

Endoscopic assisted Occipital
Ventriculo-Peritoneal Shunt for
Pagetoid HydrocephalusJoel Caballero García^{1*}, Adolfo Michel Giol Álvarez², Iosmill Morales Pérez¹ and
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Abstract

Hydrocephalus secondary to bone remodeling of cranial base in Paget's disease is rare with few cases reported in the post TC era. There were not previous reports of endoscopic assisted ventriculo peritoneal shunts in these cases. We describe an elderly lady, diagnosed to have Paget's disease who suffered dementia, gait disturbances and urinary incontinence. Obstructive hydrocephalus secondary to cranial base crowding was present. Fiberoptic intubation was doing and an endoscopic assisted occipital ventriculo-peritoneal shunt was inserted. She improved immediately following CSF diversion. Hydrocephalus in Paget's disease is an uncommon and challenging complication. Timely surgery yields good results. There are some anesthetic and surgical precautions that we need to take account in order to ensure good results. Endoscopic visualization ensures an optimal colocation of ventricular catheter far too choroid plexus minimizing the risk of shunt failure and a subsequent reintervention in these difficult cases.

Introduction

Paget's Disease (PD) is a chronic disorder characterized by increased but disorganized bone remodeling accompanied initially by an increase in osteoclastic bone resorption followed by a secondary increase in osteoblastic activity [1]. Although about half of the cases are asymptomatic and are incidentally diagnosed, 10-30% of patients experience pain, skeletal deformity, pathologic fractures or neurologic symptoms [2].

Hydrocephalus secondary to bone remodeling of cranial base in PD is rare with less than 20 cases reported in the post Computer Tomography (CT) era [3-17]. Cerebrospinal fluid diversion is the treatment of choice in symptomatic hydrocephalus, but the anatomical distortion of cranial vault increase the risk of suboptimal shunt colocation during a blend procedure, future dysfunction and subsequent reintervention in these complex patients. There were not previous reports of endoscopic assisted ventriculoperitoneal shunts in these cases.

Case report

We report a 60-year-old black female high school teacher with a known history of PD treated with oral bisphosphonates presented with bilateral neurosensorial deafness for five years and progressive urinary incontinence, gait abnormalities and memory deficits (Hackim-Adams syndrome) for 2 years.

Clinical exam showed bilateral moderate neuroasensorial deafness with no focal motor or sensory deficits. Her head was enlarged (circumference 71 cm) with the characteristic morphology of PD. Laboratory tests were normal except for markedly elevated alkaline phosphatase (1390 units). Skull X-rays showed patchy osteolytic and sclerotic areas with thick cranial vault and basilar impression (Figure 1).

The head CT scan revealed in the bone window an enlarged skull with thickening of the vault bone and widening of the diploe measuring almost 3 cm. The basal skull bones including the clivus and petrous were also enlarged. Brain window showed dilatation of both lateral ventricles and anterior part of third ventricle with periventricular edema. The subarachnoid space and fourth ventricle was collapsed (Figure A-C).

The patient was planned for an endoscopic occipital ventriculoperitoneal shunt in order to treat the hydrocephalus providing an optimal ventricular catheter colocation far too choroid plexus thus preventing a future obstruction and complications of a blend procedure in distortional anatomy for Paget's disease. A restricted mouth opening, a short neck with limited extension predicted difficulty with tracheal intubation and a nasotracheal intubation was doing without significant problems. Positioning was difficult and a left side movement of the table was necessary in order to put the



Figure 1: Lateral skull X-rays showed patchy osteolytic and sclerotic areas with thick cranial vault and basilar impression. The skull has a "cotton wool patch" appearance.

antero-posterior head vertice parallel to the floor. The monitor was positioned to the left side in front of both surgeons. A "U" shape incision with inferior base and a burr hole was performed in the right side 5 cm upward and 3, 5 cm lateral to the in ion with an elliptical shape using a high speed drill in order to provided sufficient space for ventricular catheter and scope (Figure 3). Both outer and inner tables and diploe was very soft and extremely vascular. The bleeding was controlled with bone wax. Distal catheter (with a low pressure valve) was inserted subcutaneously. Before cauterization, the dura



