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Intracranial Hypertension as an Initial Presentation of Polycythemia Vera: A Case Report

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Abstract

Idiopathic intracranial hypertension, also known as pseudotumor cerebri, is a common medical condition that mainly affects young obese women. As the name indicates, it is most commonly idiopathic. However, there are many medical conditions that are considered risk factors, associations, or less commonly causative of the disorder. This paper presents a similar clinical scenario in which a middle-aged female patient presents with signs and symptoms suggestive of idiopathic intracranial hypertension, but the condition persisted despite maximum medical treatment until polycythemia vera, a blood disorder, was diagnosed and treated.

Keywords

Pseudotumor Cerebri, Polycythemia Vera, Idiopathic Intracranial Hypertension, Lumbar Puncture, Optic Nerve Sheath Fenestration

1. Introduction

Idiopathic intracranial hypertension, IIH, also termed pseudotumor cerebri, was first described by Heinrich Quincke in 1893. It is characterized by high intracranial pressure without a demonstrable cause. In 2013, Friedman *et al.* proposed the term primary pseudotumour cerebri for the idiopathic form, and secondary pseudotumour cerebri when there is an identifiable cause [1]. Classically, pseudotumour cerebri affects middle-aged obese women. However, a wide spectrum of ages from both genders regardless of their weight may be affected with the disorder. Secondary causes may include medications such as antibiotics (tetracycline, minocycline, doxycycline, nalidixic acid and sulfa drugs), vitamin A, hormones including oral contraceptives, the chronic use of or withdrawal of corticosteroids, and lithium.

Medical conditions associated with pseudotumour cerebri include endocrine disorders, e.g., hypothyroidism and hypoparathyroidism, obstructive sleep apnea, asthma, anaemia, as well as other blood disorders. The most significant risk factor for IIH is weight gain. Although pregnancy is another significant risk factor, it is thought to be due to the associated weight gain and hormonal changes.

Classic symptoms include chronic occipital headache that tends to occur in mornings, pulsatile tinnitus, nausea, vomiting, blurred vision, and in severe cases double vision due to the involvement of unilateral or bilateral sixth cranial nerve palsy. Patients usually will have some of the previously mentioned risk factors. An eye exam would usually be normal apart from Papilloedema, which is the most significant finding. It suggests the presence of fluid around both optic nerves and it may also be evident on Optical Coherent Tomography, OCT of the optic nerve, which demonstrates the thickness of the retinal nerve fiber layer, RNFL. The other key factor to be assessed is visual field defects. The most common visual field defect seen is enlargement of the blind spots. In fulminant or advanced cases, however, the field defect may progress to result in tunnel vision. Once IIH is suspected, neuroimaging should be done. Lumbar puncture, which should be done after a normal neuroimaging to confirm the diagnosis classically reveals elevated CSF pressure and normal CSF composition.

Polycythemia vera is a rare clonal hematopoietic stem cell disorder that was first described in 1892 by Louise Henri Vaquez. Polycythemia vera is known to be a chronic hematological malignancy in which there is an acquired increase in hemoglobin/hematocrit level accompanied by characteristic bone marrow morphology and a JAK2 mutation [2]. The high viscosity impairs the circulation and obstructs the flow of blood through the vessels, causing blood clots. It acts as a switch that triggers blood cell production by the bone marrow. Treatments of polycythemia vera include low-dose aspirin, hydroxyurea, busulfan, interferon Alfa (Intron A, Roberson A), and phlebotomy.

2. Case Presentation

This paper discusses a 48-year-old Asian hypertensive female patient who presented to the neuro-ophthalmology clinic complaining of blurred vision. She also gave a one-year history of left temporal headache associated with postural changes and not relieved by simple analgesics. There was no diplopia, no nausea, no vomiting, and no tinnitus either. Apart from Zestril to treat her hypertension, she was not taking any other medications or supplements, and there were no other medical problems or known allergies. Moreover, she had no past ophthalmic problems. Interestingly, she was not overweight, nor did she have any recent weight gain. Further history did not reveal any other risk factors suggesting IIH.

On exam, she had 20/25 best corrected visual acuity in both eyes. The anterior segment exam was within normal limits. Intra-ocular pressure was 12 mmHg bilaterally and both pupils were round, regular and reactive. The posterior segment exam, however, showed bilateral grade 4 optic disc swelling, blurred margins, and

no cupping. The venous pulsations were not appreciated in either eye. The retina and macula were within normal limits in both eyes.

