Medulloblastoma in a case of migraine-like headache with head tilt: a case report

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Abstract

This is the case of a 7-year-old girl with a history of migraine headaches who presented with a head tilt to the left and worsening headaches. Papilloedema was found on ophthlamoscopy. She had an intermittent history of migraine-type headaches for the past year. Oral naproxen 375 mg twice daily provided some relief of the headaches initially but they slowly worsened and head tilt appeared. A medulloblastoma was diagnosed on computed tomography and the histology was confirmed at surgical resection. There was no evidence of metastatic spread. The patient was given radiotherapy and chemotherapy and was tumor free at 6 months.

Keywords

Medulloblastoma; migraine; papilloedema; intracranial pressure.

Case presentation

A 7-year-old girl with a significant past medical history of intermittent migraine headaches for the past year presented to the clinic with headaches increasing in frequency over the past week. She had a low-grade temperature of 37.2–38.0°C over the past several days, increased irritability, occasional cough and decreased appetite but she denied nausea or vomiting. The headache was located bitemporally and at the right occipitotemporal area and was pounding in nature. It was reduced by lying down in a dark room. The headache was relieved by Motrin or Tylenol by mouth but kept returning after several hours.

She was placed on oral naproxen 375 mg twice a day, which improved the headache bringing it from 6/10 to 2–3/10 in intensity. However the headache returned at night. She was admitted to the ward to for monitoring and evaluation. Suspecting possible meningitis, a careful ophthalmologic examination was performed. Papilloedema was suspected on ophthlamoscopy, and head tilt was seen to the left by the time she was examined in the ward. Any coughing episode increased the headache intensity temporarily. She did not complain of diplopia and merely said that tilting the head relieved some of the headache. No neck stiffness was found and the Brudzinski sign was negative. There was no photophobia and no ataxia. No other neurological abnormality was seen. She had no nausea or vomiting; she could tolerate fluids and food by mouth but had a reduced appetite. The remainder of the examination was within normal limits.
There was a positive family history for migraine headaches in a maternal aunt, beginning at 12 years of age, and there were primary brain tumors and primary and metastatic colon cancers on the father’s side of the family and metastatic breast cancer on the mother’s side of the family.

Suspecting an intracranial mass lesion, a contrast-enhanced magnetic resonance imaging (MRI) study of the brain was performed and this revealed a primary brain tumor. Once a primary brain tumor was diagnosed based on the MRI scan with radiological evidence of impending foramen magnum herniation, immediate consultation with a pediatric neurosurgeon in a tertiary care facility was arranged. Based on the clinical features and the MRI findings, intravenous dexamethasone was started immediately and arrangements were made to transfer the patient with her family to the tertiary care center.

**Investigations**

MRI with gadolinium contrast revealed a right-sided posterior fossa hemispheric mass lesion measuring 6 cm in diameter with a 4 cm medially enhancing mural nodule. Figs. 1–4 show the tumor causing a pressure effect on the brain, herniation of the cerebellar tonsils 12 mm below the posterior margin of the foramen magnum and a mass effect on the brain stem with impending cerebellar herniation and brain stem compression. There was obstruction of the Aqueduct of Sylvius, leading to dilatation of the third and lateral ventricles with obstructive hydrocephalus. The tumor was considered to be a multicystic astrocytoma; or less likely a medulloblastoma. Gadolinium-enhanced MRI of the spine was performed in the tertiary care facility to look for metastatic deposits but none were seen. Cerebral spinal fluid (CSF) was examined to check for neoplastic cells and this too was negative.

**Fig. 1.** Right posterior fossa hemispheric cystic mass measuring 6 cm with a 4 cm medially enhancing mural nodule.

**Fig. 2.** Downward herniation of the cerebellar tonsils which project 12 mm below the posterior margin of the foramen magnum.
Differential diagnosis

- Primary brain tumor;
- Brain abscess due to the well-circumscribed margin and the appearance of fluid within a cystic cavity; history of low-grade fever and cough preceding admission also supported this diagnosis;
- Non-neoplastic hydrocephalus (before obtaining the MRI).

