheterization showed bilateral transverse sinus stenosis; transverse sinus stenting was recommended. The patient had a left transverse sinus stent inserted. He was given a diagnosis of IIH.

Conclusion: IIH can be atypically presented and occur later in life. Patients with normal body mass index and those 50 years or older make up a small proportion of patients with IIH but appear to have better visual outcomes than more typical patients with IIH. This case underscores the importance of considering IIH diagnosis in patients with unexplained visual or ocular presentations.

Background

Idiopathic intracranial hypertension (IIH) is a disorder of increased intracranial pressure in the absence of cerebrospinal fluid (CSF) outflow obstruction, masses, or other secondary etiology¹. The incidence of IIH was 3.3 per 100 000 in women and 0.3 per 100 000 in men ($P \leq 0.001$)².

The prevalence of IIH is lower in the elderly population³, and its atypical presentations further reduce clinical suspicion, which may lead to missed or delayed diagnosis. Early diagnosis and intervention are crucial; if left untreated, severe optic disc oedema may, ultimately, lead to permanent vision loss³. Typically, patients present with headache, dizziness, pulsatile tinnitus, visual disturbances, blurred vision, diplopia, photophobia, visual field defects, and papilledema on fundoscopy. While a visual field defect is a common manifestation of increased intracranial pressure (ICP), to our understanding, no cases have been reported in the literature of IIH presenting with an altitudinal field defect resembling that of anterior ischemic optic neuropathy (AION)⁴.

Case presentation

The patient is a 64-year-old Egyptian man who presented to the Department of Neurology with a 1-week history of acute onset of left eye ptosis and a transient episode of complete visual loss in the same eye,

lasting a few minutes. Vision recovered spontaneously, but he noted a persistent inferior altitudinal visual field defect. Concurrently, he developed unilateral blurred vision without associated aura, photophobia, phonophobia, diplopia, loss of consciousness, seizures, or sensory disturbances. The symptoms significantly affected his daily activities. He denied prior similar episodes or a family history of neurological or autoimmune diseases. The other systems review was unremarkable.

The physical examination upon admission found the patient was alert, conscious, and hemodynamically stable. Neurological examination revealed persistent left partial ptosis without diurnal variation or fatiguability. Field testing showed a left inferior altitudinal defect. Pupils were round, regular, and reactive bilaterally, and the optic disc appeared pale bilaterally. Cranial nerves were intact. Neurological examination revealed no facial weakness, dysphagia, or limb weakness. There was no rigidity or spasticity. Muscle power was 5/5 in all limbs, with intact sensation and reflexes. There were no cerebellar or meningeal signs, and gait was normal. The patient has developed bilateral sequential upper limbs superficial thrombophlebitis during his hospital stay. Laboratory investigations on admission showed normal Antithrombin III, Protein C&S levels. No Prothrombin G20210A mutation or Methylenetetrahydrofolate reductase (MTHFR C677T) mutation was detected. Serological workup was negative for Anti-neutrophil cytoplasmic antibody (cytoplasmic pattern) (ANCA-C) and Anti-neutrophil cytoplasmic antibody (Perinuclear pattern).

Inflammatory markers were normal (ESR: 13 mm/h, CRP: 0.03 mg/dL). During the follow-up visit, the patient was found to have a positive homozygous factor V mutation.

Brain magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) were performed to rule out any ischemic stroke or aneurysm. Brain MRI showed bilateral frontal and parietal subcortical ischemic foci, lacunar infarcts, and subcortical arteriosclerotic encephalopathy (Fig. 1). Left sphenoid sinusitis with possible fungal colonisation, no gross left orbital infection.

Because the diagnosis remained uncertain and a vascular cause was suspected based on the presentation, onset, and history of recurrent thrombosis, computed tomographic angiography (CTA) of the cerebral vessels was performed to evaluate the cerebral arteries. Bilateral transverse sinus stenosis was incidentally identified during the venous phase of the study, prompting us to consider lumbar puncture and to re-evaluate the patient's history for headache. The patient then reported a mild daily headache, which was aggravated by prolonged speaking.

