Clinical Reasoning: A 32-Year-Old Woman With Tunnel Vision and Back Pain

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Abstract

The incidence of new onset visual disturbances in emergency departments across the country is frequent. A detailed history of events and thoughtful physical examination may produce a diagnosis; however, atypical cases may require further diagnostic testing to explain symptoms. We present a case of presumed increased intracranial pressure with atypical findings on diagnostic testing, which allowed our team to explore a broader differential diagnosis. This clinical reasoning article will benefit students, residents, and attendings alike to continue to uncover etiologies for symptoms of increased intracranial pressure and review differential diagnoses in similar presentations.

Section 1

A 32-year-old right-handed woman presented to clinic for bilateral visual disturbances described as episodes of transient tunnel vision lasting seconds. This progressed over weeks to include a persistent bilateral visual impairment described as peripheral visual darkening. The episodes were associated with pulsatile tinnitus and a holocephalic headache without well-defined migrainous features. Her history was otherwise unremarkable except for a 1-year history of lower back pain, which had progressed in intensity and now included intermittent radiation to posterior thighs. She denied any preceding illness, trauma, weight changes, weakness, bladder, or bowel symptoms. Bedside examination was noted for bilateral disc edema and a normal body mass index (BMI). The remainder of the examination including color vision, visual acuity, motility, and confrontational visual fields was unremarkable.

Questions for Consideration:

1. What is the localization of transient visual disturbances?
2. What is the differential diagnosis in this patient with suspected increased intracranial pressure (ICP)?

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Section 2

The patient presented with a transient visual impairment which may localize throughout the afferent visual pathway including orbits, optic nerves, optic radiation, occipital lobes, and systemic or intracranial factors that may affect anywhere along this pathway. The differential for new, transient visual disturbances in a young adult can be broad and includes inflammatory/autoimmune (optic neuritis), infectious (neuroretinitis), structural (aneurysms and tumor), or other etiologies (migraines and idiopathic intracranial hypertension [IIH]). Although one might find it difficult to link the localization of the lower back pain to the visual symptoms, a single unifying lesion might provide a more logical explanation than 2 separate lesions.

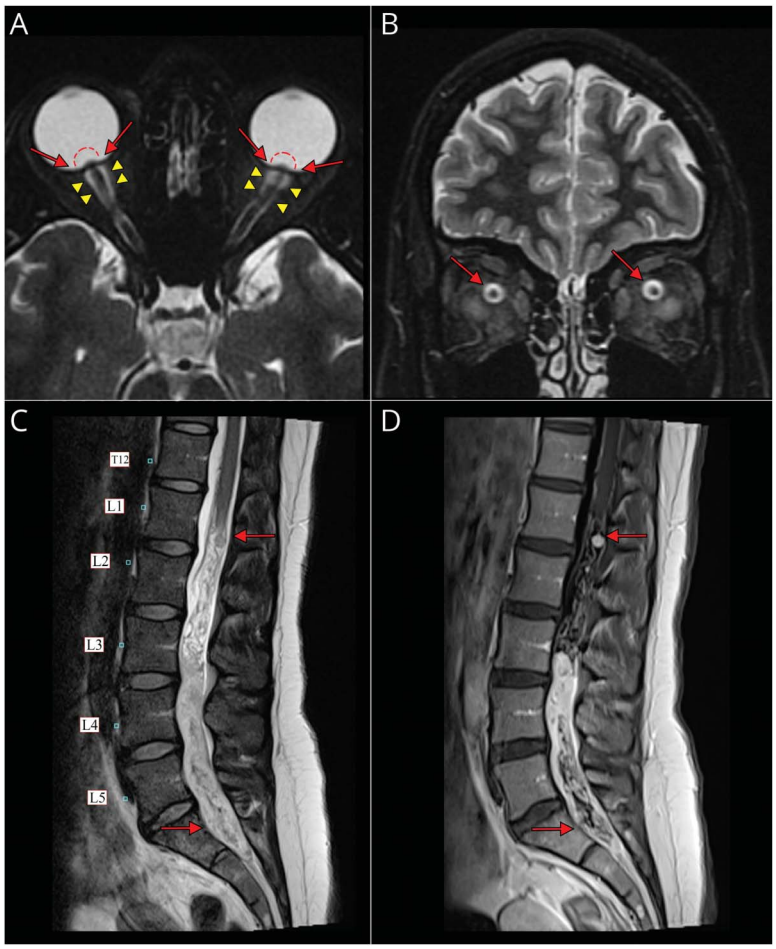
Headache, visual impairment, and pulsatile tinnitus in this population may raise suspicion for IIH, formerly pseudotumor cerebri.[1,2] The differential at this time was broad, and secondary causes of intracranial hypertension must be excluded before IIH diagnosis. These conditions include intracranial mass lesion, obstruction of venous outflow (venous sinus thrombosis or compression), decreased CSF absorption (secondary to CNS infection or hemorrhage), increased CSF production (choroid plexus tumor), medication-induced, and endocrine or metabolic disorders (thyroid disease, Addison disease, etc).[3]

An MRI of the brain with gadolinium performed 4 months later showed mild prominence of the optic nerve subarachnoid space and bilateral intraocular protrusion of the optic nerve (Figure 1, A and B). There were no clear precipitating factors such as weight gain, medications, systemic illnesses, autoimmune, or hematologic conditions that may contribute to symptoms; however, overall history and examination were concerning for intracranial hypertension—warranting further diagnostic testing. The following month, a fluoroscopic-guided lumbar puncture was performed; however, it was unsuccessful after multiple attempts. Repeat neuro-ophthalmologic examination revealed worsening disc edema in both eyes, and initial Humphrey visual field testing showed enlarged blind spots in both eyes (Figure 2A).

