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Cerebral Hemosiderin Deposits On Magnetic Resonance Imaging In A Patient With Pseudopapilledema

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Abstract

Pseudopapilledema is apparent optic disc swelling that emulates characteristics of papilledema but is usually secondary to multiple underlying causes. We review an interesting case in which a developmentally delayed male is worked up for abnormal appearing optic nerves and upon brain imaging is found to have significant unique findings. Physical examination showed no focal findings, however the optic disc bilaterally appeared elevated with blurred margins. A subsequent MRI illustrated hemosiderin deposition in the bilateral cerebral convexities. Humphrey Visual Field testing demonstrated an enlarged blind spot; however a lumbar puncture performed by a neurologist had normal opening pressure. Several hypotheses for the patient's radiographic findings are discussed; chronic causes such as leaky microanerysms, subdural hematoma, and recanalized venous thrombus or neonatal causes such as traumatic birth injury and shaken baby syndrome. Due to the social dynamics of the patient and a Department of Children and Families investigation which revealed an unsafe home environment, it was hypothesized that such findings were secondary to neonatal traumatic injury, likely due to shaken baby syndrome. A management plan for the patient involves annual follow up with routine visual field testing.

INTRODUCTION

Pseudopapilledema in the context of cerebral hemosiderin deposition has not been presented in the literature. We review an interesting case in which a developmentally delayed male is worked up for abnormal appearing optic nerves and upon brain imaging is found to have hemosiderin deposition in the frontal cerebral convexities. We summarize possible etiologies of such findings and discuss management follow-up plan for such patients.

CASE REPORT

A healthy 16 year old patient with mild developmental delay was examined by a neuro-ophthalmologist for irregular and swollen optic nerve heads bilaterally. The patient, a 16 year old healthy Caucasian male was initially seen by a referring ophthalmologist who recommended additional evaluation of the optic nerve by a specialist. The patient denied headaches, nausea, vomiting, or any visual changes including changes to visual acuity, color vision, or any horizontal/vertical diploplia. A previous CT scan failed to identify a cranial mass or cerebral edema to account for the suspected elevated intracranial pressure.

Initial examination of the optic nerve illustrated adequate color vision function using the Ishihara color tiles (12/12 bilaterally). Visual acuity was measured at 20/25 OU without correction, and tonometry by applanation measured intraocular pressures of 10 and 11, well below the upper range of normal. Pupils were equally reactive to light and accommodation, brisk, with no observable relative afferent papillary defect. Both confrontational visual fields and extraocular movements were full. Slit lamp examination of the lids and lashes showed no meibomian gland dysfunction, a white and quiet conjunctiva and sclera, a clear cornea, and deep and quiet anterior chamber. The iris was round, and the lens was clear.

An evaluation of the optic disc illustrated significant findings in both eyes. Specifically, the optic discs lacked a true physiologic cup and appeared tilted inferiorly. The nerve margins were blurred and had an elevated appearance. It was difficult to differentiate whether the appearance of the patient's optic nerve was secondary to intracranial pathology or a congenital anomaly.

Due to the patients lack of prominent visual or intercranial symptoms an MRI was initially deferred and the patient was followed up one month later to undergo Humphrey visual field testing and a dilated fundus exam. On follow-up, the

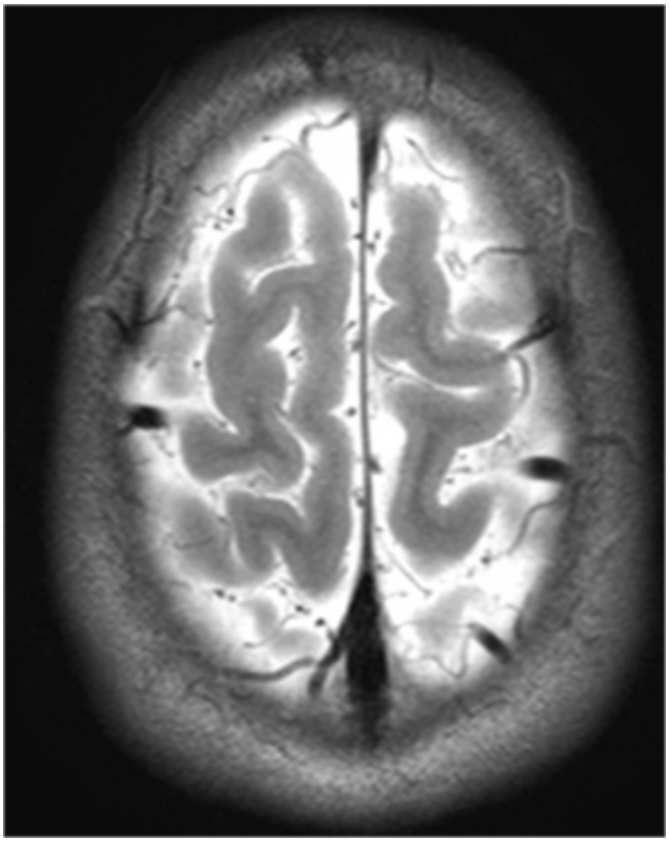
patient's visual field examination illustrated an enlarged blind spot but no arcuate or scotomatous changes. The patient denied any recent headaches or changes in visual acuity, however, complained of an unrelated left lower lid painless nodule for the past 2-3 weeks. Physical examination showed a pigmented sessile lesion on the left lower lid margin involving the meibomian gland. The slit lamp examination was unchanged from previous visit, and the optic discs continue to have irregular margins with minor protrusion bilaterally.