Patient underwent a lumbar puncture for CSF opening pressure. Initial CSF opening pressure was 500 mm H₂O. Serial Lumbar punctures (on acetazolamide) were done at intervals, showing the following readings: 2 weeks later, it was 500 mm H₂O, one month later, it was 150 mm H₂O, and 2 months later, it was 400 mm H₂O.

It was recommended to go for a diagnostic central venous catheterization. The cerebral angiography showed bilateral transverse sinus stenosis (Fig. 2), seen at the junction of the transverse sinus with the sigmoid sinus on both sides. The plan was to start by stenting one side, which is usually enough to

control the clinical condition. If stenting of one side is not enough to control his symptoms within 3–6 months, the other side will be stented afterwards. The decision was to start stenting the left transverse sinus stenosis (Fig. 3), as the left transverse-sigmoid-jugular system was larger than the right side, so it was supposed to be the dominant side responsible for draining the majority of venous outflow from the brain.

The diagnosis of IIH was made clinically based on the International Classification of Headache Disorders, 3rd Edition (ICHD-3)⁵, which includes signs and symptoms of increased intracranial pressure (headache, papilledema); absence of localizing neurological signs, except for possible abducens nerve palsy; normal neuroimaging; and normal cerebrospinal fluid composition with elevated opening pressure.

Initially, the patient was started on Acetazolamide, which was gradually increased to 3 gm/ day, with an observed improvement in headache and vision. Moreover, the dose of Acetazolamide was reduced to 750 mg/day after stenting of the transverse sinus with a complete improvement in ptosis, field defect, and headache.

The patient was co-managed by a multidisciplinary team, including neurologists, interventional radiologists, and ophthalmologists. A formal multidisciplinary meeting was convened, and clinical decisions were made collaboratively across departments.

Discussion

Idiopathic intracranial hypertension (IIH) can present with a broad spectrum of ocular and visual manifestations. In this case, the patient is a male over 50 years old who developed acute, unilateral partial ptosis accompanied by an inferior altitudinal visual field defect. Notably, there was no evidence of papilledema or significant headache. Taken together, these features were more consistent with a vascular etiology than with typical IIH.

Visual field defects in IIH are known to improve with appropriate treatment and may arise through several mechanisms. The primary mechanism is believed to involve CSF pressure–induced disruption of axonal transport, resulting in intraneuronal optic nerve ischemia at the level of the optic nerve head. Another frequent mechanism is enlargement of the blind spot, which typically occurs due to peripapillary hyperopia and represents a refractive cause of visual loss⁶. Less frequent causes of visual loss in idiopathic intracranial hypertension include neurosensory detachment, resulting from fluid tracking from the optic disc to the fovea, and refractive visual impairment associated with choroidal folds. In this case, the patient demonstrated an inferior altitudinal field defect, a presentation that does not typically align with descriptions commonly reported in the literature.

A literature review shows that patients with IIH and mild visual loss typically develop optic nerve–related visual field defects that follow localized nerve-fiber bundle patterns. Abnormalities are more frequent in the inferior hemifields than the superior ones, most commonly presenting as a partial inferior arcuate defect⁷.

Oculomotor palsy is an uncommon manifestation in IIH, generally attributable to elevated intracranial pressure⁸. In the present case, however, the patient uniquely exhibited oculomotor involvement as partial ptosis alone. This atypical finding prompted a thorough evaluation for secondary causes, all of which were excluded. Consequently, we propose that partial third nerve palsy may represent a non-localizing sign of IIH, though secondary etiologies should always be ruled out. The distinctive feature of this case lies in the combination of unilateral inferior altitudinal field defect with unilateral partial ptosis. While third nerve palsy has been reported in several atypical presentations of IIH, only one previously documented case fulfilled the established diagnostic criteria⁹.

