Case Report: Medulloblastoma Presented with Bilateral Sixth Cranial Nerve Palsies

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Abstract

A 16 years old healthy man presented with history of increasing frequency of headache and vomiting , notes of blurred vision and binocular horizontal diplopia for 1 month. Clinical findings revealed bilateral sixth nerve palsies with bilateral optic disc swelling. With all of these findings, this seem to be a case of raised intracranial pressure. Neuroimaging demonstrated a midline posterior fossa tumor with obstructive hydrocephalus. The patient underwent ventriculoperitoneal shunt and subtotal tumor removal. Pathological examination revealed histological characteristic findings of medulloblastoma. Patient was received the craniospinal radiation and chemotherapy to improve the survival. On follow-up visits, ocular examination revealed his diplopia improved and resolved of swollen disc both eyes.

Introduction

Medulloblastoma is the most common malignant brain tumor in children[1]. Early symptoms are secondary to increased intracranial pressure such as headache and vomiting. The earliest sign are nonlocalized and caused by increased intracranial pressure. Visual disturbances more commonly are a result of papilledema. Patient may develop double vision as the sixth cranial nerve becomes stretched from the hydrocephalus.[2] Neurological deficits in extremity coordination and gait are common. Later signs due to brain stem invasion may cause conjugate gaze palsy and combination with deficits of multiple cranial nerves. [3]

We describe the presentation of medulloblastoma in a young man presented with chronic intermittent headache and vomiting with increasing frequency of headache, blurred vision and binocular horizontal diplopia. The fundus evaluation revealed papilledema. The neurological examination revealed bilateral sixth nerve palsies and truncal ataxia.

The presence of bilateral sixth nerve palsies and papilledema is seem to be evidence of increased intracranial pressure require urgent neuroimaging.[4] Isolated sixth nerve palsies may be non-localizing signs but the presence of ataxia is also the localizing sign suggested the cerebellar involved lesion.[5]

Case report

A 16-year-old healthy man presented with chronic headache , increasing frequency of headache during the past 2 months. Headache was became worse in the morning and associated with intermittent vomiting. He also notes of blurred vision and binocular horizontal diplopia about 1 month. The parents also noticed that patient had gait imbalance. He had history of closed head trauma for 1 year ago. There was no significant

drug history and no family history of any ocular abnormalities.

Ocular examination revealed best corrected visual acuity were 20/30 in both eyes. Extraocular movement examination revealed limited abduction of both eyes with esotropia 20 prism diopter in primary position and increased esoropia in both ipsilateral gazes (Figure 1).

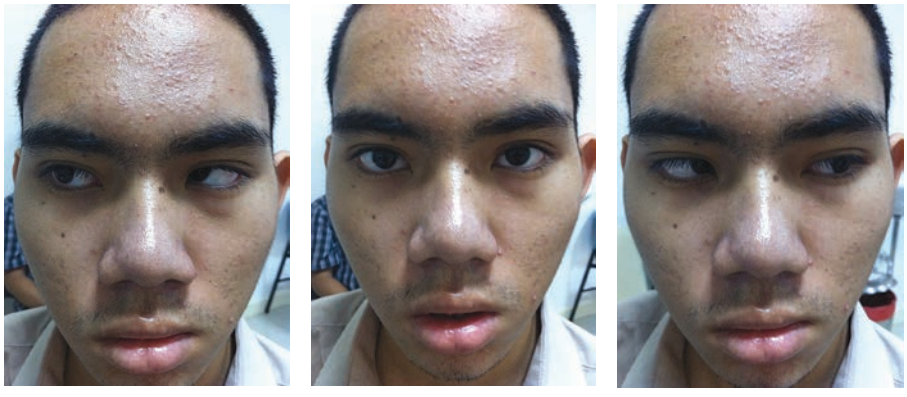


Figure 1 Extraocular eye movement examination show mild esotropia in primary gaze and abduction deficit in both horizontal gazes.

Slit lamp examination : no abnormalities of anterior segment in both eyes. The pupils were 3 mm react to light both eyes without afferent pupillary defect (RAPD). The fundus examination reveal bilateral optic disc swelling with inferior peripapillary flame shaped hemorrhage in the right eye . (Figure 2). Neurological examinations revealed truncal ataxia .The color vision test showed normal and the perimetry showed the enlarged blind spot in the both eyes.

