Signal change following posterior fossa tumour resection: evidence of hydrocephalus

Key words: Posterior fossa tumour, hydrocephalus, fourth ventricle, mutism, children.

This report discusses two patients who had neuro-imaging changes suggestive of uncontrolled hydrocephalus following the removal of a fourth ventricle tumour. Patient one had a low-density change on a computed tomography (CT) scan and patient two had increased signal intensity on a T2-weighted magnetic resonance (MR) scan. Both patients were treated for hydrocephalus and subsequently improved with complete resolution of these signal changes. Progressive signal change in the region surrounding the resection bed following removal of fourth ventricular tumours may be due to evolving hydrocephalus. This finding might be a useful indicator of disordered cerebrospinal fluid (CSF) circulation that requires treatment.

Patient 1
A 13-year-old boy presented to the emergency department with a one-month history of nausea and vomiting. On neurological exam he had diminished gag reflex bilaterally. There was no papilloedema. An MR scan showed a posterior fossa mass occupying the fourth ventricle and extending through the foramen magnum, suggestive of an ependymoma (Figure 1a and 1b). There was no evidence of metastatic disease.

An uncomplicated posterior fossa craniotomy was performed with gross total excision of the mass, and pathological analysis confirmed an ependymoma. He became acutely hypertensive shortly after his arrival in the intensive care unit. A CT scan showed a tumour bed haemorrhage with extension into the third and fourth ventricles, and hydrocephalus. An external ventricular drain (EVD) was inserted and the operative site re-explored. Following the second procedure, the patient was noticed to have cranial nerve palsies affecting cranial nerves (CN) V (loss of sensation, right side of face), VII (bilateral facial motor weakness), IX (no cough, gag or swallow) and XII (inability to protrude tongue completely and fasciculations), and was aphonic after extubation.

In addition to the ventriculomegaly that had developed as a result of the tumour bed haemorrhage, low-density changes on CT were found in the resection bed and in the walls of the fourth ventricle (Figure 2a). No other periventricular low density was seen. The EVD was maintained and, over the course of 10 days, several attempts were made to wean him from the drain by gradual elevation. In all attempts, his level of consciousness deteriorated and his ventricles increased in size. It was decided that longer term CSF drainage was required and a ventriculoperitoneal shunt was inserted. Two days later, a CT scan was performed that showed reduction in the size of the lateral ventricles and reversal of the tumour bed low-density change (Figure 2b). The patient has recovered speech but bulbar cranial nerve dysfunction remains.

Patient 2
A nine-year-old girl presented to the emergency department with a one-week history of progressive headache, nausea and vomiting, and a two-day history of worsening vision and unsteady gait. On physical examination she was alert and oriented while showing left-directed horizontal nystagmus and congruent lateral deviation to the left. She had bilateral papilloedema, a positive Romberg's
Patient 1

sign, an ataxic gait and mild sitting ataxia. MR (Figure 3) and CT scans of the brain showed a midline fourth ventricular mass lesion with accompanying hydrocephalus. Periventricular signal change was present adjacent to the frontal and occipital horns of the lateral ventricles. There was no evidence of metastatic disease. An uncomplicated posterior fossa craniotomy was performed with gross total excision of the mass. Pathological analysis confirmed medulloblastoma. The patient was kept intubated overnight and was extubated the following day. A CT scan on the first postoperative day showed decreased ventricular size and no residual tumour. Aside from a mild left facial weakness suggestive of lower motor neuron damage, she displayed no abnormal behaviour, was appropriately responsive and exhibited normal spontaneous speech. Her extraocular movements were normal. On the second postoperative day, the patient complained of severe headaches, developed a left-sided CN VI palsy and a right internuclear ophthalmoplegia. A CT scan showed ventricular dilatation and an EVD was inserted. Approximately 72 hours after the patient’s operation, she was noted to be aphonie. During the next 24 hours, the measured intracranial pressure was low. The patient continued to be aphonie and demonstrated limb and truncal ataxia. The EVD was removed on the eighth postoperative day. On the fourteenth postoperative day, a surveillance MR scan showed that her lateral ventricles were increasing in size and there was significant signal change in the lateral and posterior walls of the resection bed (Figure 4a). The periventricular signal change seen around the lateral ventricles pre-operatively had resolved. An endoscopic third ventriculostomy (ETV) was performed 18 days after the excision of the tumour. The periventricular signal change seen around the lateral ventricles pre-operatively had resolved. An endoscopic third ventriculostomy (ETV) was performed 18 days after the excision of the tumour. By the seventh day post ETV, the signal change around the fourth ventricle had improved (Figure 4b). Within 24 hours, the patient’s speech partially returned and she was able to understand and answer questions using two- to five-word sentences.
Discussion

