Normal Pressure Hydrocephalus: Scintigraphic Findings on SPECT/CT Image

Tzyy-Ling Chuang[1], Ming-Chin Hsu[2], Yuh-Feng Wang[1,3]

[1]Department of Nuclear Medicine, Buddhist Dalin Tzu Chi General Hospital, Chia Yi, Taiwan
[2]Department of Neurology, Buddhist Dalin Tzu Chi General Hospital, Chia Yi, Taiwan
[3]School of Medicine, Tzu Chi University, Hualien, Taiwan

A 65-year-old woman sustained a head injury, subarachnoid hemorrhage (SAH), and hydrocephalus. A ventriculoperitoneal shunt was subsequently implanted. However, the patient developed general weakness and mutism. Clinical evaluation revealed that the shunt was patent. Cisternography was performed by direct intrathecal administration of 6 mCi of technetium-99m diethylene triamine pentaacetic acid (^{99m}Tc-DTPA) by means of a lumbar puncture. The planar images showed the classic heart-shaped pattern of normal pressure hydrocephalus (NPH). Cisternography using SPECT/CT was also performed and revealed that the butterfly shape of increased radioactivity was clearly and definitely located in the bilateral lateral ventricles with the help of CT images. Compared with traditional planar images, the SPECT/CT makes the diagnosis of NPH more accurate and easier.

Key words: SPECT/CT, normal pressure hydrocephalus, cisternography

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For correspondence and reprints contact: Yuh-Feng Wang, M.D., Department of Nuclear Medicine, Buddhist Dalin Tzu Chi General Hospital. 2 Minsheng Road, Dalin Township, Chiayi 622, Taiwan. Tel: (886)5-2648000 ext. 5700, Fax: (886)5-2648508, E-mail: nment@ms7.hinet.net

Introduction

Differentiating a normal pressure hydrocephalus (NPH) from other forms of dementia and cerebral atrophy remains a challenge. Cisternography is a useful tool in the evaluation of cerebrospinal fluid (CSF) and is also helpful in selecting patients with presumed NPH for shunting. Treatment with shunting procedures can improve the clinical symptoms in patients with NPH. A typical finding of NPH in a cisternography is a heart shape of radioactivity seen in an anterior view of skull and presents as a butterfly shape in SPECT/CT image. The butterfly sign results from reflux of radiotracer into bilateral lateral ventricles post intrathecal injection of the radiopharmaceutical. When a typical heart-shaped activity presents, the diagnosis of NPH is very possible. However, occasionally it is difficult to differentiate ventricular activity from the normal filling of the ambient and quadrigeminal cisterns which may overlap with the lateral ventricle activity on the planar images. Even with the help of a SPECT image, the differentiation sometimes may be still not easy due to the lack of landmark.

With the help of advanced development of scintigraphic camera, SPECT/CT becomes more and more popular worldwide. With the aid of CT images, the co-registered SPECT/CT images with butterfly sign may be able to reduce the difficulty in interpretating a cisternogram. In this study, we evaluated the CSF condition in a patient with NPH using a SPECT/CT scanner and compared the findings between the traditional planar images and the SPECT/CT images.

Case Report

A 65-year-old woman had a past medical history of

hypertension and diabetes mellitus under medication control for one year. She suffered from a traffic accident with traumatic subarachnoid hemorrhage (SAH) and left femoral fracture in January 2007. Generalized weakness, abulia and akinetic mutism were noted and a head CT scan showed dilated ventricles with transependymal resorption. Her score for Glasgow Coma Scale was 7. Her pupil size was 2.5 mm with positive light reflex and her blood pressure was 143/89 mmHg measured at our emergency room. A few months after the accident, she was admitted for ventrico-peritoneal shunt (VP shunt) surgery under the presumptive diagnosis of NPH in March 2007. However, her symptoms did not improve and the head CT scan showed persistent dilated ventricles. Under the diagnosis of VP shunt dysfunction, she underwent VP shunt revision on June 25, 2007. During the surgery, slow distal CSF flow was noted and was improved by clearing the blocked distal shunt catheter. The patient improved after the surgery with some verbal output for only one day and then returned to baseline abulia and akinetic mutism. A neurologist was consulted and a CSF flow imaging was suggested. A typical pattern (heart-shaped) of NPH was shown on the planar imaging (Figure 1), in which the butterfly-shaped increase of radioactivity was clearly and definitely located in the bilateral ventricles with the aid of CT images (Figure 2). The patient underwent another shunt revision on July 6, 2007, to replace previous VP shunt with a medium pressure one, and an associated endoscopic third ventriculostomy was done. The patient tolerated well with the procedure, but the symptoms, specifically abulia with akinetic mutism state and four limbs weakness remained unchanged till discharge from the hospital.

