

## CASE REPORT

# Occam's razor in the management of ventriculoperitoneal shunt dysfunction: Diagnosis and management of an unusual pediatric case

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## ABSTRACT

Ventriculoperitoneal (VP) shunt surgery is probably the commonest surgical procedure in neurosurgery. Belying its technical simplicity is the myriad complications associated with it. Shunt malfunction is a common complication associated with this surgery, second only to shunt related infections, which may be associated with it. Sterile cerebrospinal fluid (CSF) eosinophilia (CE) has been reported with VP shunts, which may or may not be related to the dysfunction. Eosinophilia in the CSF has also been associated with a number of other conditions including parasitic infestations in the brain. This may be unrelated to the shunt surgery. We present a case of a child, operated earlier for hydrocephalus, who presented with sub-acute loss of vision and bilateral oculomotor paresis. CSF from a chamber tap revealed eosinophilia. The commonest presenting symptom of shunt malfunction is raised intracranial pressure. There are no reports in the literature of VP shunt malfunction presenting with bilateral oculomotor paresis and decreased visual acuity. The associated CE complicated the clinical picture, especially since the initial brain radiology was normal. We discuss the clinical differential diagnosis of this very interesting presentation, management dilemmas and outcome in this child. This rare clinical presentation was found to be the result of a shunt malfunction and not due to any rare parasitic infestation of the brain. Occam's razor dictates that the simplest explanation in a given situation is usually the most accurate, as is seen in this case.

**Key words:** Eosinophilia, eosinophilic meningoencephalitis, Occam's Razor, pediatric, shunt malfunction, ventriculoperitoneal shunt

## Introduction

Ventriculoperitoneal shunt malfunction (VPSM) is quite common and may occur in up to 50% of operated pediatric patients.<sup>[1]</sup> It usually presents with features of raised intracranial pressure.<sup>[1]</sup> Visual loss as a presenting feature is extremely rare as is the involvement of the oculomotor nerve.<sup>[1,2]</sup> Shunt malfunction may be misleading in unusual presentations because there may be no radiological signs

(in up to 10% of patients), symptoms or signs of raised increased intracranial pressure (ICP) and even the absence of papilledema.<sup>[1,3,4]</sup> Shunt malfunction may occur in the setting of infections, which may be responsible for cerebrospinal fluid (CSF) eosinophilia (CE).<sup>[5]</sup> We present a child with unusual clinical features and CE with normal radiology. The diagnosis was missed until the child deteriorated clinically, and repeated radiology revealed overt hydrocephalus.

## Case Report

A 6-year-old child, operated elsewhere soon after birth, for myelomeningocele repair and a ventriculoperitoneal (VP) shunt presented with a history of progressive decrease in bilateral visual acuity and bilateral ptosis. The child had traveled to his remote village about 1-month prior to the commencement of symptoms. He was of normal mentation and had discontinued schooling 2 weeks prior due to worsening vision. On examination the child was conscious, alert and oriented. He had bilateral ptosis and restricted ocular movement [Figure 1a]. His vision was limited to perception of light in both eyes. There was no papilledema on fundoscopy. He had recent history of fever 1-week prior to presentation and had no symptoms or

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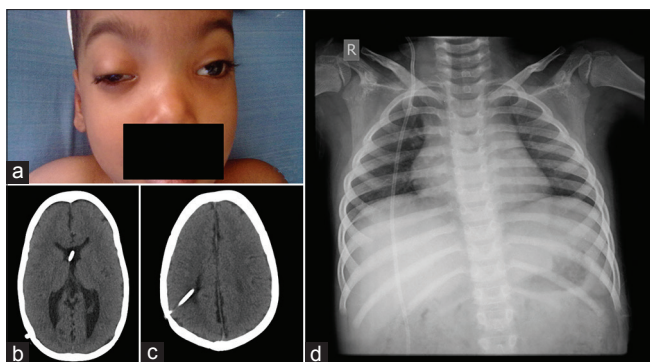
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signs of raised intracranial pressure. The computed tomography (CT) scan done at presentation revealed no evidence of hydrocephalus with the shunt tip *in situ* [Figure 1b and c]. Chest and abdomen were screened, but no evidence of shunt fracture or disconnection of the tube could be identified [Figure 1d]. In view of the preceding fever, a chamber tap for CSF analysis was done [Table 1]. The CSF revealed florid eosinophilia [Figure 2] and an elevated protein level but normal CSF sugar levels. The child had a mild fever during the first 2 days after admission. Blood tests did not reveal any evidence of infection with normal total counts and erythrocyte sedimentation rate. His CSF, blood and urine cultures were sterile. A repeat chamber tap (which proved to be a little difficult and only 1 ml could be aspirated) at this point revealed further elevation of the CSF count and protein but CSF sugars remained normal [Table 1]. A working diagnosis of eosinophilic meningoencephalitis (EME) as made and a battery of tests were undertaken to ascertain both infective and noninfective causes of CE [Table 2]. He was started empirically with Albendazole and steroids considering the possibility of parasitic infestation as a cause of CE. On closer evaluation, he was found to have a mild cough with expectoration, and the sputum culture grew *Staphylococcus aureus*. He was managed with appropriate antibiotics and the fever subsided. There was no change in the vision or bilateral 3<sup>rd</sup> C. N palsy. A repeat CT scan of the brain revealed a slight increase in the size of the ventricles but the sulci, gyri were well visualized and the child had normal mentation and no signs of raised ICP [Figure 3c]. A lumbar puncture was done (as the chamber had collapsed, and no further CSF could be aspirated) which revealed clear CSF with a cell count of only 2 cells with normal biochemistry [Table 1]. About 36 h after this the child started to become progressively drowsy, and a repeat CT scan of the brain revealed further dilatation of the ventricles with effacement of the sulci [Figure 3d]. He had no papilledema or features of raised ICP at this point. He underwent a left sided VP shunt. Intra-operatively, the CSF was clear but under high pressure. An attempt was made to remove the previous shunt. At the cranial incision, the