Treatment

Pre-transfer treatment was aimed at reducing the intracranial pressure by elevating the head of the bed by 30 degrees, giving intravenous dexamethasone 4 mg/kg at once and at 6 h while awaiting an improvement in the weather for helicopter transport. After transfer, surgery was performed to remove the primary tumor. A temporary shunt to relieve the intracranial pressure was placed at surgery but following removal of the primary tumor, CSF shunting was not needed for long. After excision the patient had an uneventful recovery. She was treated with radiation therapy to the posterior fossa to prevent tumor recurrence as well as to the craniospinal axis. Adjuvant multi-agent chemotherapy with vincristine, cisplatin and cyclophosphamide was also given.
Outcome and follow-up

The patient has remained tumor free for 6 months. She has suffered some delayed effects of radiation therapy such as hypothyroidism, treated with oral thyroxine replacement.

Discussion

Medulloblastoma is a highly malignant primary neoplasm that originates in the cerebellum or posterior fossa. Previously, medulloblastomas were thought to represent a subset of primitive neuroectodermal tumors (PNET). However, recent gene expression profiling has shown that some medulloblastomas have a distinct molecular profile and are distinct from other PNET tumors. This prompted the World Health Organization to place them in a class of their own[1]. The histologic appearance is of a highly cellular tumor with little cytoplasm and dark-staining round or oval nuclei.

Medulloblastoma is the most common malignant brain tumor of childhood, causing 33% of all infratentorial tumors in children. (Gliomas are the most common tumors of the central nervous system (CNS) overall in childhood causing 50–60% of brain tumors in children). The incidence of medulloblastomas is 20% with a peak incidence between 5 and 9 years of age. The 5-year survival for medulloblastomas is 50–80%.

Approximately 2–5% of children with medulloblastomas have inherited disorders with germline mutations of specific genes that predispose to the development of medulloblastoma. These include Gorlin syndrome and the less common Turcot syndrome, which consists of medulloblastomas in conjunction with familial adenomatous polyposis and adenomatous polyposis coli[2].

Typically children with medulloblastoma present with morning headache, vomiting and lethargy. Truncal or limb ataxia and duration of symptoms for less than 3 months prior to diagnosis are common. Head tilt due to cranial nerve dysfunction (4th or 6th cranial nerve dysfunction) can occur[3]. Hydrocephalus is present in 75% of patients at presentation.

MRI studies showed the tumor mass compressing the 4th ventricle leading to obstructive hydrocephalus. This tumor is often not seen on computed tomography (CT) scan. In one-third of cases, medulloblastomas metastasize throughout the CNS following CSF pathways[4]. Elevated intracranial pressure (ICP) can result from vasogenic edema caused by the tumor. In this case the cerebellar tonsils were herniating through the foramen magnum when the imaging study was done. General measures to reduce the intracranial pressure include:

- Rapid treatment of hypoxia, hypercarbia, and hypotension;
- Elevation of the head of the bed from 15 to 30 degrees;
- Aggressively treating fever with antipyretics and cooling blankets;
- Control of shivering in intubated patients with muscle relaxants (e.g. vecuronium, rocuronium);
- Administering prophylactic phenytoin or phenobarbital to patients who are at high risk of developing seizures;
- Maintaining adequate analgesia to blunt the response to noxious stimuli.

Specific measures to reduce ICP include mannitol, hypertonic saline, hyperventilation and/or barbiturate-induced coma in patients who require intubation and ventilation. Corticosteroids are not useful in most cases of increased ICP, however they are useful short-term for vasogenic edema caused by tumors or brain abscesses such as in this patient. Indomethacin[5] is a new

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Table 1. Prognostic factors in children with medulloblastoma[8]