Discussion

The clinical triad of NPH is ataxia, dementia, and urinary incontinence. “Dementia” is one kind of mental disturbance, characterized by apathy, irritability, and severe disorientation. “Ataxia” is defined as difficulty in movement, particularly in the initiation of motion. Patients with severe NPH may develop the picture of akinetic mutism as did our patient. Spinal-fluid examinations such as CSF pressure, CSF cell morphology studies and chemical laboratory data are unremarkable in the diagnosis of NPH [1], because CSF pressure may be abnormally low or high instead of the expected normal pressure and the cell and chemical studies often reveal no associated abnormalities.

CSF is formed primarily in the choroid plexus of the ventricles and enters the subarachnoid space at the base of the brain. It eventually ascends over the cortex and is absorbed through the pacchionian granulations of the pia arachnoid villa into the superior sagittal sinus. NPH was first described as an obstruction in the cortical subarachnoid space causing impairment of CSF reabsorption so that CSF refluxes into the lateral ventricles [1]. The pathogenesis is usually a precipitating event such as hemorrhage, infection or tumor which blocks the CSF pathways. Continued CSF formation increases CSF pressure, however, the vascular volume decreases with equilibration of intracranial pressure [2].

Pneumoencephalography (PEG) was an invasive X-ray procedure first used in 1919 to diagnose intracranial lesions with withdrawal of small amounts of CSF and replacement with air. It was replaced later with cerebral angiography and radionuclide cisternography. For most neurological diseases, head CT has replaced PEG and cerebral angiography, especially in the diagnosis of hydrocephalus, in which CT shows symmetrical dilated ventricles. Meanwhile, radionuclide cisternography is still valuable in preoperative assessment of patients with dementia and clinically suspected NPH because it reveals the physiology of dynamic CSF circulation [3,4].

In the past, ^{131}I-tagged human serum albumin (HAS) was used for cisternography [1]. However, ^{131}I has the disadvantages of beta particle emission, radiation exposure, and poor resolution even with high energy collimators. The only advantage of ^{131}I was the longer half life which allows delayed imaging. Currently, cisternography is performed by intrathecal injection of approximately 222 MBq (6 mCi) of ^{99m}Tc-DTPA into the lumbar subarachnoid space by means of lumbar puncture. ^{99m}Tc-DTPA is used because it is a gamma-emitting tracer (no particle emission), not lipophilic, not metabolized, and not absorbed across the ependyma before it reaches the arachnoid villi [5]. As compared with ^{131}I-labeled albumin, ^{99m}Tc-DTPA has the favorable characteristics of ready availability, greater administered activity with reduced radiation dose to patient, and advantageous photon energy (140 keV), producing higher-resolution images.