Questions for Consideration:

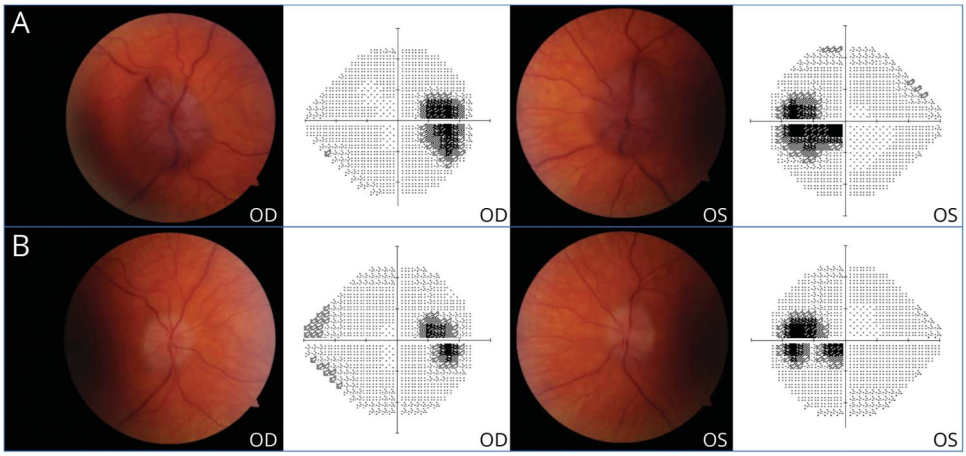
1. What is the next step after failed lumbar puncture?
2. What is the differential diagnosis of spinal cord lesions identified by imaging?
3. What is the management of myxopapillary ependymomas?

Figure 1 MRI of the Brain, Orbits, and Lumbar Region



(A) Axial MRI view highlighting prominence of the optic nerve subarachnoid space (yellow triangles) and bilateral intraocular protrusion of the optic nerve (red arrow and ellipse). (B) Coronal MRI view noting the prominence of the subarachnoid space around the optic nerve. (C) MRI T2 Sagittal and (D) MRI T1 fluid-attenuated inversion recovery postcontrast images of large intradural extramedullary heterogeneous and contrast enhancing mass involving the caudal thecal sac associated with the cauda equina nerve roots from approximately L3-S2.

Figure 2 Fundus Photographs Before and After Surgical Resection of Ependymoma



(A) Fundus photographs preoperatively showing optic nerve edema in both eyes, and the Humphrey visual field test showing enlarged blind spot in both eyes. (B) Fundus photographs 1 month postoperatively showing improved optic nerve edema in both eyes, and the Humphrey visual field test showing resolving blind spot enlargement in both eyes.

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Section 3

Unguided lumbar punctures may be unsuccessful for a variety of reasons (e.g., medical hardware, scar tissue, mass lesions, degenerative spine disease, and body habitus). The rate of failure is approximately 19% for bedside procedures, most notably in patients with higher BMI and ultimately requiring fluoroscopic guidance.[4] Guided lumbar punctures, as in our patient, are less likely to fail. Guided lumbar punctures may not be pursued or fail because of medical instability, patient compliance, body habitus, or pregnancy. Gadolinium-enhanced MRI lumbar-spine was undertaken after the unclear failed lumbar puncture and progressive lower back pain for evaluation of structural disease within the spine. MRI lumbar-spine showed a large intradural extramedullary mildly heterogenous T2 hyperintense mass filling the thecal sac from the level of L3-S2 with innumerable leptomeningeal/intradural enhancing foci (Figure 1, C and D).

The differential diagnosis for spinal cord lesions is extensive with main differentiation between neoplasm and mimics of neoplasm. Spinal cord lesions can be divided between extradural (outside the surrounding dural sac) and intradural (within the meninges), which are further divided into extramedullary (lesions outside the spinal cord) and intramedullary (lesions within the spinal cord). Extradural masses are typically tumor-based, most often metastases from prostate, breast, or lung.[5] Several intradural-extramedullary, non-neoplastic lesions include arachnoid cyst, disc extrusion, infectious, and granulomatous disease.[6] Several intradural-intramedullary, non-neoplastic lesions include vascular malformations (cavernous hemangioma and dural arteriovenous fistula), cord infarction, demyelinating disease (tumefactive MS), and infectious (abscess and tuberculosis). Intradural-extramedullary spinal tumors are most commonly schwannoma and meningioma, while intradural-intramedullary spinal tumors are typically ependymoma and astrocytoma.[6] Ependymomas account for 60% of intradural-intramedullary tumors, of which 10%–15% are myxopapillary ependymoma (MPE), which was ultimately confirmed in our patient (Figure 1, C and D).