In selected patients with IIH secondary to venous sinus stenosis and elevated ICP, venous sinus stenting can abolish the venous pressure gradient, lower ICP, and lead to functional, neurological, and ophthalmological improvement¹⁰.

Venous sinus stenosis is increasingly recognized as a key factor in the rise of intracranial pressure (ICP) and is now considered central to the pathophysiology of idiopathic intracranial hypertension (IIH). Elder et al. reported transverse sinus stenosis in 10–90% of IIH patients, compared with 6.8% in the general population¹¹. Stenosis usually produces flattening or collapse of the dural sinus contour. Extrinsic stenosis can begin with a small ICP elevation that, through a positive-feedback mechanism, causes a larger secondary increase in ICP¹². The external pressure on a collapsible conduit depends on flow and thus alters its effective diameter. Consequently, modest initial ICP rises, triggered by factors such as weight gain or hormonal change, compress the collapsible transverse sinus, impair venous outflow, reduce CSF absorption into the venous system, and further elevate ICP, perpetuating the cycle. Interventions that relieve sinus stenosis, such as venous stenting, therefore have the potential to interrupt this vicious circle.

Cerebrospinal fluid (CSF) shunting is indicated for patients with visual decline, papilledema, or marked systemic symptoms of raised intracranial pressure. Optic nerve sheath fenestration (ONSF) is the preferred initial intervention for vision loss from severe papilledema when headache is not prominent¹¹.

Both procedures, however, carry substantial postoperative risks and appreciable recurrence rates. The use of transverse sinus stenting in this case aligns with findings from prior studies.

Conclusion

IIH may manifest with a wide range of ocular abnormalities and visual field defects. Findings such as partial ophthalmoplegia and specific visual field defects raise suspicion for IIH. For patients with venous sinus stenosis, a marked trans-stenotic pressure gradient, and elevated ICP, venous sinus stenting can effectively eliminate the gradient, reduce intracranial pressure, and lead to significant functional, neurological, and ophthalmic improvement. Patients with a normal body mass index or those aged 50 years and above constitute a relatively small subset of IIH cases, yet they tend to experience more favourable visual outcomes compared with the typical IIH population.

Recommendations

This case underscores the importance of maintaining a high level of suspicion for idiopathic intracranial hypertension whenever ocular or visual field changes are observed, regardless of the patient's age or gender.

Abbreviations

IIH Idiopathic intracranial hypertension

CSF Cerebrospinal fluid

ICP Intracranial pressure

AION Anterior Ischemic Optic Neuropathy

MTHFR Methylene tetrahydrofolate reductase

ANCA-C Anti-neutrophil cytoplasmic antibody (cytoplasmic pattern)

ANCA-P Anti-neutrophil cytoplasmic antibody (Perinuclear pattern)

ESR Erythrocyte sedimentation rate

CRP C-reactive protein

MRI Magnetic resonance imaging

MRA Magnetic resonance angiography

CTA Computed tomography angiography

ICHD-3 International Classification of Headache Disorders, 3rd Edition

ONSF Optic nerve sheath fenestration

Declarations

Ethics approval and consent to participate

Not Applicable

Consent to publish

The authors obtained written informed consent from the patient for publication of any data or images related to the case, with the understanding that these materials may be publicly accessible on the

internet.

Availability of data and materials

All data generated or analysed during this study are included in this published article.

Competing interests

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Authors contributions

AR, NM, and HM conducted the patient interview and clinical assessment, while FY performed the CTA evaluation and placed the venous sinus stent. All authors contributed to writing the manuscript and have read and approved the final version.