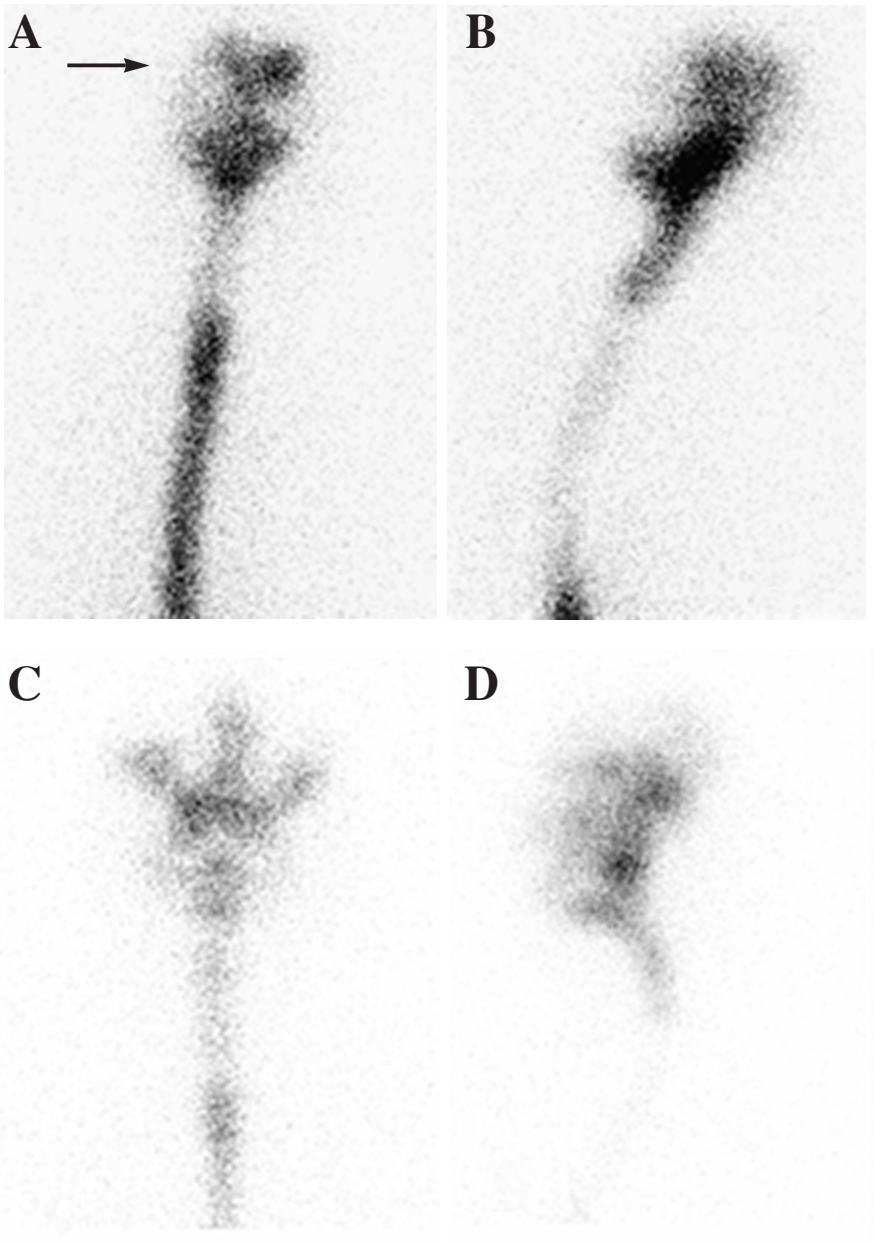

Figure 1. The cisternography was performed in a 65-year-old woman with NPH by directly administering 222 MBq (6 mCi) of ^{99m}Tc-DTPA via a lumbar puncture. (A) Planar anterior view and (B) lateral view of the head show a heart-shaped structure (arrow), as well as activity in the lateral ventricles at 120 min after injection. The cisternograms in a healthy patient are shown for comparison: (C) Planar anterior view shows the typical Neptune's triumvirate (trident pattern). Nevertheless, on lateral views (D), the normal cisternographic picture is hard to differentiate from the findings of NPH.

The classic cisternographic pattern of NPH on planar view consists of early entry of the radiotracer into the lateral ventricles and a central, heart-shaped structure instead of the usual trident pattern. In addition, activity in the lateral ventricles persists, and the radiotracer does not ascend over the superior aspect of the convexities on delayed images [6]. The cisternographic hallmark of NPH is the pattern of retrograde CSF flow into the lateral ventricles. Occasionally it is difficult to differentiate ventricular activity from the normal filling of the ambient and quadrigeminal cisterns which may overlap on the planar images [4].

