CASE REPORT

Dandy-Walker Malformation with Refractory Epilepsy: Role of Radionuclide Scintigraphy in Assessing Ventriculo-Peritoneal Shunt Patency

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ABSTRACT

Dandy-Walker malformation (DWM) is an uncommon congenital anomaly characterised by enlarged posterior fossa involving the cerebellum and fourth ventricle. Most of DWM patients with hydrocephalus require ventriculo-peritoneal shunt (VPS) especially in those with symptoms such as headache and seizures. In cases with worsening hydrocephalus although on VPS, computed tomography and magnetic resonance imaging can be performed to assess ventricular sizes, and an obstructed shunt may be confirmed with iodinated contrast media injection into shunt reservoir under fluoroscopy guidance. VPS scintigraphy is one of infrequently performed nuclear imaging studies that has the capability to assess shunt patency and detect site of blockage with minimal radiation exposure. We describe here a 32-year-old lady with underlying DWM and hydrocephalus on VPS complicated by refractory epilepsy. Radionuclide scintigraphy has proven to be a safe, simple and useful tool in evaluating shunt function and localising the site of VPS obstruction in this patient.

Keywords: Dandy-Walker malformation, Refractory epilepsy, Ventriculo-peritoneal shunt, Shunt patency, Radionuclide scintigraphy

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INTRODUCTION

Dandy-Walker malformation (DWM) is characterised by the triad of enlargement of the posterior fossa, complete or partial agenesis of the cerebellar vermis and cystic dilatation of the fourth ventricle. It is frequently found in association with hydrocephalus (1). The mainstay of treatment to this disease with hydrocephalus-related symptoms is surgical cerebrospinal fluid (CSF) shunt i.e. ventriculo-peritoneal shunt (VPS). Symptoms which get worse after shunt placement may indicate malfunction or obstruction of the VPS. There are few imaging modalities that are able to evaluate shunt patency such as radiography, ultrasonography and computed tomography. Radionuclide shunt scintigraphy is another modality which is simple and safe to be performed, however it is infrequently done.

CASE REPORT

A 34-year-old Chinese lady was admitted to Hospital Pulau Pinang in late 2016 for frequent episodes of breakthrough seizure although with treatment, and was monitored in the ward for more than a week. At the age of three months, she was diagnosed with epilepsy and DWM complicated by obstructive hydrocephalus. A VPS was inserted since then to improve her symptoms. However, she had multiple episodes of shunt blockage confirmed by computed tomography (CT), requiring few revisions. The first revision was done at the age of nine years and the second revision was performed at the age of 14 years after she presented with severe progressive headache. At the age of 31 years, a new left VPS was inserted, again due to VPS malfunction.

During the admission, she had four episodes of generalised tonic-clonic seizure lasting for few minutes associated with loss of consciousness. However, there was spontaneous recovery of consciousness following each seizure episode. She had an episode of status epilepticus and a CT of the brain was subsequently performed to assess her anomalies, demonstrating elevated tentorium with enlarged posterior fossa and posterior fossa cystic lesion (Fig. 1). Proximal end of bilateral VPS was noted in-situ. Nonetheless, in comparison with her previous CT, there was no worsening of hydrocephalus seen.

As a result of her refractory epilepsy, she was later referred to Nuclear Medicine Department for VPS scintigraphy to assess her shunt patency. Approximately 0.6ml of Tc99m-diethylene triamine pentaacetic acid

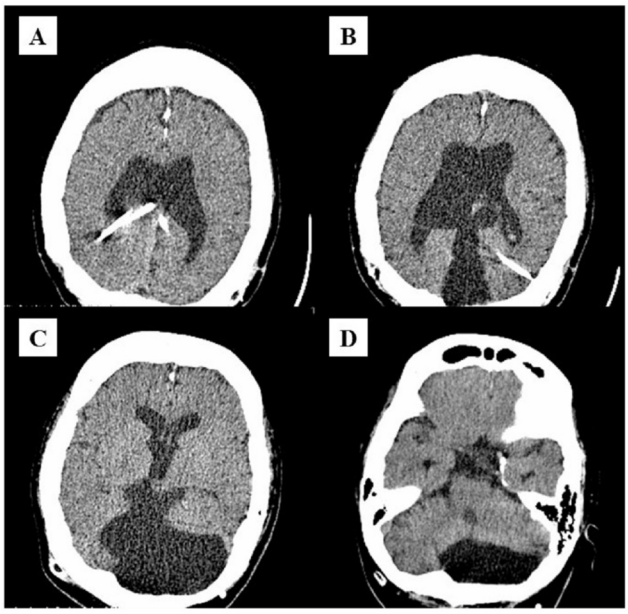

Figure 1 A-D: Axial CT brain, showing enlarged posterior fossa with presence of cystic lesion and hydrocephalus in keeping with Dandy-Walker malformation. Bilateral shunt tubes (proximal end) noted in-situ on image A and B.

