
Trigeminal neuralgia in a patient with Dandy-Walker malformation

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Summary Background. Trigeminal neuralgia may be idiopathic or secondary to a number of cranial pathologies. We report a novel case of trigeminal neuralgia associated with Dandy-Walker malformation, which may be an etiologic factor. Case description. A 32-year-old male presented with shock-like pain in the somatosensory distribution of the right trigeminal nerve, which was refractory to all medication. MRI revealed a cystic lesion in the posterior fossa and a hypoplastic vermis. The pain was diagnosed as trigeminal neuralgia and was thought to be secondary to the Dandy-Walker malformation. The trigeminal neuralgia was treated successfully with radiofrequency thermocoagulation rhizotomy (RF-TR). Conclusion. Trigeminal neuralgia may be associated with Dandy-Walker malformation, however an etiologic relationship is not proven. We suggest that traction on the trigeminal nerve may be significant. The posterior fossa cyst of Dandy-Walker malformation may be a complicating factor when considering microvascular decompression to treat the trigeminal neuralgia. Collapse of the cyst at surgery may destabilize the posterior fossa and further deform the trigeminal nerve. We suggest that RF-TR, which is minimally invasive and reliable, may be preferable.

INTRODUCTION

Trigeminal neuralgia is a painful condition of the face characterized by paroxysmal, lancinating and shock-like pain confined to the somatosensory distribution of the trigeminal nerve. There is a hypersensitivity to non-nociceptive stimuli. Medication is the initial treatment of choice. If medical treatment fails, surgical treatment may be chosen, considering the general condition and age of the patient, and the pathology accompanying the trigeminal neuralgia. A number of conditions may accompany trigeminal neuralgia. Cerebellopontine angle schwannomas and other tumors, multiple sclerosis and Chiari malformation have been previously reported.2–4,7 Trigeminal neuralgia with Dandy-Walker malformation, as reported here, may be coincidental; however, there may be an etiological association between Dandy-Walker malformation and trigeminal neuralgia. We suggest that trigeminal neuralgia associated with a Dandy-Walker cyst may be caused by traction and direct deformation of the nerve, rather than vascular compression as is usually thought to be the case.

Dandy-Walker malformation is a congenital condition characterized by paroxysmal, lancinating and shock-like pain confined to the somatosensory distribution of the trigeminal nerve, which had been occurring for 15 years. The pain was refractory to all medication and was provoked by external factors. It originated at the right ear and radiated to the upper lip and the chin. He had been diagnosed with trigeminal neuralgia at another clinic, but medical treatment had failed. His physical examination was normal. On neurological examination, he had hyperalgesia in the distribution of the second and third divisions.

CASE REPORT

A 32-year old male was referred to our clinic with shock-like pain confined to the somatosensory distribution of the right trigeminal nerve, which had been occurring for 15 years. The pain was refractory to all medication and was provoked by external factors. It originated at the right ear and radiated to the upper lip and the chin. He had been diagnosed with trigeminal neuralgia at another clinic, but medical treatment had failed. His physical examination was normal. On neurological examination, he had hyperalgesia in the distribution of the second and third divisions.
of the trigeminal nerve. MRI showed a cystic lesion occupying almost the entire posterior fossa and a hypoplastic vermis (Fig. 1). CT scan with bone windows revealed a very large and deformed foramen ovale (Fig. 2). Based on the radiological findings, the trigeminal neuralgia was thought to be related to the Dandy-Walker malformation. Treatment was with RF-TR to the second and third branches of the trigeminal nerve through the right foramen ovale (Fig. 3). A lesion was made at 70 °C for 60 seconds for each division. Post-operatively, the pain resolved. He developed hypoesthesia confined to the distribution of the second and third divisions of the trigeminal nerve. At 6 months follow-up, he remains pain-free.

**DISCUSSION**

Trigeminal neuralgia is characterized by paroxysmal, shock-like pain in the somatosensory distribution of the trigeminal nerve. It is the most common of the cranial neuralgias. The annual incidence of trigeminal neuralgia is four or five per 100,000. The incidence of Dandy-Walker malformation is one per 25000 or 35000. In over 2500 patients with trigeminal neuralgia in our hospital, this is the only case in which Dandy-Walker malformation may be an etiological factor. It is also the first such report in the literature.

Surgical procedures for the treatment of trigeminal neuralgia include ganglion ablative procedures, microvascular decompression and stereotactic radiosurgery. Microvascular decompression is recommended for young patients with persistent pain and no significant medical or surgical risk factors, as was the case in our patient. Radiosurgery may also be used in the treatment of trigeminal neuralgia, but access and cost may be prohibitive and in our experience, results are less satisfactory. RF-TR is the most commonly used method in the treatment of trigeminal neuralgia in the literature.
most age groups as it is minimally invasive and controlled and selective lesions can be achieved. It can be applied to all three divisions of the trigeminal nerve if needed.5,10,11 Glycerol rhizotomy, another commonly performed ablative procedure, was not used in this case due to the large cerebrospinal fluid space in the posterior fossa into which glycerol may escape. Therefore, RF-TR, a reliable and effective method, was performed with a fine needle to treat our patient.

Cystoperitoneal shunting has also been used in the treatment of Dandy-Walker malformation.6 In the patient presented, it is possible the facial pain may have resolved after a shunt procedure. This would also strengthen the evidence of an etiological relationship between the trigeminal neuralgia and Dandy-Walker malformation. However, we considered the complications of cystoperitoneal shunt to high in this patient.

REFERENCES

INTRODUCTION
Autosomal dominant polycystic kidney disease (ADPKD) is a systemic disorder associated with intracranial manifestations including intraparenchymal haemorrhage and aneurysmal subarachnoid haemorrhage.1 Imaging studies have demonstrated an increased incidence of intracranial arachnoid cysts in patients with ADPKD.2 Chronic subdural haematoma (CSDH) as a complication of arachnoid cyst is well documented3–6 but its association with arachnoid cyst in ADPKD has not been reported.7–8 We report a case of CSDH complicating an intracranial arachnoid cyst in a patient with ADPKD.

CASE REPORT
A 27-year old man presented to his family physician with six months history of intermittent generalized headache. MRI demonstrated an arachnoid cyst occupying the anterior and middle parts of the left middle cranial fossa, and foreshortening of the left temporal lobe (Galassi Type II).9 There was no evidence of haemorrhage (Fig. 1). He was managed conservatively with oral analgesics.

He later presented to our unit with four days history of vomiting and worsening headache. There was no history of recent trauma. He was conscious, orientated and neurological examination was normal. His systolic and diastolic blood pressures were persistently above 170 mmHg and 110 mmHg respectively. This was initially attributed to pain and anxiety.

Blood investigations were normal including coagulation profile and renal function. CT scan showed a left frontotemporal CSDH with extension into the temporal arachnoid cyst (Fig. 2a, b). There was no subarachnoid haemorrhage.

Surgical drainage was performed through two burr holes under general anaesthesia. Intra-operatively, liquefied subdural haematoma was found and there was no acute component. The subdural space was irrigated with warm 0,9% sodium chloride solution and a subdural catheter was inserted for post-operative drainage. The arachnoid cyst was not explored.

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