<table>
<thead>
<tr>
<th>Factor</th>
<th>Favorable</th>
<th>Unfavorable</th>
</tr>
</thead>
<tbody>
<tr>
<td>Extent of disease</td>
<td>Nondisseminated</td>
<td>Disseminated (especially brainstem infiltration)</td>
</tr>
<tr>
<td>Size of primary tumor</td>
<td>Less than 3 cm (completely resected)</td>
<td>More than 3 cm</td>
</tr>
<tr>
<td>after surgery</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Histologic features</td>
<td>Undifferentiated</td>
<td>Foci of glial, ependymal or neuronal differentiation</td>
</tr>
<tr>
<td>Age (years)</td>
<td>≥4</td>
<td>&lt;4</td>
</tr>
</tbody>
</table>

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experimental approach to reducing ICP as it provides analgesia as well as cerebral vasoconstriction. Hypothermia is another experimental approach. Contraindicated treatments in cases of increased ICP include nitroglycerin, nitroprusside, ketamine, prolonged propofol infusions and hypotonic saline (e.g. 5% dextrose in water (D5W)).

The only effective treatment is neurosurgery. Surgical resection of all or most of the tumor is possible in most cases unless it has spread to the 4th ventricle or cerebellar peduncle. Postoperative lumbar puncture to look for tumor cells and gadolinium-enhanced MRI to look for leptomeningeal involvement (for neuroaxial drop metastasis) should be performed. One-third of all cases have metastasized throughout the CNS by the time the diagnosis is made.

Complications of resection include cerebellar mutism syndrome which occurs in 25% of patients and consists of decreased speech (often mutism)[6], cerebellar dysfunction, behavior changes (irritability or apathy) and other neurologic anomalies. The mutism usually resolves in weeks to months of surgery but the dysmetria and cerebellar ataxia often persist.

Complications of radiotherapy, particularly in young infants, include significant neurocognitive deficits, endocrine sequelae such as hypothyroidism and hypopituitarism and growth deficits[7]. Chemotherapy can lead to cushingoid features and immunosuppression.

Radiotherapy after surgery is very important. Chemotherapy is also frequently used in disseminated disease. New treatment options in poor risk candidates for radiotherapy (young infants) include high-dose chemotherapy with autologous stem cell transplantation[9].

The case reported here was unusual in that the child lacked many of the presenting features including vomiting, ataxia, morning headache and lethargy. Past history of chronic migraines was a further confounding factor. A family history of primary brain tumors and colonic cancer might point to genetic predisposition to tumors as discussed earlier.

Vague symptoms of low-grade fever and cough for more than 1 week pointed toward possible meningitis. In view of that, a lumbar puncture might have been performed resulting in catastrophic cerebellar herniation. However, a rise in body temperature can also occur in response to central dysregulation of the temperature mechanism in the brain in the presence of a tumor.

The presence of papilloedema and a head tilt are not uncommon in the presence of severely raised ICP secondary to intracranial tumors. Even though the presence of these signs pointed toward an intracranial space occupying lesion, the absence of these signs should not completely remove the suspicion of a space occupying lesion and a brain MRI or CT head should be ordered where there is the slightest possibility of an intracranial lesion.

After the diagnosis of an intracranial space occupying lesion with raised ICP, timely dexamethasone was administered leading to a reduction in ICP while awaiting transfer to a tertiary facility with neurosurgical capabilities.

The presence of a head tilt and the finding of papilloedema were important factors in obtaining an imaging study. It is important to remember to get a CT head or a brain MRI at the slightest suspicion of an intracranial lesion as these signs are often absent in intracranial space occupying lesions. The case was also unusual in that her tumor was on the right side, which is more common in adults; a central cerebellar tumor location is more common in children.

A similar case of a 36-year-old male smoker complaining of occipital headaches associated with photophobia, diplopia, nausea, vomiting and ataxia has recently been published[10]. An MRI of the brain showed two focal lesions of the medulloblastoma in the right cerebellar hemisphere. After prolonged treatment for migraine he was finally diagnosed as having a brain tumor.

Teaching points

- Migraine-like headaches with head tilt may indicate underlying intracranial masses.
- Lumbar puncture for suspicion of meningitis without performing a CT scan can lead to catastrophic cerebellar herniation if signs of increased ICP are missed[11].
- Intravenous dexamethasone if administered promptly at the first sign of raised ICP caused by malignant vasogenic edema and abscesses can prevent cerebellar herniation[12].

References