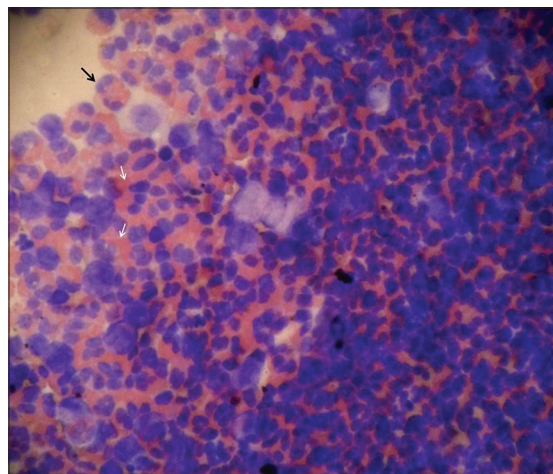
chamber, in continuity with the lower end was found separated from the upper end of the shunt tube, and the burr hole was found completely fused. The upper end could not be accessed. On retrospective analysis and on adjusting the window width appropriately, bony fusion obliterating the burr hole was identified on previous CT scans [Figure 3f]. Postoperatively the child rapidly improved in sensorium and had a finger counting at 3 feet at the time of discharge. His 3<sup>rd</sup> cranial nerve palsy did not show much improvement at discharge. Post op CT scan revealed a decrease in the size of the ventricles [Figure 3e].

## Discussion

The typical presentation of a VP shunt malfunction is heralded by symptoms and signs of raised intracranial pressure and enlarged ventricles on radiological evaluation.<sup>[2,6]</sup> The “classical” symptoms attributed to VP shunt include headaches, vomiting and drowsiness.<sup>[6]</sup> This drowsiness has a high degree of correlation with acute VP shunt blockage.<sup>[6]</sup> Decreased visual acuity has rarely been documented in the literature but is considered an atypical presentation along with other presentations such as seizures, abdominal pseudocyst, syringomyelia, cranial nerve palsies, and hemiparesis.<sup>[3,4,6,7]</sup> Although rare, decreased visual acuity could be a portent of permanent visual impairment such as has been reported in 1.8% of all children experiencing an episode of raised intracranial pressure due to shunt malfunction.<sup>[7]</sup> It has also been reported that in such cases the patient need not have features of raised intracranial pressure or papilledema.<sup>[2,3,4,7]</sup> Cerebral infarcts in the posterior cerebral artery territory have been documented.<sup>[3]</sup> There are reports of patients presenting with isolated oculomotor paresis as the only clinical sign of hydrocephalus.<sup>[2]</sup> Presentation with both decreased visual acuity and bilateral oculomotor paresis in the same child due to shunt malfunction, a combination of two atypical presenting symptoms, as seen in this case, has not been reported in the literature.



**Figure 1:** (a) The clinical photograph of the child demonstrating bilateral ptosis. (b and c) The computed tomography scan images of the child at presentation demonstrating normal-sized ventricles and no radiological evidence of shunt malfunction. (d) The chest and abdomen X-ray demonstrating no obvious fracture in the shunt tube



**Figure 2:** Photomicrograph cytospin preparation of cerebrospinal fluid showing numerous eosinophils with binucleation (black arrow) and orange-red refractile cytoplasmic granules (white arrows) (Leishmann stain, x400)

Shunt malfunction has been reported at around 39% during the first postoperative year and around 53% during the 2<sup>nd</sup> year.<sup>[1,2]</sup> It has also been reported that if the shunt malfunction occurred within 2 years of surgery, it was most likely due to a ventricular end malfunction and beyond 2 years is usually due to a distal end malfunction.<sup>[2]</sup>

The assessment of shunt malfunction begins with a detailed neurological history and assessment and the “classical” symptoms of shunt blockage have been positively correlated in over 70% of cases.<sup>[6]</sup> In this case the initial symptoms were atypical.