The visual field test was done. It had fair reliability and had been repeated several times during the management of this case, all of which showed similar results. The right eye showed severe enlargement of the blind spot and inferior nasal field defect, while the left eye showed severe enlargement of the blind spot and peripheral field defects (Figure 1 and Figure 2). These findings suggested that she had IIH. Magnetic resonance imaging including MRI, MR Angiography and MR Venography of the brain and orbit was done. It revealed Papilloedema with bilateral posterior flattening of the sclera, as well as prominent subarachnoid spaces around the optic nerves bilaterally, prominent Meckel's cave, and a partially empty sella (Figure 3). Bilateral transverse sinus stenosis and left sigmoid sinus stenosis were also noted. MRV showed dural venous sinus thrombosis (Figure 4).

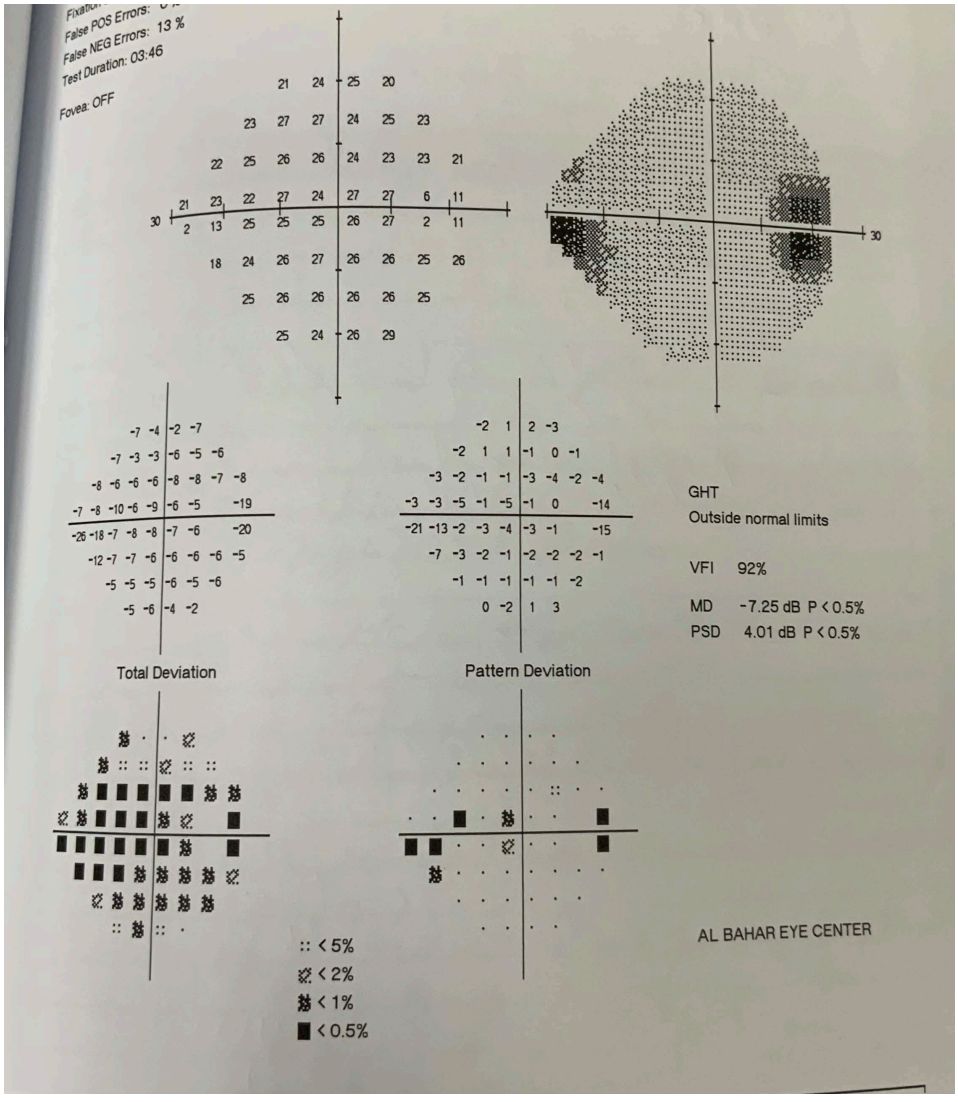

Figure 1. 24/2 Visual field test of the right eye showing enlargement of the blind spot and inferior nasal field defect.