She was started on acetazolamide before undergoing neurosurgical resection of the tumor. Intraoperative CSF analysis noted a white blood cell count of 4/mm^3, glucose of 77 mg/dL (serum 128 mg/dL), and protein of 184.1 mg/dL. Owing to the rarity of MPE, annual occurrence of 0.05–0.08 per 100,000 people, nonsurgical management (radiotherapy or chemotherapy) can vary; however, surgery remains the primary treatment with preference for gross total resection as in our patient.[7] On neuro-ophthalmology follow-up 1 month postoperation, there was improvement in the disc edema (Figure 2B). She also reported improvement in her headaches and vision. One-year post-operatively, the patient reports no visual disturbances and her corrected visual acuity is 20/15 in both eyes.

Discussion

Increased ICP may be caused for a variety of reasons. Our patient had many symptoms of increased ICP (headache, pulsatile tinnitus, and transient visual obscurations [TVOs]) which prompted a suspicion of IIH. Particularly TVOs, defined as transient bilateral vision loss for seconds and more pronounced during postural changes, are well described in IIH. The pathophysiology of TVO is uncertain; however, it is believed to be due to transient optic nerve ischemia from disc edema.[8] The current pathogenic mechanism of IIH remains unknown; however, there are proposed etiologies including abnormalities within cerebral venous outflow (venous stenosis or hypertension), increased CSF outflow resistance at the arachnoid granulation, increased abdominal and ICP related to obesity, and abnormalities of vitamin A metabolism.[9]

The diagnosis of IIH remains one of exclusion. Supporting criteria include progressive headaches, evidence of intracranial hypertension (e.g., enlarged blind spot and optic disc edema), and increased CSF pressure (>20 cm H_2O in nonobese individuals and >25 cm H_2O in obese individuals) with normal CSF composition.[3,10] Improvement in headache after withdrawal of CSF further support IIH if other criteria are met.[10] A detailed neuro-ophthalmologic examination is vital to identify and grade evidence of edema. On examination, the most common signs are optic disc edema (typically bilateral and symmetric, although may be unilateral),[11] confrontational visual field abnormalities (nasal or temporal loss with visual blurring), and unilateral or bilateral abducens nerve palsy.[3] It is important to note that papilledema is reserved for patients with optic disc edema because of raised ICP. Because optic edema is a meaningful criterion for IIH and evaluation of ICP, one should also consider other causes of optic disc swelling including malignant hypertension, diabetic papillopathy, pseudopapilledema, neuroretinitis, sarcoidosis, and ischemic optic neuropathy.

In patients with suspected elevated ICP, evaluation with neuroimaging is essential. MRI of the brain with gadolinium including MR venography and orbits is the preferred test for evaluating secondary causes of elevated ICP. Brain parenchyma and ventricles will appear normal in IIH; however, subtle abnormalities including distention of perioptic subarachnoid space, flattening of the posterior sclera, and intraocular protrusion of the prelaminar optic nerve can be seen, as in our patient (Figure 1, A and B). Further findings can include tortuosity of the optic nerve, enhancement of prelaminar optic nerve, empty sella, narrowing of the transverse venous sinus, and mild tonsillar herniation.[12] Once neuroimaging and ophthalmologic evaluation have been completed, lumbar puncture must be undertaken to assess opening pressure and CSF composition, which are elevated and normal in IIH, respectively. If the lumbar puncture fails, patients exhibit myelopathy symptoms, or CSF profile is abnormal in cells or protein, spinal imaging with gadolinium-enhanced MRI is recommended.

There are no data estimating incidence of spinal tumor and ICP.[13] Since 1970s, there have only been 29 reported cases of increased ICP without hydrocephalus in the setting of a spinal cord tumor; 28 of 29 cases presented with evidence of optic disc edema.[14] The authors further clarify that patients with

disc edema who also have neurologic signs or symptoms of myelopathy or elevated CSF protein, particularly in patients with an atypical demographic for IIH, should raise suspicion for a spinal tumor thus prompting spinal imaging and neurosurgical evaluation.

The pathophysiology of increased ICP in patients with spinal tumors has been hypothesized. Some authors believe that the elevation in CSF protein decreases CSF absorption at the arachnoid villi, although elevated protein levels may not always be identified in spinal tumors.[15] The more supported hypothesis involves the release of tumor-generated proteins and inflammatory markers that hinder CSF absorption, such as fibrinogen or transforming growth factor β, which may lead to hydrocephalus or increased ICP.[15]

Although there are rare reports of symptom resolution with medical management alone,[14] surgical intervention, chemotherapy, and/or radiotherapy typically are required to decrease CSF protein burden and pressure without the need for a prolonged medical treatment course.[14] Similar to our patient, most patients' papilledema resolved and visual symptoms improved after resection of the spinal tumor.[14]

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