Figure 3: On the left, the patient position with the antero-posterior head vertice parallel to the floor. The monitor was positioned to the left side in front of both surgeons. A "U" shape incision with inferior base and a burr hole was performed in the right side 5 cm upward and 3,5 cm lateral to the in ion. On the right, endoscopic visualization of right ventricle showing the corpus callosum (cc), the choroid plexus (p), the frontal horn (f) and the catheter (c). Discussion.

was opened in an X fashion. Fourth centimeter of ventricular catheter was inserted 15 grades cephalic and 15 grades medial to the nasion point to reach the occipital horn and atrium. Then the guide was gradually removed and the catheter advanced in order to reach the frontal horn of right lateral ventricle in a blond fashion until 9 cm of catheter was introduced. A 0 grades neuroendoscope (GAAB system) was introduced for the first surgeon following the inserted catheter immobilized by the assistant until it reaches the ventricular cavity. Then an anatomic identification of landmarks structures of right lateral ventricle (corpus callosum, septum, choroid plexus, frontal and temporal horn) was carried out. The fenestrated portion of ventricular catheter was collocated in optimal position (in the frontal horn, far too choroid plexus). Finally a posterior septostomy was carried out and the GAAB system was removed stopped any bleeding in their trajectory with Ringer solution irrigation. The distal catheter was introduced in to peritoneal cavity under directed visualization of peritoneum by means a periumbilical incision. The patient was extubated in the operation room and monitored in the ICU for 24 hours. The urinary catheter was removed by the first 24 hours without urinary dysfunction. She improved remarkably becoming independently ambulant and coherent by the second day. 24 hours postoperative CT scans show a correct ventricular catheter collocation in the frontal horn of right lateral ventricle without complications and she was subsequently discharged in the third postoperative day. At 3 months TC scan showed a normal ventricular system with the catheter in the right ventricle frontal horn (Figure 2 D-F).

Discussion

First described by Sir James Paget in 1876 PD of bone is actually the second most common bone disease after osteoporosis [2]. Normal bone is replaced by a disorganized, hypertrophic and softened osseous structure that is prone to deformity and fracture. Interesting, although Paget’s disease of bone is commonly reported in countries with Caucasian populations and it is to be rare in oriental and African populations [1] our patient was afro descendent maybe due the fact of mixture nature of our population. In spite of that, the other characteristics of our patient were very similar to those reported

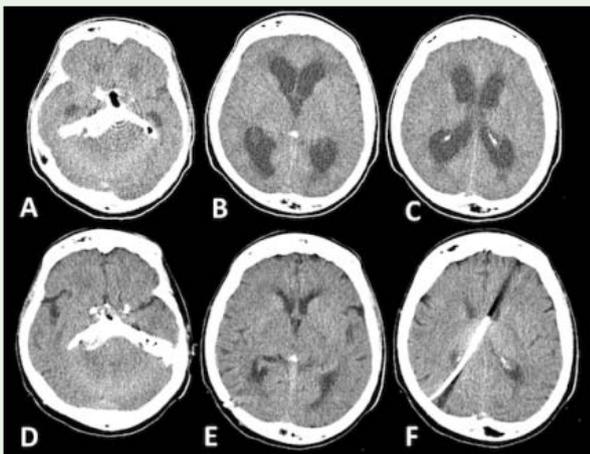


Figure 2: Head CT scans of the patient. A-C: preoperative CT scan showed an enlarged skull with thickening of the vault bone and widening of the diploe. The basal skull bones including the clivus and petrous were also enlarged. The third and both lateral ventricles were markedly dilated with periventricular edema. The subarachnoid space and fourth ventricle was collapsed. D-F: CT scan obtained 3 months after surgery showed a normal ventricular system with the catheter in the right ventricle frontal horn. Subarachnoid space can observe. Is possible that the tip of catheter were crossing the wide septostomy hole to the contralateral frontal horn, but far to the choroid plexus.

by previous studies: the slight predominance of female gender, the mean age at diagnosis, the distribution of affected bones and the predominance of polyostotic PD of bone [2-18].

