

High-Pressure Normocephalus-Raised Intracranial Pressure with False Localizing Signs

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Abstract

Normal-sized ventricles and absence of papilledema do not rule out shunt failure and raised intracranial pressure (ICP). Raised ICP can present with false localizing signs which may be cranial nerve palsies or extensive polyradiculopathy. Our patient with a history of ventriculoperitoneal (VP) shunt presented with rapidly progressive vision loss without papilledema, as well as multiple cranial nerve palsies and radiculopathy. Imaging did not reveal hydrocephalus, however, cerebrospinal fluid (CSF) manometry revealed high CSF opening pressure. After lumbar thecoperitoneal shunting, vision did not improve, but the rest of cranial nerve palsies and radiculopathy improved. In a patient in whom VP shunt is *in situ*, headache and vomiting should prompt evaluation for raised ICP though there is no ventriculomegaly of papilledema. Vision can be saved if raised ICP is suspected, CSF opening pressure measured at presentation and prompt surgery is performed.

Keywords: False localizing signs, raised ICP-raised intracranial pressure, shunt failure

WHAT IS KNOWN?

1. In a patient in whom ventriculoperitoneal shunt is *in situ*, headache and vomiting should prompt evaluation for shunt malfunction and raised intracranial pressure (ICP)
2. Vision can be saved raised if the diagnosis is suspected and cerebrospinal fluid opening pressure measured at presentation.

WHAT IS NEW?

1. Normal-sized ventricles and absence of papilledema do not rule out shunt failure and raised intracranial pressure
2. Shunt failure can present with false localizing signs which may be cranial nerve palsies or extensive polyradiculopathy.

INTRODUCTION

Ventriculoperitoneal shunts are commonly inserted for the treatment of raised intracranial pressure. Shunt failure is a common complication and early diagnosis and treatment can significantly improve the outcome. Failure to diagnose shunt failure can lead to significant deficits which may be irreversible.

CASE REPORT

This 11-year-old girl presented with headache for 3 weeks, weakness of all the four limbs for 2 weeks, and rapidly progressive vision loss for 10 days. At 16 months of age (10 years back), she underwent fenestration of posterior fossa arachnoid cyst with ventriculoperitoneal (VP) shunt insertion when she had become lethargic and lost motor milestones. She improved and returned to baseline and was doing fine.

Three weeks before presentation to our center, she developed persistent frontal headache and vomiting which progressively increasing. On the 7th day of illness, she was evaluated elsewhere by an ophthalmologist. Visual acuity was reportedly normal. There was no papilledema. Magnetic resonance imaging (MRI) was done, which showed a posterior fossa arachnoid cyst with VP shunt *in situ* and normal-sized ventricles [Figure 1]. Cerebrospinal fluid (CSF) examination revealed acellular CSF with normal biochemistry, but opening pressure was not measured. She also had aching over the limbs. Headache decreased after the CSF drainage (25 mL).

During hospitalization, she developed symmetrical proximal as well as distal weakness of all the four limbs without sensory loss. She also developed difficulty in chewing and jaw drop without facial sensory loss. There was left-sided facial, palatal, tongue, and neck weakness. Eleven days after the onset of headache, she developed horizontal binocular diplopia. Then, she started having diminution of vision in both the eyes, which progressed over 5 days to complete blindness with worsening of headache. There were no seizures, ataxia, fever, and loss of weight or appetite. There were no other systemic complaints. She received pulse methylprednisolone with no improvement.

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At presentation at our center (3 weeks after onset of illness), she was conscious, drowsy, and had a normal general physical examination and hemodynamically stable without any features of Cushing's triad but had neck stiffness.

Examination revealed absent perception of light bilaterally without papilledema. Pupils were dilated and not reacting to light, with ptosis of the right eyelid with bilateral impaired abduction of the eyeballs. Facial sensation was normal with diminished corneal reflex bilaterally, with jaw weakness. There was no facial weakness, palatal, or tongue weakness. Neck flexors as well as extensors were weak.

There was symmetrical flaccid quadriparesis. All deep tendon reflexes were absent and plantars were extensor. Sensory examination was normal. Finger–nose test was normal and tandem walking could not be evaluated.

Routine investigations were normal. MRI of the brain was done [Figure 1]. Lumbar puncture showed elevated CSF pressure >40 cm CSF. CSF was acellular with sugar of 63 mg/dL (corresponding Random blood sugar (RBS) of 87 mg/dL), protein was 26 mg/dL. Other investigations such as CSF culture, acid–fast bacillus staining, malignant cytology, cryptococcal antigen, and GeneXpert were negative. After CSF tapping of 50 mL, her weakness and cranial nerve deficits except vision improved and she also had relief in headache.

Visually evoked potentials were not recordable. Nerve conduction study was suggestive of radicular involvement.

Clinically, she had a meningeal process with contiguous involvement of the cranial nerves on the left side, bilateral 6th nerve involvement as well as polyradiculopathy. Possibility of chronic meningitis, shunt malfunction, or other causes of raised intracranial pressure (ICP) such as chronic sinovenous thrombosis was considered. ICP rise due to idiopathic ICP or drug-induced causes were ruled out because modified Dandy's criteria for IIIH were not fulfilled. Moreover, since the shunt was already *in situ*, if it was functional, the patient was unlikely to be having features of raised ICP. Thus, shunt malfunction was diagnosed.

Treatment and follow-up

Since the patient had loss of vision as well as multiple deficits, lumbar thecoperitoneal shunt (LP shunt) insertion was performed to relieve the raised ICP immediately. LP shunt was preferred in this case over VP shunt revision, as the shunt revision was technically difficult in normal-sized ventricles, and our center has a large experience in LP shunting in patients with malignant idiopathic intracranial hypertension.

