UNUSUAL VISION IMPROVEMENT IN A CASE OF SLIT VENTRICLE SYNDROME AFTER ENDOSCOPIC THIRD VENTRICULOLOSTOMY

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ABSTRACT: Slit ventricle syndrome is a rare condition where brain compliance is reduced. It is associated with intermittent intracranial hypertension. It has been typically observed in older hydrocephalic children operated in early infancy, even though it has been observed occasionally in young children and adults. Endoscopic third ventriculostomy (ETV) is difficult in slit ventricle syndrome because of collapsed and fibrosed ventricles. In our case, patient had symptomatic improvement after endoscopic third ventriculostomy.

KEYWORDS: Slit ventricle syndrome, Endoscopic third ventriculostomy, ventriculo-peritoneal shunt.

INTRODUCTION: The term 'slit ventricle syndrome' (SVS) refers to the occurrence of headache, vomiting, possibly some degree of consciousness impairment and visual deterioration in children with hydrocephalus where previously Ventriculo-Peritoneal (VP) shunt was done. On computerized tomography (CT) scan collapsed, slit like ventricles are seen which is diagnostic.

It is a rare condition associated with decreased brain compliance and intermittent intracranial hypertension. It has been typically observed in older hydrocephalic children operated in early infancy, even though it has been observed occasionally in young children and adults.

Doing Endoscopic third ventriculostomy (ETV) is difficult because of the collapsed and slit like ventricles which are hard to tap.

In our case patient had relief of headache, decreased irritability and Improvement of vision after the procedure.

CASE REPORT: 12 Years male child had a H/o recurrent episodes of Severe headache in bifrontal region, Vomiting, Irritability and Diminution of vision in his left eye more than right eye since one month. He was operated 8 years back for a posterior fossa tumor (histopathology report not available) along with right sided ventriculo-peritoneal shunt.

CT scan brain was done showing slit like ventricles (fig. 1) with ventricular end insitu. Patient observed in ICU for two days. Ophthalmological examination shows Bilateral optic atrophy changes. Patient had persistence of symptoms even with medical management. Diagnostic guarded lumbar puncture was done, which relieves headache. In view of fibrosed and slit like collapsed ventricles, theco-peritoneal shunt was done and previous VP shunt was removed at the same time. Post operatively patient recovered gradually. Headache and irritability were subsided. He was discharged on 5th postoperative day.

Ten days after discharge he came with complaints of diminution of vision in both eyes, severe head ache and irritability. On ophthalmological examination there is decreased visual
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acuity with perception of light in both eyes and left sided lateral rectus palsy. MRI brain was done showing mild enlarged ventricles compared with previous brain scans (fig. 2 & 3). So he was taken up for endoscopic third ventriculostomy (ETV) as a final option. After ETV irritability subsided, headache relieved completely and visual acuity improved to hand movements bilaterally. Patient was followed for 6 months and his vision is improved that he is attending his classes.

DISCUSSION: The classic definition of small ventricles, a slowly filling shunt reservoir, and chronic intermittent headaches form the basis of the clinical diagnosis of slit ventricle syndrome.¹ Original descriptions of slit ventricle syndrome management predicted a more complex interplay of ventricular volume & pressure, descriptions of small ventricles (normal volume ventricles) and shunt revision for occluded catheters.²

There are five clinical presentations of patients with shunt headache differentiated by history, symptoms, and ICP monitoring results as described by Rekate and associates.³ ⁴ ⁵

1) Intermittent shunt malfunction: Episodic high pressure symptoms are the mainstay, sometimes associated with activity.

2) Elevated ICP with functioning shunt: This has been termed cephalocranial disproportion. An example is seen in shunted children with craniofacial syndromes. Headaches, vomiting, and papilledema can lead to visual loss elevated ICP is left untreated.

3) Elevated ICP with malfunctioning shunt (normal volume hydrocephalus): This has been termed normal volume hydrocephalus, or shunt pseudotumor. The patient may have morning headaches unrelieved by analgesics or visual changes similar to cephalocranial disproportion symptoms above.