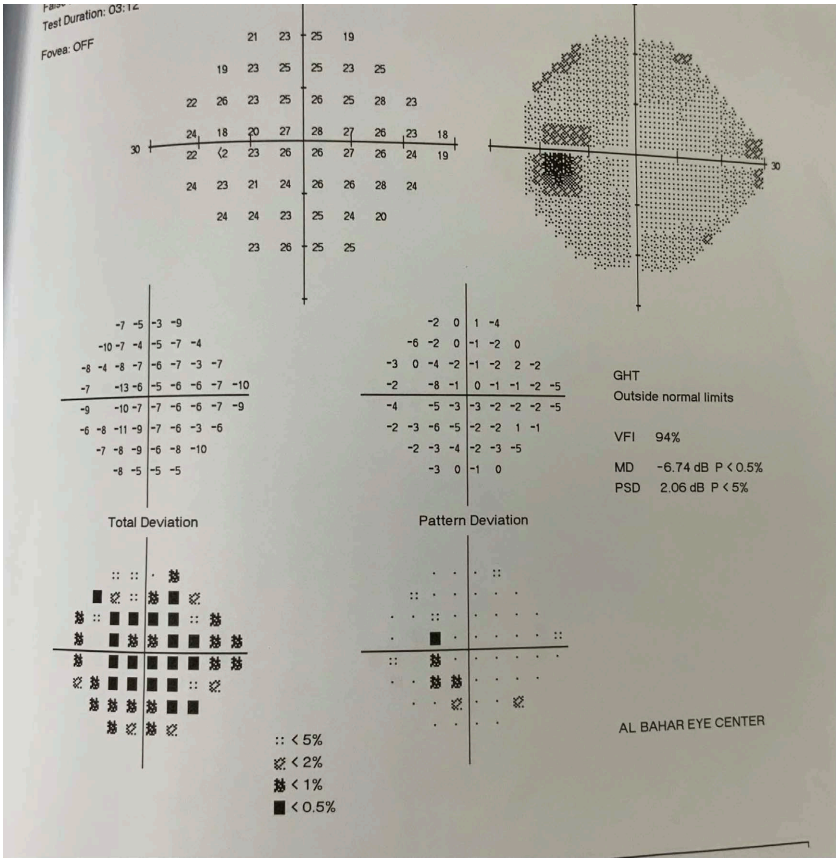

Figure 2. 24/2 Visual field test of the left eye showing enlargement of the blind spot, and mild peripheral field defects.

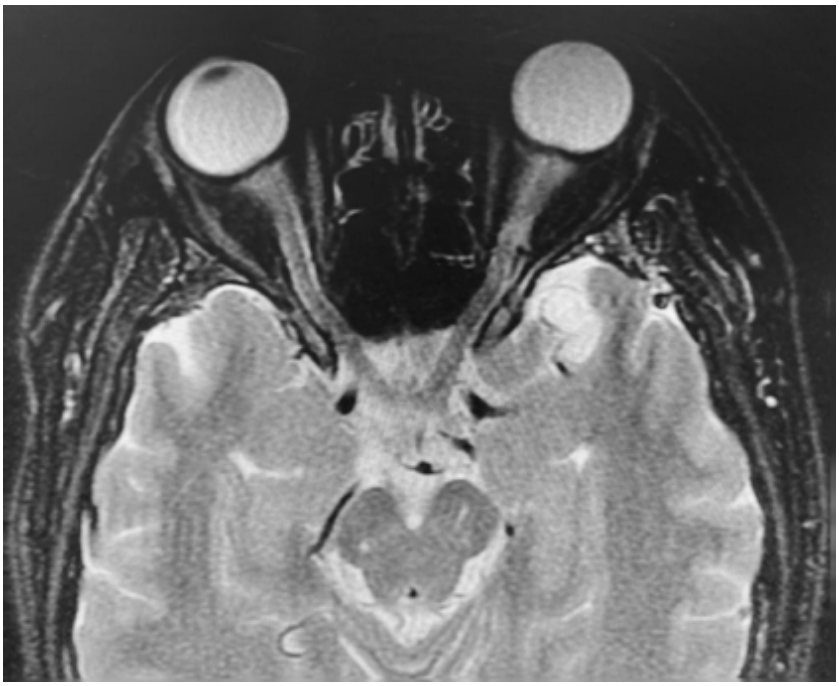

Figure 3. MRI shows CSF around both optic nerves, tortuous optic nerves, and posterior flattening of both globes.