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Figures

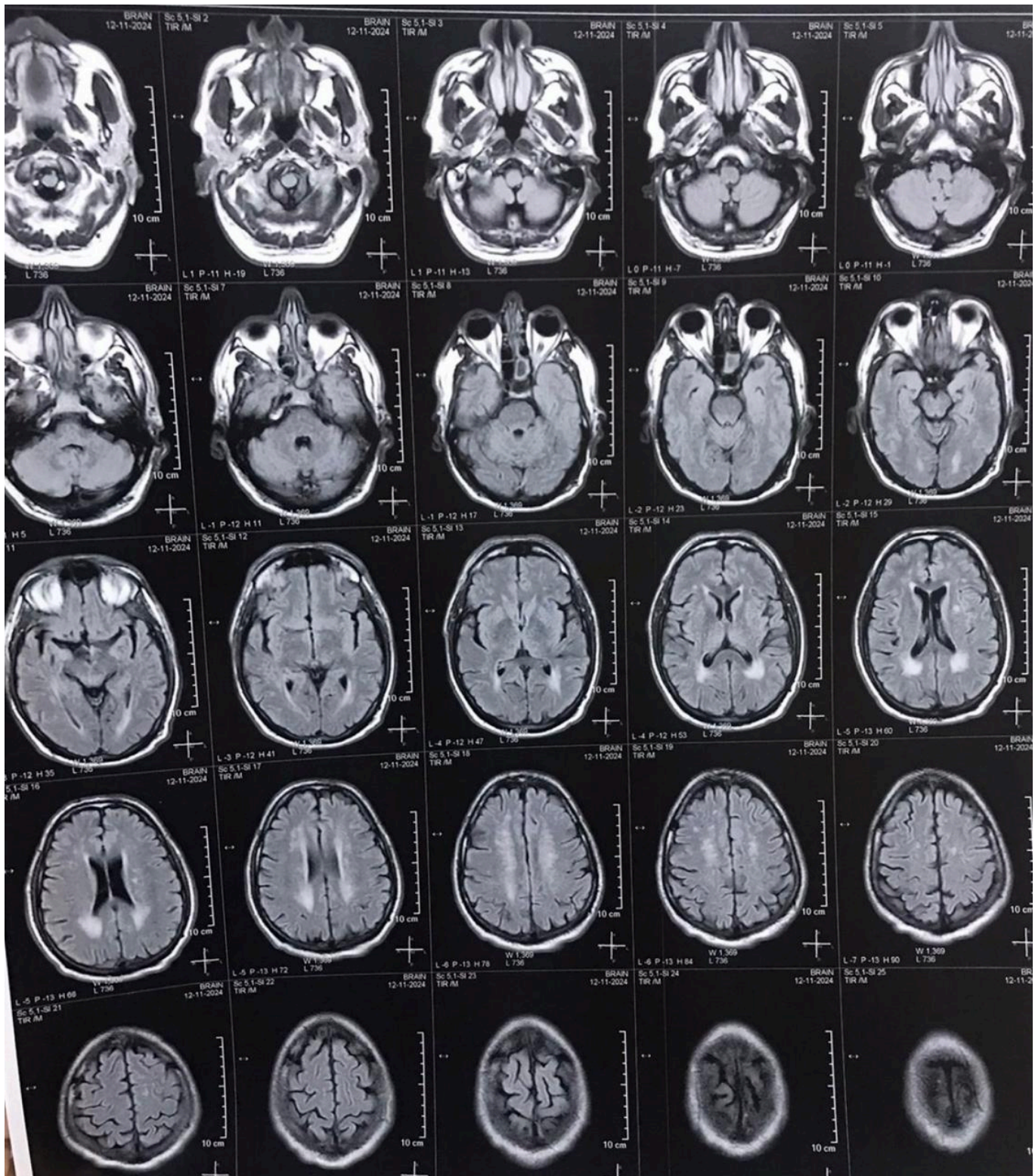


Figure 1

Brain MRI showed bilateral frontal and parietal subcortical ischemic foci, lacunar infarcts, and subcortical arteriosclerotic encephalopathy.