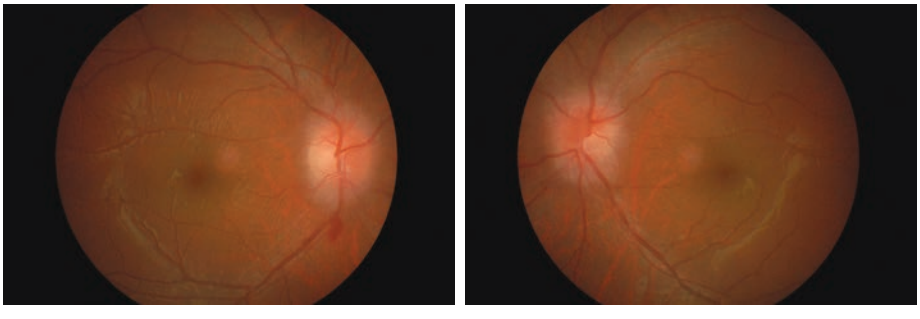
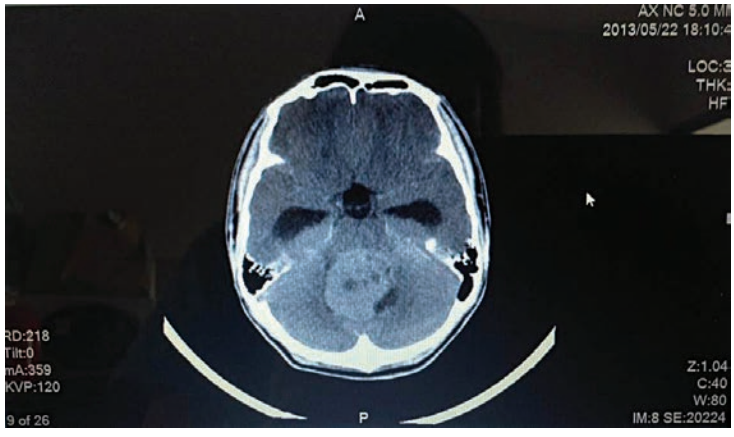


Figure 2 Fundus photographs show bilateral optic disc swelling with peripapillary flame shaped hemorrhage in the right eye.

This case is diagnosed bilateral sixth cranial nerve palsies with papilledema. Computerized tomography brain demonstrated a large heterogenous high density mass within fourth ventricle, likely arise from the ventricle floor . Bilateral tonsilar herniation and diffuse brain swelling is presented. The possible diagnosis are medulloblastoma or ependymoma with obstructive hydrocephalus. (Figure 3)



Figures 3 Axial plain CT brain demonstrating evidence of hyperdensity mass involving the fourth ventricle. Shifting of the midline structures is observed.

Further magnetic resonance imaging of brain and whole spine were done for metastasis evaluation. There was about 3.5x3x6x4.1 cm fourth ventricular mass causing moderate hydrocephalus and mild tonsilar herniation (Figure 4). There was no evidence of leptomeningeal metastasis and no gross abnormality detected in spinal cord (Figure 5).

Figure 4 Magnetic resonance imaging brain demonstrating large heterogenous intraventricular mass in the enlarged fourth ventricle size about 3.5 x 3.6 x 4.1 cm in AP,transverse and CC dimensions respectively.

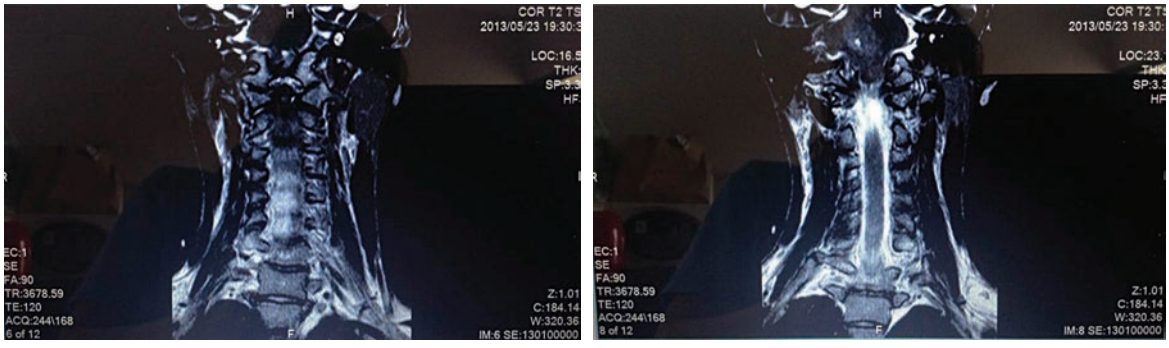


Figure 5 Magnetic resonance imaging whole spine demonstrating the spinal cord is normal size and signal intensity.

