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Case Report

Can Visual Complaints be Only Symptom in Slit Ventricle Syndrome? A Difficult Diagnosis to be Made

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Abstract

Background: Slit ventricle syndrome (SVS) is a rare symptomatic condition with radiological findings of small sized ventricles in a case of VP Shunt. Diagnosis may be difficult because radiological imaging of small ventricles can be misinterpreted as normal sized ventricles. SVS is a rare condition of reduced brain compliance with intermittent intracranial hypertension.

Cases: Author reported two cases of ventriculoperitoneal (VP) shunted patients with only visual disturbances which on evaluation were found to be SVS.

Conclusion: SVS is a difficult diagnosis to be made based on laboratory investigations, clinical scenario and imaging. Presence of visual symptoms in shunted patients should not be ignored, closely watched and evaluated for raised ICP as they can be due to SVS.

Keywords: Slit Ventricle; Shunt Malfunction; Over-drainage; Ventriculoperitoneal (VP) Shunt

Introduction

A Slit ventricle syndrome is a condition used to describe intermittent severe headaches following ventriculoperitoneal shunt for hydrocephalus [1,2]. VP shunt is most common treatment of hydrocephalus. VP shunt itself can lead to lot of complications. Infection, malfunction, obstruction and over-drainage of cerebrospinal fluid are most common complications. 5-55% of shunted patients have over-drainage of CSF [3]. Headache, nausea, vomiting, disturbances of consciousness, seizures, hemiparesis and rarely visual disturbances can be the main clinical presentations of these patients [3].

Becker and Nulsen first described this syndrome (1960) and stated that it could be due to inadequate CSF drainage secondary to small ventricles in shunted patients [4]. Most of these patients are asymptomatic, and require treatment when they suffer from severe headache. Radiological examination may reveal normal or slit ventricles. Accurate diagnosis may be difficult and adequate management may be delayed because neuro-radiology can be misleading as normal working shunt [5].

Patients with slit ventricles are prone to have further shunt complications as SVS is itself a complication of cerebrospinal fluid (CSF) shunting procedure. Exact cause of SVS and its pathogenesis is still poorly understood. Author reported two cases with visual disturbances with diagnostic difficulties which later turned out to be SVS.

Case Reports

A 14 year male child admitted to outdoor department with chief complaints of visual blurring. The child underwent ventriculoperitoneal shunt at the age of 1 year for congenital hydrocephalus. Computerised tomography (CT) of head did not show any ventriculomegaly or other signs of raised intracranial pressure with VP shunt in situ. Ophthalmological examination revealed optic disc swelling with visual acuity of 20/20 in right eye and 20/25 in left eye. Patient did not consent for indoor admission and was advised to follow up after 2 weeks. The patient again presented after 2 weeks with increased visual blurring with CT head showing no ventriculomegaly or any signs of raised intracranial pressure (Figure 1a). However, fundus examination (Figure 1b) revealed severe optic disc swelling with exudates as compared to previous examination. Patient was planned for ICP monitoring which revealed high ICP and SVS was suspected. Patient was planned for shunt revision and symptoms improved following shunt revision. Patient 's visual acuity gradu-

ally improved and regained almost complete visual acuity within 6 months.

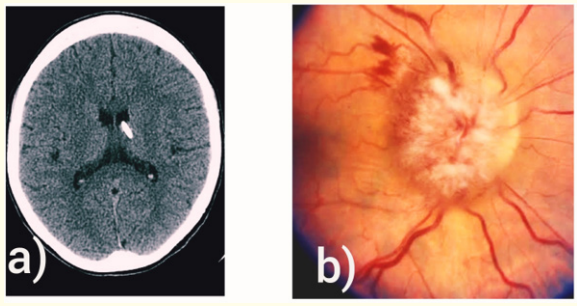

Figure 1: a: Computerised tomography (CT) of head suggestive of normal looking ventricles with VP shunt in situ with no ventriculomegaly or flattened sulci or enlarged subarachnoid space suggestive of raised ICP.

Figure 1 b: Fundus examination revealed severe optic disc swelling with exudates.

Another 8 year female child presented with atypical features of visual blurring and diplopia for last 10 days. The child underwent VP shunt for congenital hydrocephalus at 18 months of age. Patient had no features of headache, vomiting, seizure, or loss of consciousness. Patient had a visual acuity of 20/40 in right and 20/20 in left eye. Fundus examination revealed optic disc swelling with increased tortousity of vessels. CT head revealed no ventriculomegaly with nearly normal looking ventricles with VP shunt in situ (Figure 2a). After 1 week, visual blurring had increased and fundus examination revealed severe optic disc swelling (Figure 2b). Patient again went for CT head which was almost similar to previous one with no signs of ventriculomegaly or raised ICP. Patient was subjected to ICP monitoring and revealed raised ICP. Shunt revision was planned in view of suspected features of SVS and visual symptoms improved following shunt revision. Over a period of 6 months, visual acuity and diplopia had improved gradually.