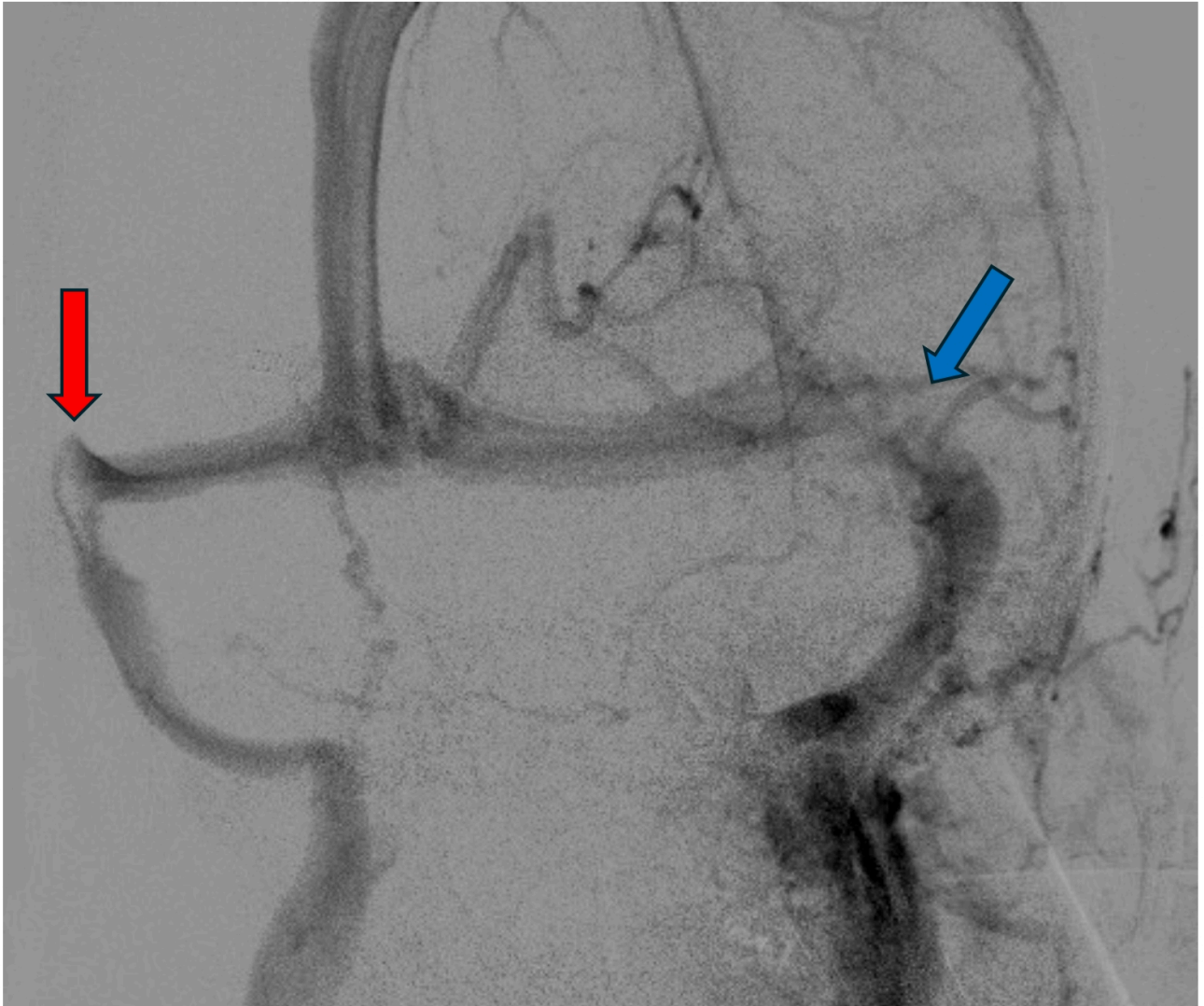


Figure 2

The cerebral angiography showed bilateral transverse sinus stenosis (Red arrow showed left transverse sinus and blue arrow showed right transverse sinus), seen at the junction of the transverse sinus with the sigmoid sinus on both sides.