The most common solid tumours in the paediatric population are brain neoplasms, accounting for 16% of all malignancies, 43% of which occur in the posterior fossa. The most common tumours include astrocytomas (52%), medulloblastoma/primitive neuroectodermal tumours (21%) and ependymomas (9%). The current overall five-year survival for all brain tumours in childhood is 67%.

Hydrocephalus is a common complication of posterior fossa tumours and results from derangement of CSF production, circulation, or drainage. Non-communicating hydrocephalus results from obstruction within the ventricular system while communicating hydrocephalus results from decreased CSF reabsorption or increased production. Diagnosis is suspected by characteristic clinical signs and confirmed with imaging studies. The clinical picture varies with age but typical symptoms include irritability, headache, vomiting, poor appetite, lethargy, change in personality and deterioration in school performance. Infants (whose fontanelle is open and cranial sutures have not fused) often present with accelerated head growth, a bulging anterior fontanelle and, occasionally, prominent veins. Clinical signs include papilloedema, muscle weakness, brisk deep tendon reflexes, spasticity, clonus and a Babinski reflex. The ‘sun-setting’ sign is also classically observed when the dilated suprapineal recess impinges on the tectum, forcing the eyes to deviate downward. Imaging studies confirm the diagnosis and can help to identify the underlying pathology and the severity of the hydrocephalus. CT, MRI and transcranial ultrasonography in the infant (through a patent fontanelle) are the most common diagnostic studies. Plain film x-rays of the skull may show separation of the cranial sutures.

Treatment of hydrocephalus depends on the underlying pathology. Diuretic medications (furosemide and acetazolamide) and EVD insertion can provide temporary relief, but long-term treatment usually requires extracranial shunting of CSF with a ventriculoperitoneal or ventriculo-atrial shunt, or a ventriculostomy. Major complications of shunts are occlusion and infection requiring shunt revision.

Hydrocephalus secondary to brain tumours, in particular those occupying the fourth ventricle, are managed in a variety of ways. The most effective treatment is gross total resection of the tumour in order to restore normal CSF circulation. However, obstructive hydrocephalus may remain in 15-30% of patients and requires further treatment. In the early postoperative period, it is not always apparent that the hydrocephalus has resolved. In this period, clinical signs and any changes in ventricular size or draining CSF are used to determine if further treatment is needed.

Perventricular low densities on CT are often seen in the lateral ventricles in hydrocephalus. It is thought to represent transependymal flow of CSF associated with increased diffusion. It is less commonly seen in the fourth ventricle.

Using pre-treatment clinical features, Riva-Cambrin et al have devised a model to predict the need for hydrocephalus treatment following tumour resection. The parameters are age <2 (score of 2), papilloedema (score of 1), moderate to severe hydrocephalus (score of 2), cerebral metastases (score of 3), and specific tumour pathologies (score of 1). Patients with a score of 0-4 are deemed low risk, while those at 5-10 are high risk. Pre-operatively, both patients in this report were considered to be at low risk for hydrocephalus intervention following resection of the tumour (a score of 3 for patient 1, and 4 for patient 2). Subsequently, their postoperative hydrocephalus was treated conservatively via EVD with the expectation that it would resolve without further treatment.

CT low density or T2 signal change is not normally seen around the tumour site following resection of a posterior fossa tumour. The observed effect of CT low density is presumed to be interstitial fluid, which is likely to be an indication of locally obstructed CSF flow. This is either due to blockage of interstitial fluid flow back into the ventricular system, or the influx of CSF from pressurised ventricles into the periventricular tissues. Thus, the phenomenon of signal change surrounding the posterior fossa tumour bed in the setting of post-resection hydrocephalus suggests that the CSF circulation disturbance is not controlled.

Establishing risk of hydrocephalus pre- and postoperatively is valuable in counselling families and outlining treatment plans. Such marked signal changes in the tumour bed following resection of a posterior fossa tumour may be another sign of ongoing hydrocephalus requiring intervention.

References