Oculogyric crisis is an underrecognized oculodystonic reaction most often caused by medications. The exact prevalence of this condition is not available. The reaction is most often associated with the use of neuroleptics; therefore, physicians and surgeons who typically do not prescribe these drugs often do not encounter such cases. The patient we present here posed a diagnostic challenge for several years and underwent repeated shunt revisions with no noticeable improvement. Once the correct diagnosis was suspected and addressed she was completely symptom free. We therefore present this case to highlight an additional differential diagnosis when considering VP shunt dysfunction.

**Case Report**

**History.** This 24-year-old woman with congenital hydrocephalus, bilateral VP shunts since infancy, and a history of epilepsy was referred to the emergency department on multiple occasions with a questionable ventriculoperitoneal (VP) shunt dysfunction. Symptoms included nausea, vomiting, altered level of consciousness, ataxia, and vertical eye deviation. The patient underwent multiple revisions of the VP shunt with transient and questionable improvement. During her visit to the neurology clinic, OGC from carbamazepine was suspected, and the dose was reduced. The patient has been completely asymptomatic for the past 18 months. The authors report this case to increase the awareness of carbamazepine-induced OGC as one of the differential diagnoses for VP shunt dysfunction. (DOI: 10.3171/JNS/2008/109/11/0944)

**Key Words** • oculodystonic reaction • oculogyric crisis • ventriculoperitoneal shunt dysfunction

Oculogyric crisis is an underrecognized oculodystonic reaction most often caused by medications. The exact prevalence of this condition is not available. The reaction is most often associated with the use of neuroleptics; therefore, physicians and surgeons who typically do not prescribe these drugs often do not encounter such cases. The patient we present here posed a diagnostic challenge for several years and underwent repeated shunt revisions with no noticeable improvement. Once the correct diagnosis was suspected and addressed she was completely symptom free. We therefore present this case to highlight an additional differential diagnosis when considering VP shunt dysfunction.

**Case Report**

**History.** This 24-year-old woman with congenital hydrocephalus, bilateral VP shunts since infancy, and a history of epilepsy was referred to the epilepsy clinic in November 2003 for daily episodes of impaired balance, nausea, and vomiting with associated headaches. Some of the more severe episodes were associated with an upward deviation of the eyes. Episodes lasted from several hours to several days. Her mother stated that the patient could intermittently follow commands during most of these episodes. Epilepsy was considered unlikely and she was sent to her neurosurgeon for evaluation of a possible VP shunt malfunction. With persistent symptoms and despite no change in the ventricular size on CT scans, she underwent revision of the ventricular catheter of her VP shunt in January and April 2004 with no significant improvement. Testing of the VP shunt in September 2005 did not demonstrate obstruction or malfunction of the device. The patient was reassessed from the epilepsy clinic in March 2006. At this time prolonged episodes of headaches, nausea and vomiting, decreased levels of consciousness, and ataxia were occurring 2 to 3 times a week.

The patient’s history was significant for 23 shunt revisions by 6 years of age. She was first witnessed to suffer epileptic seizures in the neonatal period, with seizure recurrence at 3 years. At that time she was treated with phenytoin and valproic acid. At 6 years of age carbamazepine was added. At the time of her visit to the epilepsy clinic in March 2006, the patient was taking a total of 2100 mg of controlled-release carbamazepine, 1500 mg of valproic acid, and 15 mg of clobazam in divided doses for her epilepsy. Her past seizures had consisted of focal
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motor activity with secondary generalization. She had not experienced these episodes over the past 6–8 years.

Examination, Treatment, and Outcome. On examination, the patient followed simple commands. None of the recent episodes in question were witnessed in the epilepsy clinic. She had a wide-based gait and she was moderately ataxic. Motor examination was remarkable for right spastic hemiparesis. Muscle stretch reflexes were 3/4 in all 4 extremities. An OGC was suspected at this visit and the dose of carbamazepine was gradually reduced to 600 mg twice a day. The patient has been completely asymptomatic for the past 18 months.

Discussion

The first detailed report on OGC was published in 1928 in association with a case of chronic epidemic encephalitis.6 An OGC is an involuntary forced conjugate ocular deviation of the eyes that is sustained for a variable period of time, lasting intermittently from hours to days. Although more often described with the use of neuroleptics and antiemetics,3 it has also been described in association with carbamazepine.1,4 Patients who experience OGC associated with carbamazepine may also demonstrate ataxia, headache, drowsiness, and lethargy, which are well-known dose-related side effects of this drug, and can mimic the picture of VP shunt dysfunction. Approximately 56% of patients with VP shunts experience shunt blockage in the course of a long-term follow-up.7 The clinical signs of shunt failure are attributed to the acute increase in intracranial pressure and may include headaches, drowsiness, ataxia, diplopia, and an increase in seizures. In a retrospective review of 197 patients with shunt-treated hydrocephalus, 17% of patients developed seizures.5 Therefore, this population is more likely to be taking antiepileptic drugs such as carbamazepine. Consequently these patients are more likely to present with OGC and other carbamazepine-related side effects. It is important to note that the carbamazepine levels during the occurrence of these symptoms can be within therapeutic range; OGC attributed to carbamazepine has been reported in individuals with normal drug levels as was true with our patient. These symptoms are attributed to carbamazepine epoxide, a metabolite that is not measured routinely.

Because our patient needed multiple shunt revisions in the past, her most recent symptoms were also attributed to shunt dysfunction, and she underwent shunt revisions with no significant or lasting improvement. Our patient did not have a change in the ventricular size on CT scanning before or after the procedure. It is important to note, however, that 16% of patients with proven hydrocephalus do not show changes in the ventricular size. Drowsiness is by far considered the best predictor of VP shunt dysfunction.2 Headaches and vomiting are other indicators of a blocked shunt. Our patient’s symptoms dramatically improved and completely resolved within weeks of tapering down the carbamazepine, which is in keeping with published data on carbamazepine-induced OGC. She continues to be asymptomatic at 18 months of follow-up.

Interestingly, in our literature review we did not come across medication toxicity and OGC as one of the differential diagnoses of VP shunt failures. We conclude that OGC is a rare but potentially reversible dose-related side effect of carbamazepine, and should always be in the differential diagnosis, especially when shunt revisions do not seem to alleviate the patient’s symptoms.

Disclaimer

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

References


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