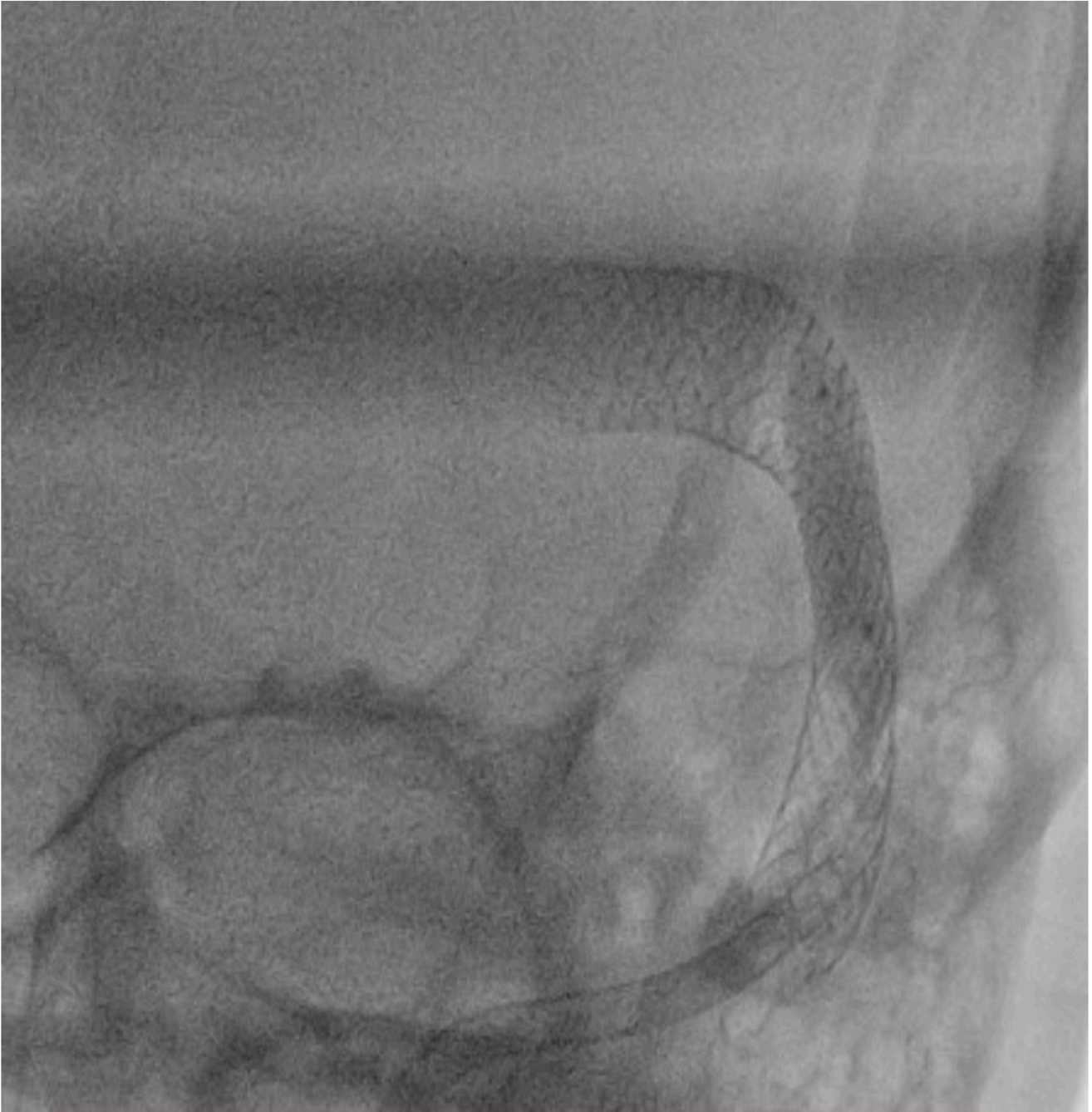


Figure 3

Cerebral Angiography after stenting the left transverse sinus stenosis.