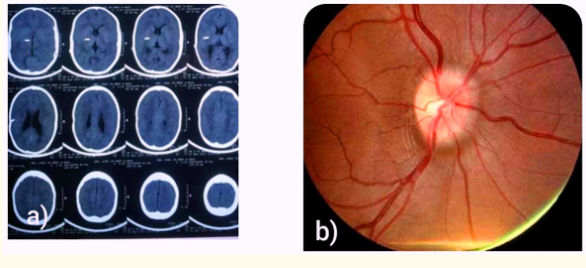

Figure 2: a: CT head revealed no ventriculomegaly with nearly normal looking ventricles with VP shunt in situ.

Figure 2 b: Fundus examination revealed severe optic disc swelling.

Discussion

The term SVS refers to intermittent presence of headache, vomiting, some degree of visual disturbances, impaired consciousness in patients of hydrocephalus with shunts [6]. However pathogenesis related to SVS is still not clear. These patients have generally intermittent severe headache, normal sized ventricles on radiology and slow refilling valve mechanisms.

However, recent studies suggest that lack of dilation of ventricles is due to reduced brain compliance [7]. Most of patients with SVS does not develop any clinical symptoms, and remain asymptomatic during their lifetime. However, symptoms of SVS are common in young children with normal size[1] or enlarged ventricles [8].

Rakate., *et al*. [9,10] described clinical presentation of patients with frequent shunt headaches:

- Intermiitent malfunction of shunt associated with higher pressure.
- Raised ICP with functioning shunt described as cephalocranial disproportion.
- Raised ICP with malfunctioning shunt in normal pressure hydrocephalus.
- Low ICP correlated with shunt over-drainage
- Headache not associated with shunt function uncommonly related to migraine, chronic headache and headaches relieved with rest.

Various hypothesis have been proposed to explain pathophysiology in slit ventricle syndrome. With shunt malfunction, ependymal wall becomes coapted to shunt catheter openings, causing transient obstruction leading to elevation of ICP. Decreased brain parenchyma compliance with variations to cerebrospinal fluid volume have been proposed as one of mechanism. Chronic shunting lead to periventricular gliosis which may lead to inability of ventricles to dilate[11]. There is evidence that decreased brain compliance does not occur but rather there is increase in venous distension. Distended veins are easily compressed during shunt failure causing venous outflow obstruction and further increase in ICP [12,13].

Both of our patients presented only with visual symptoms which on evaluation were found to be SVS and underwent shunt revision which improved vision. However it was difficult to determine which pathophysiology fits in these cases. Management of slit ventricle syndrome involves complex interplay of ventricular volume, pressure, small ventricle and shunt revision for blocked shunt [14]. Over-drainage of cerebrospinal fluid (CSF), shunt malfunction, intracranial hypertension, decreased brain parenchyma

compliance to variations in cerebrospinal fluid volume have been proposed as mechanism of SVS syndrome.

Management of SVS requires a complete neurological examination, shunt chamber filling, sensory, motor coordination, gait and balance. The patient also requires examination of fundus for papilloedema. Computerised Tomography (CT) of head is first imaging modality, better in localising shunt location than MRI (Magnetic Resonance Imaging). However MRI brain is better CT in detailed examination of brain and has advantage of no radiation exposure. Based on history, neurological examination and imaging, management plan is decided accordingly. Many patients can be managed with observation and close follow up. Some may need admission with intravenous fluids, steroids, ICP monitoring and even surgery.

ICP monitoring is a valuable tool in differentiating high from low ICP. Intracranial hypotension is found in nearly half of these patients [15]. Low ICP may require hydration, medicines, antisphion devices or adjustment of programmable shunt, with abdominal binder for refractory low pressure [16]. High ICP may require shunt revision, addition of lumboperitoneal or cistern magna shunt type, shunt exteriorisation, antisphion devices or ETV (Endoscopic Third Ventriculoscopy) with shunt removal [17].

Diagnosis of SVS can be delayed if radiological finding and clinical features of raised ICP are absent, as in our cases visual symptoms are only clue to shunt malfunction. Visual pathways and especially abducent nerve are vulnerable to increased ICP because of their subarachnoid course [18]. Papilloedema may not be a very sensitive sign but helps in detecting shunt malfunction [19]. Raised ICP in SVS can lead to permanent visual loss and should not be ignored.

Conclusion

SVS has no unique specific clinical characteristics, natural course, incidence, age distribution, pathogenic mechanisms, so making it difficult to diagnose. Moreover, laboratory investigations, ICP monitoring and various tests used for evaluating shunt function appear to vary considerably with no unique therapeutic approach identified. In conclusion, SVS can also present with only visual symptoms in absence of other neurological symptoms. Occurance of visual symptoms in shunted patients should not be ignored, closely watched and evaluated for raised ICP.

Conflict of Interest

Nil.

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