The patient was underwent ventriculoperitoneal shunt in the next day. The cytology test of the cerebrospinal fluid specimen revealed some spindle cells in foamy background and few degenerated neutrophils. Differential diagnoses include involvement by reactive gliosis, granulomatous inflammation and neoplasm. Histological examination of the mass is recommend for definite diagnosis. The patient was underwent subtotal posterior fossa tumor removal biopsy and imprint cytology. The intraoperative findings of tumor mass locate on cerebellar vermis and protrude to fourth ventricle with brain stem invasion. The pathology report demonstrated round cell tumor with rosettes shape, most likely diagnosed of medulloblastoma, WHO grade IV. The postoperative magnetic resonance imaging brain revealed small amount of contrast and non contrast lesion around the fourth ventricle which can be either residual tumor or surgical that attended to stop bleeding intraoperative . The clinical symptoms of patient was improved . He was alert, well oriented without ataxia and did not complaint of headache or vomiting. On follow- up at two weeks, the ocular examination revealed visual acuity 20/30 and still had abduction deficit in the same amount. The fundus examination revealed improving of the sectoral swollen discs in his both eyes (Figure 6).

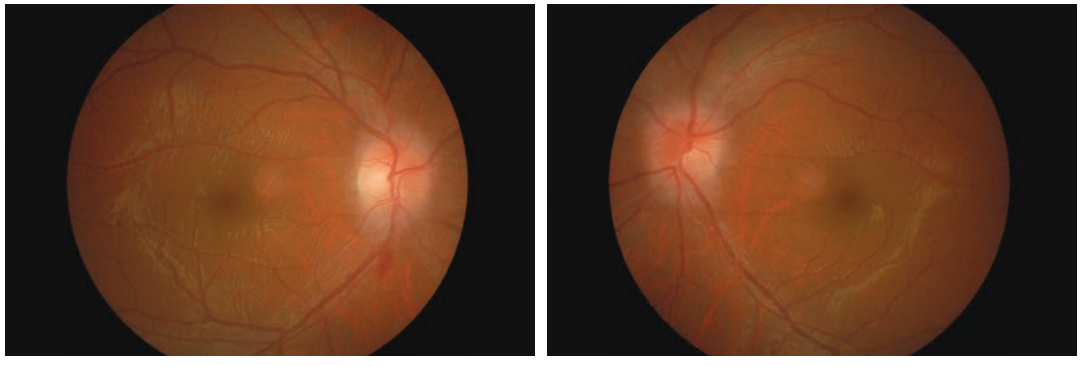


Figure 6 Fundus photographs show improved sectoral of swollen disc in both eyes

At one month follow-up, his diplopia had improved. The fundus examination revealed completely improving of swollen discs in his both eyes and the flame-shaped peripapillary hemorrhage in the right eye was spontaneously resolved (Figure 7).

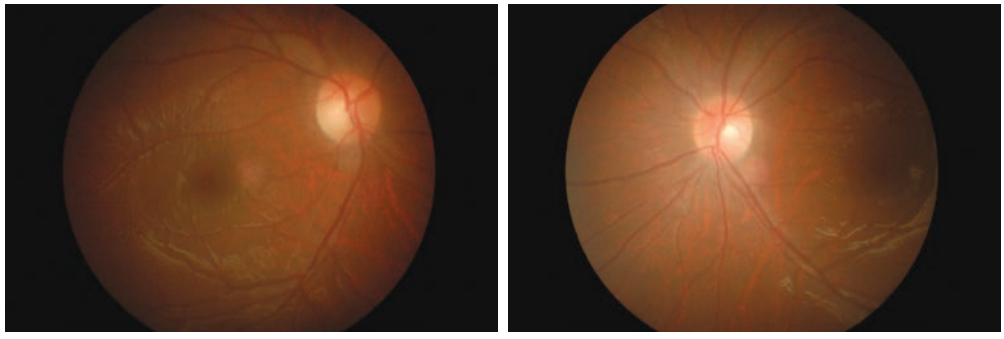


Figure 7 Fundus photographs show completely resolved of swollen discs in both eyes and resolved the previous flame- shaped peripapillary hemorrhage in the right eye.

This patient was received the whole craniospinal radiation with adjuvant chemotherapy and serial follow up of complete recovery of diplopia from sixth nerve palsies.

Discussion

Medulloblastoma is the most common malignant brain tumor in children. It is a cerebellar tumor arising predominantly from the cerebellar vermis. Patients usually have symptoms related to increased intracranial pressure as a consequence of hydrocephalus. Headache is often upon awakening that is relieved by vomiting. Initial signs of increased intracranial pressure may present with visual symptoms from papilledema. It may associated with diplopia from cranial nerves dysfunction due to extension of tumor in the brainstem or presence of raised intracranial pressure in the subarachnoid spaces through which the nerve course[6].