After surgery, patient's limb power improved. She also started walking on her own, and all other cranial nerve deficits got relieved, except for vision loss which was persistent. Subsequently, lumbar puncture was repeated,

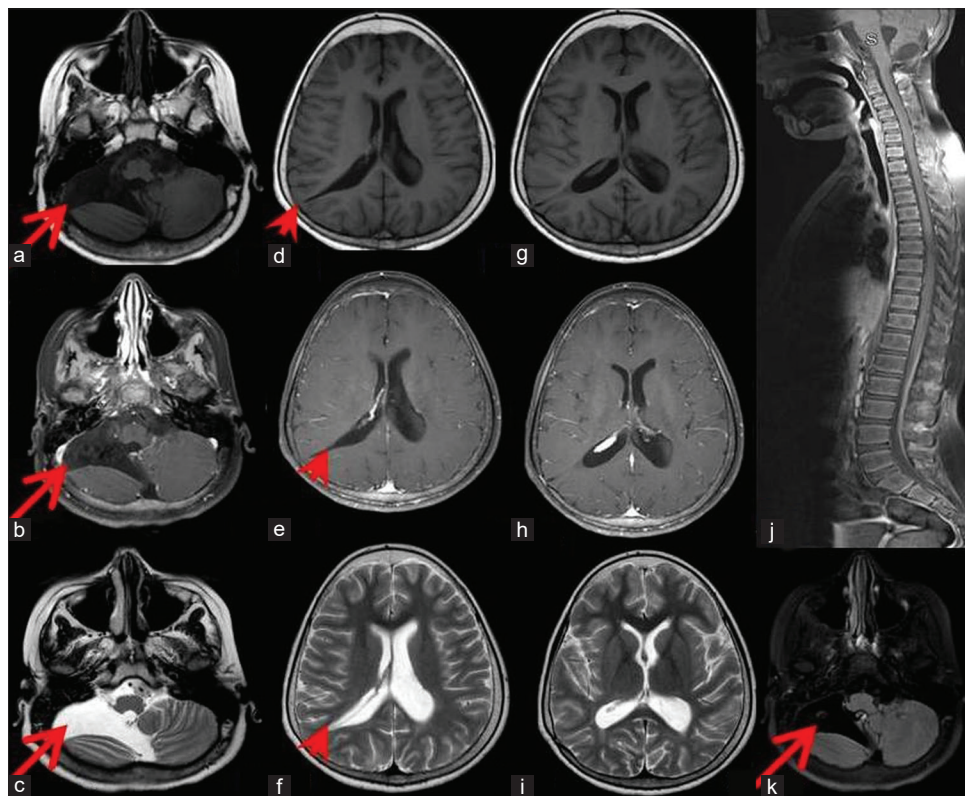


Figure 1: (a-c) Right posterior fossa arachnoid cyst (red arrow) which is leading to distortion of the brain stem. (d-f) Red arrow head shows VP shunt *in situ*. (b, e, h and j) There is no contrast enhancement on T1 post contrast images. (k) Fluid-attenuated inversion recovery image shows no brainstem lesions [(a, d and g) shows T1 axial; (b, e and h) show T1 axial with contrast; (c, f and i) T2 axial images. (j) Shows sagittal T1 contrast spine imaging]

which showed CSF opening pressure of 5 cm CSF and was clear and normal biochemically. Even after 1 year of follow-up, there was no improvement in vision. Vision did not improve probably due to the long duration of symptoms before intervention was done.

DISCUSSION

Shunt malfunction

Long-term follow-up of shunts have revealed that around 40% of shunts malfunction at 1 year and 50% at the 2nd year.^[1] Shunt failure presentations can take a variety of forms including headache, new onset of nausea, vomiting, irritability, fever, or altered sensorium. Examination may reveal papilledema, cranial nerve palsies (most commonly CN VI), hyperactive reflexes, and ataxic gait.

Shunt malfunction without hydrocephalus

Although imaging is routinely employed in diagnosis of shunt malfunction, patients may present with no evidence of hydrocephalus on imaging.^[2,3] Thus, the absence of hydrocephalus should not be used to rule out failure of the shunt and raised ICP when the clinical suspicion is high due to other features such as headache or cranial nerve palsies.

False localizing signs

“Neurological signs have been described as ‘false localizing’ if they reflect dysfunction distant or remote from the expected anatomical locus of pathology, hence challenging the traditional clinicoanatomical correlation paradigm on which neurological examination is based.”^[4]

False localizing signs have been reported in relation with raised ICP of which 6th nerve palsy is the most common, and there are reports of ophthalmoplegia, 5th and 7th nerve palsies, trigeminal neuralgia, hemifacial spasm, and hearing deficits.

Raised ICP can cause dysfunction of various structures in the neuraxis. Our patient had multiple cranial nerve dysfunction, 2nd, 3rd, 5th, 6th, 7th, 9th, 10th, 11th, and 12th cranial nerves. Furthermore, there were features of radicular involvement. Previous cases have reported radiculopathy in raised ICP due to other causes.^[5,6] However, this is the first report of shunt malfunction-related raised ICP with these features.

Raised intracranial pressure without papilledema

Our patient did not have papilledema, though she had complete vision loss. Studies have shown that in children with hydrocephalus and raised ICP up to 41% may have no papilledema.^[7] However, there are no cases which have reported vision loss in raised ICP without papilledema. Moreover, this patient had normal-sized ventricles, suggesting a “high-pressure normocephalus sans papilledema.”

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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