This disease has skeletal, cardiovascular and neurological complications that produce symptoms in about 10–30% of patients. Neurological involvement is among the more serious clinical problems encountered, and can occur for direct compression of neural structures by the enlarging bone. Deafness is the most common special sensory deficit [19] (our patient have bilateral neurosensorial deafness). Other mechanisms include softening of the bone, propensity for fractures, malignant transformation and ischemia secondary to arteriovenous shunts. Headache and deafness are the commonest neurological symptoms [14]. Skull enlargement of cranial vault and cranial base was documented in 17% of patients [19]. Basilar impression as a result of softening of the skull base occurs in about a third of the patients [15]; however it is a rare cause of obstructive hydrocephalus due to blockage of the Sylvius aqueduct [19]. More commonly, there is a slowly progressive neurologic syndrome secondary to a communicant hydrocephalus (Hackim Adams syndrome) [14]. Interesting, our patient presented with the clinical trial of normal pressure hydrocephalus; however, the CT demonstrated the overgrowth of the middle and posterior fossa bony elements with obliteration of basal cisterns, isolated dilatation of third and both lateral ventricle and absence of subarachnoid space suggesting obstructive hydrocephalus. At surgery, on tapping the ventricles Cerebrospinal Fluid was obtained under moderated pressure suggesting some released mechanism that explains the chronic presentation.

These patients are not only a surgical challenge but anesthetic challenge too. Frequently they have limited neck extension, a restricted mouth opening, calcifications of laryngeal cartilages and high output cardiac failure due the increasing bone flow. In this patient a nasotracheal intubation was doing without significant problems. However, Moiyadi et al. [14] encountered significant difficulty during the fibre-optic intubation because of a rigid and non-pliant upper respiratory airway cartilage and ultimately intubation with a McCoy laryngoscope was successful.

The spinal involvement and large head can limit the optimal surgical position too. Moreover, a potential instability makes neck manipulation dangerous and challenging. Moiyadi et al. encountered the same problem and had to modify the siting of the burrhole [14]. The pathological bone increased the risk of uncontrolled hemorrhage and their softness the risk of dural and neural damage during a burr hole.

The optimal management of hydrocephalus associated with Paget's disease of the skull is not well documented and is still debated, maybe due the fact of their low frequency. Previously, there have been few reports of patients treated with ventriculoperitoneal shunt [3-17]. Obstructive hydrocephalus is more commonly managed by ventriculoatrial or peritoneal shunting but can be treated by third ventriculostomy. Normal pressure hydrocephalus may also respond dramatically to shunt procedures. We do not perform a third ventriculostomy due the fact that the cranial base remodeling including sellae dorsum are occupying the interpeduncular cistern and increase the risk of procedure failure. Also, the sellae dorsum

is an important landmark for this procedure and their distortion increase the risk of basilar trunk injury. We decided to perform a ventriculoperitoneal shunt using an endoscopic guide in order to ensure a safely and optimal colocation of the catheter (in a pagetoid head without safe landmarks) minimizing the risk of future dysfunction and the subsequent reintervention in this challenge patient. And to perform a septostomy in order to prevent future failure. There is only a previous report of endoscopically assisted occipital ventriculoperitoneal shunt which demonstrated the safely and efficacy especially in hydrocephalus with a displaced ventricular system [20]. It is important to ensure a correct surgical position in order to avoid complications like pneumoencephalus and facilitate the endoscope use parallel to the floor. In fact, we recommended a rotation of the head with the occipital burr hole upper than the ventricular system. This surgical strategy has some advantages: the procedure with visual control minimizes the risk of inadequate or suboptimal shunt position, so decrease the risk of proximal catheter dysfunction due choroid plexus invasion; and we can perform a septostomy (very usefully specially if there is a risk of foramen interventricular obstruction). These advantages minimizing the stress of surgeons and patients. However, a learning curve is necessary in order to avoid complications.

As other authors mentioned, pretreatment with an antiresorptive agent 2-3 months before elective surgery will reduce blood flow in the affected bone and adjacent soft tissue and probably lessen perioperative hemorrhage. We found a very soft and extremely vascular cranial vault in spite of this treatment, easily controlled with bone wax. It is necessary take in count to be careful to avoid perforator injury as the bone may be very soft and give way easily.

The patient improved remarkably becoming independently ambulant and coherent by the second day. Actually she returned to his job (high school teacher) without any difficulty.

Conclusion

Hydrocephalus in Paget's disease is an uncommon and challenging complication. Timely surgery yields good results. There are some anesthetic and surgical precautions that we need to take account in order to ensure good results. Endoscopic visualization ensures an optimal colocation of ventricular catheter far too choroid plexus minimizing the risk of shunt failure and a subsequent reintervention in these difficult cases.

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