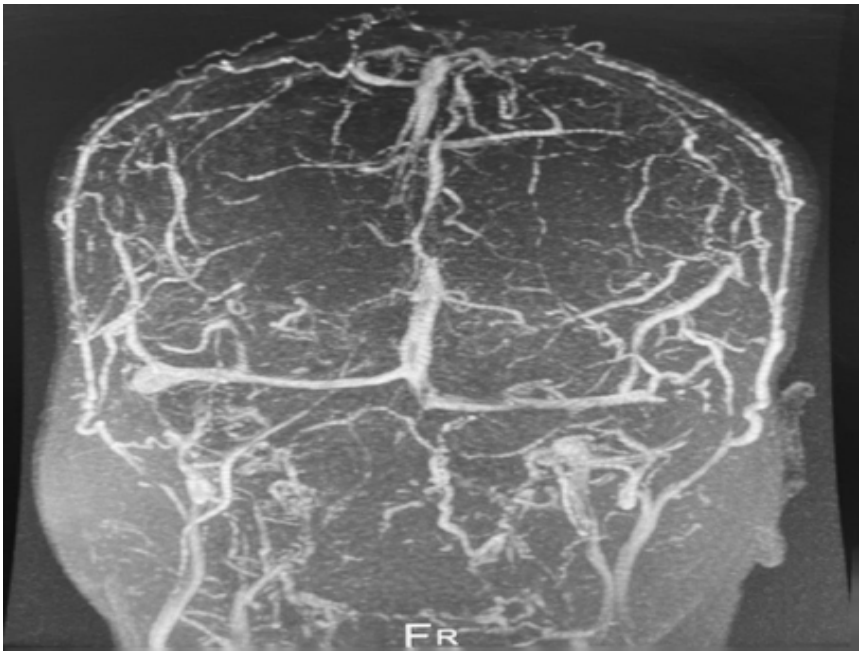

Figure 4. MRV shows bilateral transverse sinus stenosis, left sigmoid sinus stenosis, and dural venous sinus thrombosis.

These findings on neuroimaging supported the presumed diagnosis of idiopathic intracranial hypertension and the patient was thus referred to the neurology team to perform a lumbar puncture, LP, and to treat the dural venous sinus thrombosis. The lumbar puncture had an opening pressure of 310 cmH_2O and a closing pressure of 170 cmH_2O, and the CSF constituents were within normal range. This confirmed the diagnosis of idiopathic intracranial hypertension. Accordingly, Acetazolamide (Diamox) treatment was initiated with a small dose and gradually increased to reach 1.5 g/day.

On blood testing, she was found to have absolute neutrophilia, Anisopoikilocytosis, erythrocytosis and Thrombocytosis. The impression was primarily myeloproliferative disease. Further investigations included an ultrasound of the abdomen, and it showed mild nephropathy with irregular renal contour, suggesting previous illness and scarring. Mild dilatation of the Right pelvicalyceal system was also noted. The rest of the test, however, was within normal limits. She was given a small dose of aspirin until she is fully investigated by Hematology, the consultation of which was requested.

The patient was on tolerable Acetazolamide treatment, namely 250 mg 6 times/day for a duration of six months; however, there was no resolution of the Papilloedema, and there was no improvement in her visual field. Optic nerve sheath fenestration was eventually considered. Luckily, however, she was assessed by the haematology team for further investigations and diagnosis of her abnormal blood test results; a procedure which was time consuming. She was found to have JAK2 mutation and the diagnosis of polycythemia vera was made. Treatment was initiated accordingly. It was noted that the Papilloedema and patient's symptoms did

not improve until Hydroxurea treatment for polycythemia vera was commenced. She also underwent phlebotomy. The Papilloedema gradually resolved over a one-month period and her visual field became normal bilaterally without the need for optic nerve sheath fenestration surgery. Acetazolamide was successfully stopped without any relapse of symptoms. She was stable throughout the follow-up period of 6 months. Of note, during the course of follow-up, the patient at one visit presented with bluish discoloration of her hands and feet (Figure 5 and Figure 6) which was attributed to the polycythemia vera and resolved on treatment.

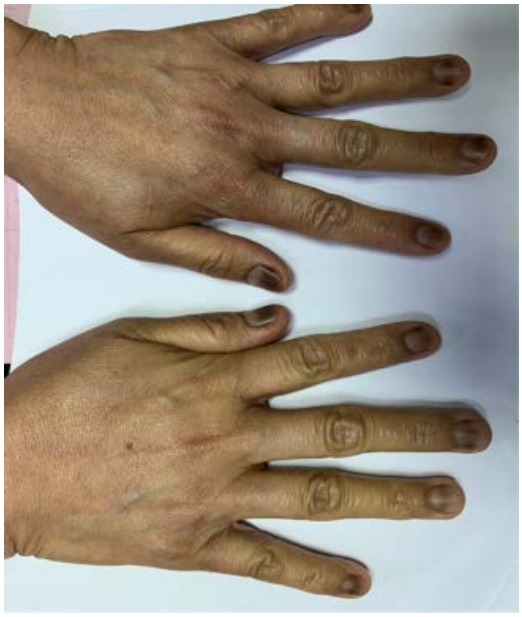

Figure 5. Bluish discolorations of the hands and fingernails.