(DTPA) (187 MBq) was injected at the reservoir of the left VPS under aseptic technique (Fig. 2, 3 and 4). Dynamic anterior and lateral view scintigrams were obtained for the first 15 minutes, followed by sequential and delayed imaging at 1 hour, 2 hours and 24 hours post tracer injection. Delayed scans revealed significant tracer hold up in the distal end of the distal limb with no obvious dispersion within peritoneal cavity, suggestive of possible obstruction at the distal end of the limb of the left VPS. In view of these findings, a revision surgery was planned. However since the last seizure attack in the ward, she had no signs and symptoms of increased intracranial pressure and her symptoms improved, hence the surgery was withhold by the neurosurgical team. Her shunt reservoir demonstrated good emptying and filling throughout her recovery in the ward. She was later discharged well with further follow-up as outpatient.

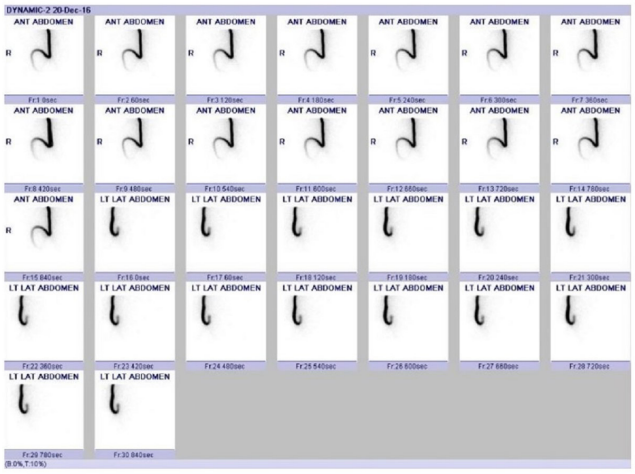

Figure 2: Dynamic VPS scintigraphy at first 15 minutes post Tc99m-DTPA injection showing good flow within the reservoir to the distal limb.

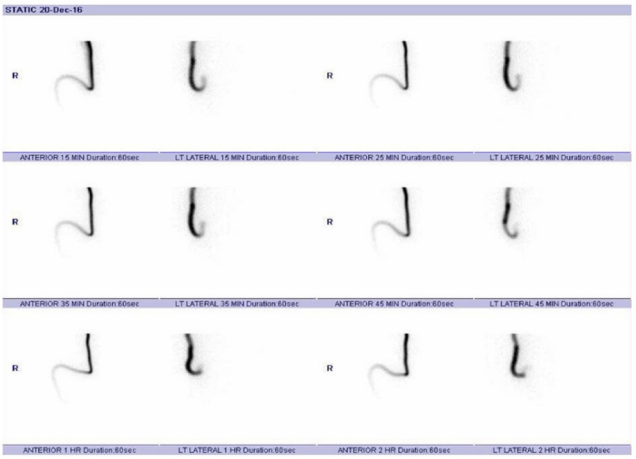

Figure 3: Sequential VPS scintigraphy at 15 minutes, 25 minutes, 35 minutes and 45 minutes post Tc99m- DTPA injection showing good flow within the reservoir to the distal limb. However, tracer hold up seen at 1st and 2nd hour post tracer injection

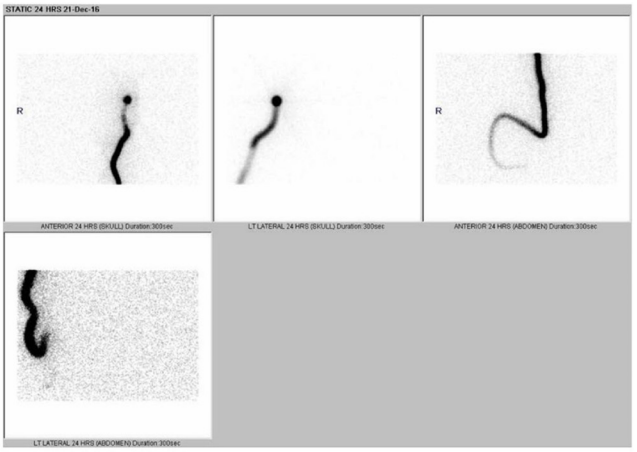

Figure 4: Persistent tracer hold up in the distal end of the distal limb with no obvious dispersion of the tracer within peritoneal cavity seen at delayed 24 hour imaging