Co-registration of SPECT/CT images during radionuclide cisternography in NPH reveals bilateral lateral ventricles in the form of the "butterfly sign". The butterfly symbol

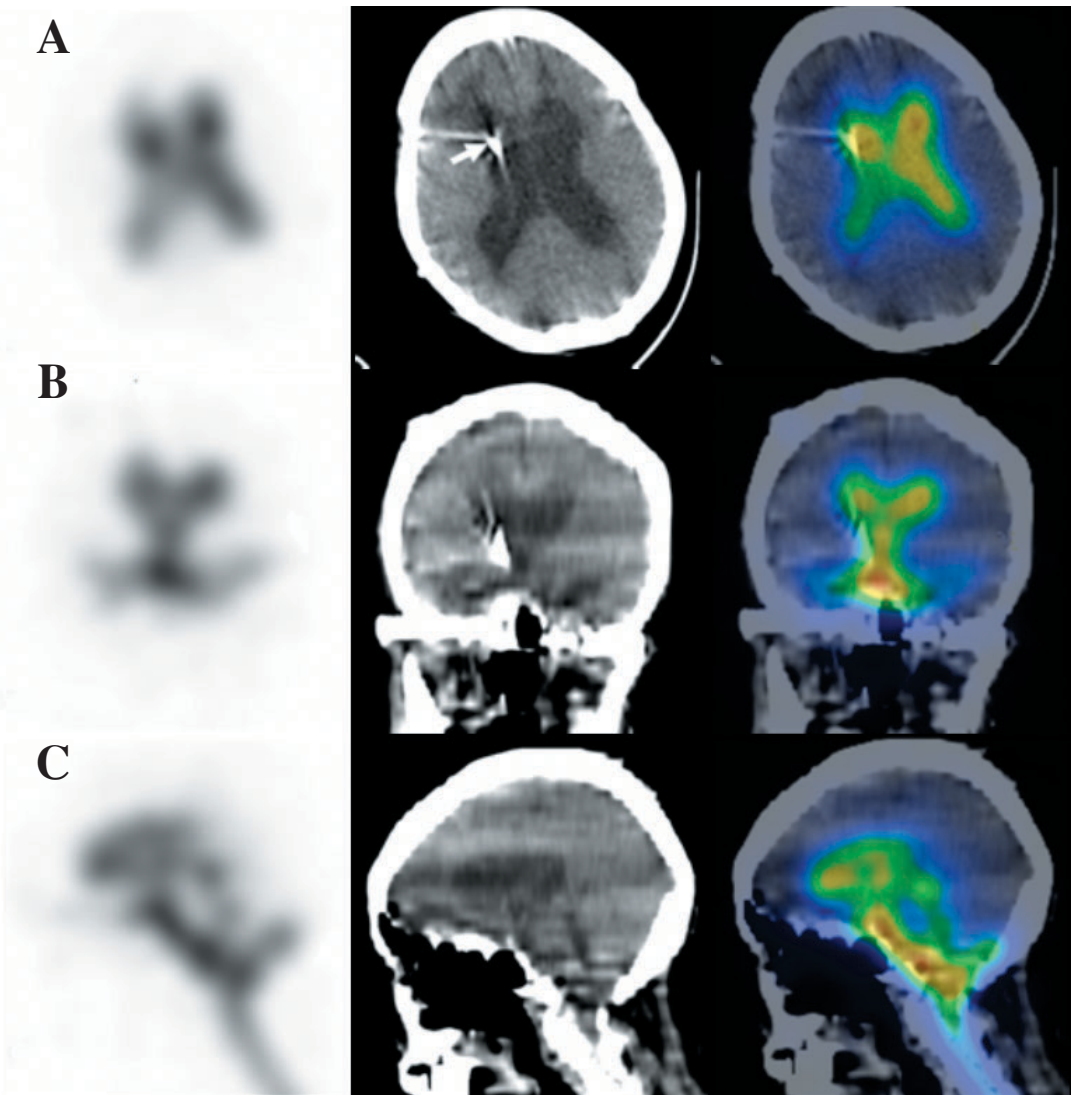

Figure 2. The cisternograms in a 65-year-old woman with NPH using a SPECT/CT scan. (A) Transverse SPECT/CT cisternogram obtained 3 h after injection shows radioactivity in the lateral ventricles, which has a butterfly shape. Arrow indicates a ventriculoperitoneal shunt. (B) Coronal view demonstrates the heart-shaped structure. (C) Sagittal views display CSF influx into the third ventricle. CT localization makes the identification of radioactivity in the lateral ventricles (ventricular reflux) easier and more accurate.

is similar to that seen in traditional CT cisternography and previous studies with transverse SPECT sections of NPH patients. Although transverse radionuclide cisternographic sections have been reported to clearly delineate the anatomic relation to the basal cisterns, the subarachnoid spaces, and the ventricles in both normal and abnormal patients [3], SPECT/CT can still assist in localizing anatomic details. In addition, SPECT/CT can demonstrate other anatomic abnormalities, such as cerebral infarction, brain hemorrhage, porencephalic cyst, tumor, and brain atrophy [7]. The three dimensional SPECT/CT images allow more exact localization of ventricular activity and may prove helpful in differentiating abnormal ventricular filling from normal activity in the surrounding cisterns.