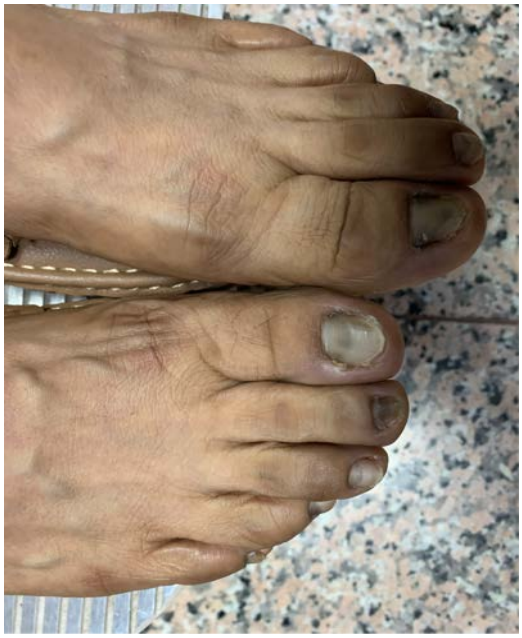

Figure 6. Bluish discolorations of the feet and toes.

3. Discussion

When the aforementioned patient had fulfilled the criteria provided by Friedman *et al.* for the diagnosis of IIH, LP was done despite the lack of risk factors of IIH, to both confirm and consequently lower the intracranial pressure; and although the pressure was indeed high, and lowered by LP, her signs and symptoms persisted despite the maximum tolerable medical treatment. Surgical intervention, along with its possible complication of vision loss was even considered in her case. The abnormal CBC, namely erythrocytosis, neutrophilia, anisopoikilocytosis and thrombocytosis, as well as the neuroimaging findings, however, had necessitated the involvement of the hematology team to explain the thrombosis and the persistent Papilloedema. Indeed, the patient's visual field deficits and the Papilloedema had only resolved after the diagnosis and treatment of the underlying blood condition, namely, polycythemia vera, a condition causing an increase in red blood cells leading to an increase in blood viscosity and consequently the risk of clotting. This in turn leads to CSF disturbance, leading to a rise in the intracranial pressure.

It is of utmost importance to further assess and investigate cases of persistent Papilloedema in patients on full treatment, however uncommon these disorders are in causing IIH, especially treatable ones. This would save countless clinic visits and unnecessary medical treatment and its associated unwanted side effects, let alone invasive surgical interventions. Eshtiaghi *et al.* showed that there is 17.8% of premature or inappropriate diagnosis of idiopathic intracranial hypertension in cases reported in peer-reviewed journals. Such a result highlights the importance of considering other secondary causes prior to labelling a patient as being idiopathic [3]. Waisberg *et al.* demonstrated that a complete blood count is important in cases of Papilloedema and may play a role in reaching a proper diagnosis [4].

Although Optic nerve sheath fenestration, a surgical procedure in which a slit is made in one or both optic nerve sheaths to relieve the trapped CSF around the optic nerves, is essential in certain cases of IIH, it remains an invasive procedure and carries the risk of significant and permanent vision loss. Considering such a procedure should thus only be done in cases where other underlying causes are completely ruled out. In the presence of signs suggesting an underlying disorder causing the high intracranial pressure, such signs shall not be overlooked, especially since such underlying disorders may be treatable. A multi-disciplinary approach may be necessary to diagnose and treat the underlying cause.

Our belief is that IIH patients should be treated in centers where multi-disciplinary teams are available and within reach. We highly recommend including a multidisciplinary team [5] and conducting further tests in cases of persistent Papilloedema on maximum tolerable treatment prior to considering surgical intervention, as attempting such an approach would save time, resources, and most importantly, vision.

4. Conclusion

Idiopathic intracranial hypertension, also known as pseudotumor cerebri, is a common medical condition that affects mostly young obese females. It causes several signs, the most significant of which is Papilloedema. IIH is confirmed by lumbar puncture, which is both diagnostic and therapeutic. Other diuretic medications are also used to treat IIH, while surgical intervention is preserved for fulminant and persistent cases. Although IIH is mostly idiopathic, there are several conditions and risk factors that must be addressed to properly fulfill the management of the condition. Thus, as of this case, I recommend that a complete blood count with differentials be a standard part of the workup for patients with IIH.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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