4) Low ICP: This has been attributed to shunt overdrainage. Low ICP symptoms progress during the day with the headache improved with recumbency. Rarely, cranial nerve 6 palsies can occur.

5) Headache unrelated to shunt function: Not uncommonly there will be a family history of migraines, episodic headache, or headaches relieved by rest.

There are different types of mechanisms which explain pathophysiology of increased intracranial pressure in slit ventricle syndrome. a) At 5 or 6 years of age there is a maximal brain growth due to this the reduction of subarachnoid CSF to buffer increases ICP during times of catheter obstruction. An inability for the spinal subarachnoid space to aid in buffering high ICP (due to a smaller than normal spinal canal) may also play a role.⁶ b) In the presence of a shunt malfunction where the ependymal wall becomes coapted to the catheter openings and causes transient obstruction there is elevation of intracranial pressure. c) Reduced brain compliance leading to high ICP has been implicated. d) Periventricular gliosis may develop after chronic shunting and contribute to the inability of the ventricles to dilate. Gliosis has been found on autopsy with and without small ventricles.⁷ e) There is some evidence that decreased brain compliance does not occur, but rather that there is increased cerebrovascular distensibility (venous). Distensible veins compress easily during increased ventricular pressure situations (i.e., shunt failure), causing venous outflow obstruction (decreased venous return) and further increases in ICP.⁸ ⁹
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A thorough neurological evaluation including mental status, shunt appearance and continuity, funduscopic examination for papilledema, cranial nerve function, sensorimotor examination, gait, balance, and coordination is essential. The patient can have a pseudomeningocele around the shunt with malfunction and papilledema with raised ICP (shunt failure, normal volume hydrocephalus, pseudotumor cases).

CT scan of the brain is the first modality used to rule out shunt malfunction. CT scans are better than MRIs for visualizing the shunt catheter location in the brain. An MRI of the brain can offer a more detailed examination of brain and ventricular anatomy and spare the child radiation. Scans will show small or slit ventricles, a paucity of subarachnoid space in elevated ICP situations (tight brain). There are other signs on MRI T2-weighted sequences indicative of elevated ICP, i.e., fluid around the optic nerve, an empty sella, or protrusion of the optic disc into the globe.

Correlation of Tests, History, examinations, and imaging: A management plan is created on the basis of the overall clinical picture, as it correlates to the imaging, with emphasis on the symptoms and signs and their severity. Some children can then be managed with cautious outpatient observation with close followup. Many will need to be admitted for IV fluids, medications, and possibly ICP monitoring or surgery.

Few nonsurgical interventions like steroids and IV fluids can reduce symptoms in the child with overdrainage or low ICP. ICP monitoring is a good starting point in evaluating a child with small ventricles, headaches, and no evidence of a shunt malfunction, as it can differentiate high from low ICPs. In one series more than half of children monitored had intracranial hypotension. High ICP will require shunt revision surgery (often difficult with small ventricles), the addition of another shunt type (lumboperitoneal or cisterna magna), or shunt externalization with attempts to increase ventricular size, using antisiphone measures and perform an ETV with the goal of eventual shunt removal. Management of refractory elevated ICP with a decompressive craniectomy (subtemporal) has been successful in several small series. Low ICPs require hydration, medication, adjustment of a programmable shunt, or the addition of an antisiphon component to the distal shunt. An abdominal binder is an option for refractory low-pressure symptoms.

CONCLUSIONS: Endoscopic third ventriculostomy in slit ventricular syndrome is one of the best procedure in relieving symptoms even with collapsed ventricles when it was done promptly and can be tried as a first procedure.

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Fig. 1: CT showing collapsed slit like ventricles

Fig. 2: MRI showing dilated ventricular system after the coperitoneal shunt
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Fig. 3: MRI showing dilated ventricular system after thecoperitonea shunt 2