DISCUSSION

Overproduction of CSF, obstruction of its flow or failure of reabsorption may subsequently cause excessive buildup in the ventricles of the brain. It may lead to increased intracranial pressure and many related symptoms such as headache, vomiting and seizures. Shunts are usually permanent treatment option to release the high intracranial pressure. There is an increased number of VPS insertion worldwide which subsequently increased the survival of children with hydrocephalus (2). The shunt system consists of a catheter with proximal end which is positioned within the lateral ventricle of the brain, a subcutaneous reservoir with a valve implanted at the mastoid bone region and the distal end which drains into body cavity that is capable to reabsorb fluid such as the peritoneum and pleural cavity. A pliable material covers the outer side of the reservoir to allow the pump to be manually depressed and also for needle access. Most neurosurgeons prefer VPS because of its easy access with less complication as compared to other shunts (2).

Shunt malfunction may occur at any time after insertion and any points along the shunt system. About 25-40% VPS malfunctioned in the first year of placement, and 63-70% within 10 years (2). Of all, ventriculo-pleural and ventriculo-atrial shunts were found to have higher chances to be malfunctioned. Undeniably, the patency and obstruction site of a shunt are essential for treatment planning (1). The diagnosis of shunt malfunction is challenging. Shunt malfunction may be due to mechanical issue or obstruction. Partial obstruction due to infection and inflammatory debris usually occur within six months of insertion. Plugging of the proximal catheter by brain parenchyma, kinking or disconnection of the tube and occlusion of the tubing tip by omentum or emboli may cause shunt malfunction. Most obstructions occur in the distal limb. The initial simple assessment can be clinically performed by examining the outer surface of the shunt's reservoir. The distal end of the catheter is likely patent if it readily depresses. If it readily refills, the proximal end of the catheter is likely to be patent. Opening pressure can also be measured by assessing the reservoir and CSF may be obtained for laboratory analysis and culture. In this patient, there is a possibility of partial or temporary obstruction at the distal end of the limb of the left VPS as her symptoms improved after her last seizure attack in the ward. There were no signs and symptoms of increased intracranial pressure with infrequent attack of seizure since then, suggesting that the obstruction may be due to kinking of the tube or the distal tip was temporarily occluded by debris or surrounding omentum. Her shunt reservoir demonstrated good emptying and filling throughout her recovery in the ward.

Radiological tool such as radiography, ultrasonography and CT can be used for initial assessment, however they have low specificity for diagnosis of shunt malfunction (2). Assessment of the tubing condition can be done by radiography examination to look for kinking or disconnection. CT is performed to assess ventricular sizes and for comparison with previous scans (3). Numerous other procedures have been proposed such as injecting radionuclide and contrast media directly into the ventricle of the brain via the shunt system, ultrasound flowmeter and thermosensitive procedure (3). However these procedures are invasive. The radionuclide CSF shuntography has proven to be a safe, effective, non-invasive and low radiation burden technique. It can be performed by injecting a small amount of radionuclide into the shunt's reservoir and acquiring images using a gamma camera (3). It usually augments clinical diagnosis and management as it may prevent unnecessary surgical intervention in certain conditions. To date, there is no reported morbidity or mortality associated with this nuclear imaging study.

Jawa et al. demonstrated the usefulness of shuntography in the evaluation of VPS in 56 children with hydrocephalus. About 52% of the study population had partial blockage of their shunt tube, 32% had normal functioning tube, and 13% had mechanical defects causing total blockage (2). Those with partial tube blockage require flushing of the tube and antibiotics treatment, while those with mechanically blocked tube require shunt revision. In previous study by Graham et al., 192 CSF shunts were assessed including 140 VPS, 28 ventriculo-atrial shunts, 15 lumbar puncture shunts and seven ventriculo-pleural shunts (4). This study found that the sensitivity, specificity and accuracy of radionuclide shuntography was 97%, 90% and 93%, respectively. Oullette et al. demonstrated the additional value of shuntography to CT in 69 paediatric patients who were suspected as having shunt obstruction. False negative rate for CT was 8.7% while radionuclide shuntography was 2.9% (5). Combined modalities of CT and shuntography has increased the sensitivity and specificity in diagnosing shunt obstruction in comparison to single modality interpretation.

CONCLUSION

This case emphasises the role of radionuclide scintigraphy in CSF shunt functional assessment although infrequently requested and performed due to limited access to nuclear imaging facilities.

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