Our patient had a history of SAH, which likely caused her NPH. Her symptoms were atypical, but radionuclide cis-

ternography revealed typical findings of NPH. The correct diagnosis of NPH is important, because this is a rare type dementia that can be treated successfully by shunting [1] if diagnosed early. Interestingly, our patient received a ventriculoperitoneal shunt, which often improves NPH [8]. The possible reasons for failure in improvement of the patient's symptoms include malfunction of shunt, misdiagnosis, coexisting vascular disease or degenerative atrophy, or irreversible cerebral damage from longstanding hydrocephalus [2]. In our case, implantation with revision of the VP shunt three times still could only temporarily and mildly improve the patient's symptoms. Irreversible cerebral damage is, therefore, likely in our case.

Some articles suggested that there was a poor correlation between a "positive" cisternogram and the results likely to be achieved by shunting. Patient selection for CSF diversion in NPH is difficult and remains a diagnostic and therapeutic challenge: many patients do not display the classic clinical and neuroimaging patterns, NPH mimics Binswanger disease or Alzheimer's disease and many patients with suspected NPH may suffer from both disorders and shunt procedures do not always result in clinical improvement. After 1993, positive radionuclide cisternography alone was not considered sufficient for shunting; a positive repeat lumbar tap or lumbar external CSF drainage is usually required [8,9]. Nuclear or CT cisternography which show that CSF dynamics is 'deficient or malfunctioning' may have a role in diagnosing communicating hydrocephalus, but it cannot predict shunt response solely. Another study demonstrated the relationship between cerebral blood flow (CBF) in ^{99m}Tc-HMPAO SPECT and shunting effectiveness. It showed that extremely poor CBF in patients predicts an unfavorable outcome from shunting with irreversible brain damage caused by the process of NPH [10].

In conclusion, we present an interesting case of radionuclide cisternography which shows the "butterfly shape" of the lateral ventricles on co-registered SPECT/CT image. As compared with the conventional planar images, SPECT/CT provides improved diagnosis of NPH. In addition, we discuss the possible etiology of shunt failure in our patient and new criteria to determine if a shunt procedure is efficacious for NPH patients.

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正常腦壓水腦症：融合電腦斷層模組之單光子斷層造影的表現

莊紫翎[1] 許明欽[2] 王昱豐[1,3]

[1]佛教大林慈濟綜合醫院 核子醫學科
[2]佛教大林慈濟綜合醫院 神經內科
[3]慈濟大學 內科學系

一位65歲女性於2007年3月遭受頭部創傷，產生蜘蛛膜下腔出血及水腦症，於是病患接受腦室腔分流 (VP shunt) 手術。然而，病患卻漸漸虛弱，並且疾病進程演變成緘默症。爲了瞭解腦脊髓液的動態，我們替病患進行腰椎穿刺，於腦脊髓膜內注射了6 mCi的鎝-99m-DTPA，並施行腦池造影。平面影像顯示典型的正常腦壓水腦症之心形顯影，我們同時收取融合電腦斷層模組之單光子斷層造影 (SPECT/CT)，其橫切面影像顯現左右兩側側腦室活性攝取的蝴蝶形狀標誌。相較於平面影像，SPECT/CT影像更容易清楚地讓核醫醫師一眼就能判別出側腦室放射活性積聚的情形，對正常腦壓水腦症的診斷變得更容易、更精確。

關鍵詞：電腦斷層模組之單光子斷層造影機(SPECT/CT)，正常腦壓水腦症，腦池造影

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聯絡人：王昱豐醫師 嘉義縣622大林鎮民生路2號 佛教大林慈濟綜合醫院核子醫學科 電話：(05)-2648000轉5700 傳眞：(05)-2648508 電子信箱：nment@ms7.hinet.net