Eventually, an MRI with Gadolinium was performed and identified significant subarachnoid venous dilation and widespread punctuate hemosiderin deposition in the subarachnoid sulci. The frontal, parietal and occipital convexities were most prominently affected (Figure 1). Susceptibility weighted imaging for iron storage confirmed iron deposits throughout the cerebral convexities with prominent dilated subarachnoid venules. (Figure 2). The dural sinuses, including the superior sagittal sinus, appeared patent. There was no evidence of arteriovenous malformations, aneurysms, or intraparenchymal hemorrhage. A benign incidental pineal gland cyst measuring 9.8 mm was acknowledged. Due to the unknown etiology of the hemosiderin deposits, the patient was referred to a neurologist who performed a lumbar puncture. Lumbar puncture was unremarkable for xanthrochromia, red blood cells, or leukocytosis and included a normal opening pressure of 190mm H2O.

Of note, a Department of Children and Families investigation performed by the state for an non-related incident revealed a harmful home environment for the child and significant signs of abuse and neglect by the parents ultimately requiring legal separation. Temporary custody of the patient was eventually awarded to the minor's grandmother.

Figure 2

Figure 2. Susceptibility Weighted Imaging of patient's brain.



There is significant abnormal hemosiderin deposition with tortuous, dilated subarachnoid veins. There is no sign of cerebral atrophy, or intracerebral hemorrhage.

{image:2}

This technique produces an enhanced contrast magnitude image which detects significant hemosiderin deposition along bilateral cerebral convexities. SWI images are sensitive to venous blood, iron, and hemorrhage.

DISCUSSION

Asymptomatic pseudopailledema in the context of abnormal hemosiderin deposition in the subarachnoid sulci has not been presented in the literature. Pseudopapilledema is a condition in which the optic nerves appear swollen during fundoscopic examination, but intracranial pressure is within normal range. This is most commonly caused by optic disc drusen which are lipid micro globules that calcify in the optic disc resulting in blurriness of the nerve margin. Other less frequent causes of pseudopapilledema include abnormal myelination of the optic nerve, severe hyperopia, and congenital abnormalities [1-Rosenberg]. Pseudopapilledema rarely causes visual symptoms but may show atypical

findings on visual field testing such as enlargement of the blind spot, and inferior nasal bundle defects [2-wall]. In contrast to papilledema, the nerve heads of pseudopapilledema are not hyperemic and do not have prominent telengectasia or capillary dilation. Spontaneous venous pulsations are also visible on dilated fundoscopic examination, an important characteristic which helps distinguish from papilledema [3 - Levin]. Furthermore, disc margins will appear to be obscured yet vessels obscuration is usually not seen.

The presence of hemosiderin deposits on T2-weight MRI have been identified in patients' with history of subarachnoid hemorrhage, leaky aneurysms or arteriovenous malformations(AVM), and from neoplasm of the dural coverings[4-Horita]. The mechanism of hemosiderin deposition in the subarachnoid space may involve leakage of red blood cells from hemorrhage (or unruptured aneurysms) and subsequent destruction and uptake of iron by macrophages [4-Horita]. Patients with significant hemosiderosis such as after a subarachnoid hemorrhage, intracerebral hemorrhage, or trauma may even develop secondary hydrocephalus because the deposition may directly obstruct cerebrospinal flow or impair absorption by the arachnoid granulations. The arachnoid granulations contain venules which drain CSF and become clogged with residual red blood cells. The hemosiderin deposits are readily picked up by T2 low intensity MRI and may remain visible one year after the inciting event [5-Takada]. These patients, however, will usually present with pain, and altered mental status. Hemosiderin deposition may be the first sign of a leaky avascular malformation, cyst or aneurysm. It is important to note that numerous other things may mimic the hypodense areas on T2 weighted imaging. For instance, calcifications, ferritin, and melanoma may appear identical to each other based solely on imaging [5 Takada]. Radiologic findings should always be taken in context of the clinical situation.

There are several hypotheses that may describe this patient's abnormal MRI findings. The normal lumbar puncture findings, including opening pressure, may suggest a chronic, congenital or neonatal etiology. Leakage of blood from the incidental pineal cyst is one possibility; however a more thorough diagnostic work-up would be required to confirm this. Another possibility that may describe the patients MRI findings includes a previous thrombus located within the right sigmoid sinus. This could account for an elevated intracranial pressure and subsequent papilledema that left the optic nerves slightly elevated once the intracranial pressure was normalized with recanalization.

However, a more likely hypothesis for the MRI findings may be due to cranial trauma endured by the patient either during or after birth. In the context of the patient's psychosocial dynamics, home environment, and associated developmental delay it was determined that the findings were suggestive of abuse in the form of shaken baby syndrome or traumatic birth injury. Unfortunately, shaken baby syndrome is a common cause of subdural hemorrhage and developmental delay in infants, however acute and chronic subdural hemorrhage on the basis of T1 and T2-weighted magnetic resonance imaging is often difficult [6 Lee]. Other pathognomonic ocular findings of shaken baby syndrome including retinal hemorrhages and detachment usually heal on their own and leave no residual findings on fundoscopic examination. Birth trauma, including chronic hemorrhage has been reported as a cause for hemosiderin deposition in the subarachnoid sinuses.

The patient's pseudopapilledema findings may be an independent factor in this patient and one which should require annual follow-up. Specifically, regular Humphrey visual field testing should be performed to evaluate for changes in vision. Due to the patient's asymptomatic presentation, additional radioimaging studies are not recommended.

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