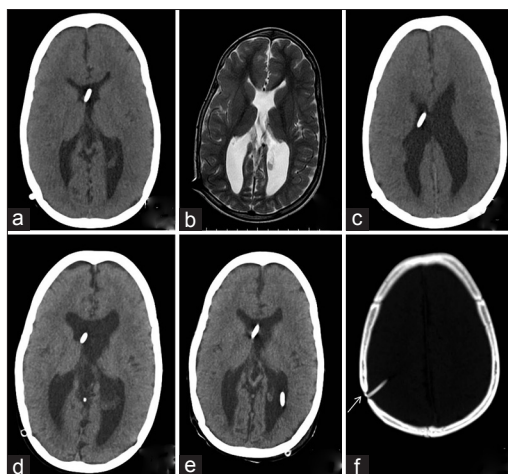
The radiology may be deceptive, and there have been reports with brain scans revealing normal-sized ventricles.<sup>[1,3,4,7]</sup> In one study, there was 84% correlation between the diagnosis of shunt malfunction and change in the size of the ventricles on CT scan, but in comparison with a baseline CT scan, prior to the malfunction.<sup>[6]</sup> In the present case, there was no change in the size of the ventricles as compared to previous scans on the initial CT scan. Shunt series radiographs have also been recommended as one of the early investigations to diagnose

shunt malfunction, which were done in this case and found to be normal.<sup>[4]</sup> The initial episodes of raised ICP may be mild and intermittent and therefore may not be adequate in enlarging ventricles sufficiently to be appreciated on imaging.<sup>[7]</sup> Alternately, it has been hypothesized that a VP shunt done early in life may be responsible for early sutural closure and sub-ependymal gliosis which makes the ventricular system resistant to subsequent dilatation when a shunt malfunction occurs, thus being responsible for normal appearing ventricles on a CT scan.<sup>[7]</sup>

Prolonged ICP monitoring has also been recommended to aid diagnosis in difficult or atypical shunt malfunction cases.<sup>[2]</sup> In retrospect, ICP monitoring would have probably clinched the diagnosis early in this child.

### Cerebrospinal fluid eosinophilia

Cerebrospinal fluid eosinophilia has been described in VP Shunt patients in whom it may be asymptomatic, or it may be responsible for VP shunt malfunction.<sup>[5]</sup> CE may also be seen in EME in a long list of pathogens [delineated in Table 3] and noninfective conditions.<sup>[8]</sup> EME has been reported in humans infected by rat lungworm, *Angiostrongylus cantonensis* (endemic in South East Asia and the Caribbean) has been reported from India.<sup>[8]</sup> Its life cycle involves snails, slugs or fish as intermediate hosts and rodents as definitive hosts.<sup>[8]</sup> Humans are accidental hosts where the worm migrates, but does not reach maturity.<sup>[8]</sup> This rare infection may occur by ingesting poorly cooked, or raw fish, slugs, snails or vegetables contaminated by an infected rat.<sup>[8]</sup> The central nervous system insult is caused by direct mechanical and toxic injury by the worm.<sup>[8]</sup> The immunologic reactions of the host also play a role.<sup>[8]</sup> Parasites such as *A. cantonensis* would be one of the primary suspect amongst parasites considering the travel history, preceding fever and ocular symptoms in this present case. Initial CT scans may be normal.<sup>[8]</sup> MRI of the brain may reveal dilated CSF spaces with peri-ventricular lucencies.<sup>[8]</sup> Diagnosis is confirmed by a Western Blot analysis.<sup>[8]</sup> Additional tests such as serology for *A. cantonensis* were being planned prior to the clinical deterioration of the child but were difficult due to the nonavailability of the reagents locally. The condition is generally self-limited with excellent prognosis.<sup>[8]</sup> A 2 weeks course of albendazole and prednisolone has been



**Figure 3:** Chronological depiction of the radiological changes observed. (a) Reveals the computed tomography (CT) brain at the time of presentation and the (b) reveals the normal appearing magnetic resonance imaging done soon after. (c) Done 5 days after the initial scan revealed slight increase in the size of the ventricles but no periventricular lucencies. (d) Demonstrates the further increase in the size of the ventricles on CT scan after the child deteriorated in sensorium. (e) Is the postoperative CT scan of the brain demonstrating a reduction in the ventricular size. (f) Demonstrates the bony fusion (arrow) obliterating the burr hole which was missed on previous CT scans.