Binocular horizontal diplopia is the main symptom of sixth nerve palsy. The sixth cranial nerve innervates the lateral rectus muscle, which is responsible for abducting the eye. Diagnosis of sixth nerve palsy is based on the clinical findings of partial or complete abduction deficit. The abduction deficit may be subtle and manifest as a small esodeviation, slowed abducting saccades and diplopia worse on ipsilateral gaze[7]. With bilateral palsies, diplopia increases on both right and left gaze. Once a diagnosis of sixth nerve palsy has been established, a complete ocular and neurological evaluation should be performed to determine if the palsy has occurred in isolation. Causes of sixth nerve palsy includes tumor, trauma, meningitis and vascular disorders[8]. The presence of isolated sixth nerve palsy, the age of patient indicate the next step management. All children and adults under 40 years of age should have neuroimaging. Adults over 40 years of age with

vasculopathic risk factors do not require neuroimaging, but should have investigation of systemic vascular disorders as first step. Neuroimaging should be performed if the palsy fails to resolve in 3 months or the patient have multiple cranial neuropathy or new neurological deficit[9].

In children and young adult, tumors cause 30% of sixth nerve palsies[10]. Tumor origins are usually brainstem glioma or posterior fossa tumors such as medulloblastoma, cerebellar astrocytoma or epndymoma. Its long course in the subarachnoid space make the sixth nerve vulnerable to stretch and have indirect effects following increased intracranial pressure result in papilledema as non-localizing sign.

We report this case of medulloblastoma whom present the mainly ocular symptoms. A careful history taking with complete ocular and neurological examination are essential for establishing the diagnosis especially evidence of increased intracranial pressure such as significant headache and bilateral optic disc swelling (papilledema). Neuromaging and cerebrospinal fluid analysis are the mainstays of the diagnostic approach for proper further management.

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รายงานผู้ป่วย: Medulloblastoma presented with bilateral sixth cranial nerve palsies

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บทนำ

เนื้องอกสมองชนิด medulloblastoma เป็นเนื้องอกสมองชนิดร้ายแรงที่พบบ่อยที่สุดในเด็ก อาการสำคัญที่พบได้ในระยะแรกเริ่มมักเป็นผลมาจากความดันในสมองที่เพิ่มสูงขึ้น ได้แก่ อาการปวดศีรษะและมักพบอาการคลื่นไส้อาเจียนร่วมด้วย อาการเดินเซจากการทรงตัวผิดปกติ ในบางรายอาจมีอาการผิดปกติทางสายตาได้จากเกิดภาวะขั้วประสาทตาบวมจากความดันในสมองสูง บางรายเกิดอาการเห็นภาพซ้อน จากเส้นประสาทที่ควบคุมกล้ามเนื้อกลอกตาทำงานผิดปกติ การให้การวินิจฉัยแยกโรคโดยอาศัยประวัติ การตรวจตาและการทำงานของระบบประสาทอย่างละเอียด ร่วมกับการตรวจภาพถ่ายรังสีพิเศษเพิ่มเติมเช่น CT หรือ MRI และการตรวจน้ำไขสันหลังจะนำมาซึ่งการวินิจฉัยที่ถูกต้อง

รายงานผู้ป่วย

รายงานผู้ป่วยอายุน้อย 1 รายที่มาด้วยอาการปวดศีรษะเรื้อรังเป็นๆ หายๆ อาการปวดศีรษะดังกล่าวเป็นมากขึ้นมา 2 เดือนร่วมกับมีอาการคลื่นไส้อาเจียน ตามัวลงทั้งสองข้างและมองเห็นภาพซ้อนในแนวนอน การตรวจพบมีตาเขเข้าในจากความผิดปกติของกล้ามเนื้อกลอกตา Lateral rectus ทั้งสองข้าง ตรวจจอประสาทตาพบมีการบวมขั้วประสาทตาทั้งสองข้าง การตรวจร่างกายทางระบบประสาทพบมีความผิดปกติในการทรงตัว ให้การวินิจฉัยเบื้องต้นว่ามีภาวะขั้วประสาทตาบวมสองข้างจากความดันในสมองสูง การตรวจเอกซเรย์คอมพิวเตอร์สมองพบเนื้องอกสมองบริเวณ cerebellum ภายหลังการทำระบายน้ำไขสันหลังเพื่อลดความดันในสมอง ผู้ป่วยได้รับการผ่าตัดเอาเนื้องอกสมองออก การตรวจทางพยาธิวิทยาพบลักษณะของเนื้องอกสมองชนิด medulloblastoma ซึ่งต้องได้รับการรักษาด้วยการฉายแสงและการให้เคมีบำบัดเพิ่มเติม การตรวจติดตามผู้ป่วยภายหลังการรักษาพบว่า ไม่มีอาการปวดศีรษะและคลื่นไส้อาเจียน ระดับการมองเห็นดีขึ้น อาการเห็นภาพซ้อนลดลงจากการทำงานของเส้นประสาทตาที่ควบคุมกล้ามเนื้อกลอกตากลับฟื้นดีขึ้นได้