**Table 1: CSF analysis**

Date	Sample	Cell count (cells/cu mm)	Differential cell count (%)			CSF glucose (mg/dl)	Corresponding RBS	CSF protein (mg/dl)	Culture sensitivity
			Eosinophils	Lymphocytes	Monocytes				
At presentation	Chamber tap	2500	95	5	0	94.6	90	2770	Sterile
3 days later	Chamber tap	7000	93	2	5	108	79	2830	Sterile
6 days later	Guarded LP	3	0	3	0	72.8	110	20.9	Sterile
7 days later	Intra-operative CSF	2	0	2	0	3.2	150	102.8	Sterile

LP – Lumbar puncture; RBS – Random blood sugar; CSF – Cerebrospinal fluid



**Table 2: Other investigations to ascertain the etiology of EME**

Investigation	Result
Anticysticercal antibodies IgG/IgM ELISA	Negative
CSF AFB	Negative
CRP	0.27 mg/L (0-3)
ESR	10 cm/h
AEC	100/cumm (20-500)
ANA	Negative
India ink for cryptococci	Negative
PS	Normocytic/normochromic
Sputum culture	<i>Staphylococcus aureus</i>

EME – Eosinophilic meningoencephalitis; CSF – Cerebrospinal fluid; AFB – Acid-fast bacilli; CRP – C-reactive protein; ESR – Erythrocyte sedimentation rate; AEC – Absolute eosinophil count; ANA – Antinuclear antibody; PS – Peripheral Smear

**Table 3: Infective and noninfective conditions associated with CSF eosinophilia<sup>1</sup>**

Bacterial	Parasitic	Fungal	Other
<i>Coagulase negative Staphylococcus</i>	<i>Angiostrongylus cantonensis</i>	<i>Candida</i> sp.	Glioblastoma
<i>Staphylococcus aureus</i>	<i>Gnathostoma spinigerum</i>	<i>Cryptococcus</i> sp.	Lymphomas
<i>Propionibacterium acnes</i>	<i>Paragonimus</i> sp.		Acute leukemias
<i>Streptococci</i> sp.	<i>Strongyloides stercoralis</i>		Carcinomatous meningitis
<i>Escherichia coli</i>	<i>Toxocara canis</i>	Viruses	Rheumatoid arthritis
<i>Pseudomonas aeruginosa</i>	<i>Loa Loa</i>	HIV	
<i>Klebsiella</i> sp.	<i>Toxoplasma gondii</i>		Drugs
<i>Enterobacter</i> sp.	<i>Taenia solium</i>		Radiographic contrast
<i>Corynebacterium</i> sp.	<i>Coccidioides immitis</i>		Ibuprofen
<i>Serratia marcescens</i>	<i>Schistosoma japonicum</i>		Ciprofloxacin
Tuberculous meningitis	<i>Fasciola hepatica</i>		
Syphilitic meningitis	<i>Trichinella spiralis</i>		Foreign bodies

CSF – Cerebrospinal fluid

recommended.<sup>[8]</sup> The other less common differentials should be considered in the diagnosis of CE outlined in Table 3. *A. cantonensis* infection is known to cause hydrocephalus requiring a CSF diversion procedure.<sup>[9]</sup> CE is known to be associated with shunt malfunction.<sup>[5]</sup> Although there are no documented cases of *A. cantonensis* causing a VP shunt malfunction, from the above discussion, it can be postulated that in such an occurrence revision of the shunt under appropriate steroid and antihelminthic cover would seem most appropriate.

### Occam's razor of parsimony or the principal of frugality

According to the principle of Occam's razor, the simplest explanation is usually the correct one. In this case, although

the child had visual deterioration, the oculomotor paresis confused the clinical diagnosis and he was initially diagnosed as EME, rather than shunt malfunction, in the absence of overt radiological shunt malfunction and was managed empirically with antihelminthic agents, antiallergic medications and steroids. In retrospective analysis, the respiratory tract infection was responsible for the fever. The eosinophilia was probably reactionary to the residual CSF in the chamber and the primary diagnosis was a mechanical shunt malfunction. The child improved rapidly after the insertion of a new shunt. Had the Occam's razor been applied to this case by perhaps ICP monitoring (as has been advocated),<sup>[2,7]</sup> there would have been a rapid diagnosis without wasting resources on inconclusive and sometimes expensive tests. The vision loss may also have proven to be permanent had the delay in intervention been prolonged.

### Conclusion

Both visual loss and oculomotor paresis are extremely rare presentations for VP shunt malfunction. Extreme clinical vigilance is required in unusual presentations of common disorders. Although EME was a distinct possibility, the simplest explanation is usually the one that's correct. Diagnosis and management should be aimed at early resolution of the offending complication to ensure a good outcome.

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