Cerebellar Medulloblastoma with Intraspinal Metastasis Presenting with Acute Paraplegia: A Case Report

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We report a 4-year-old girl with cerebellar medulloblastoma and intraspinal metastasis. This case is rare in its initial presentation with spinal cord compression and the intratumoral hemorrhage within the intraspinal metastasis. This girl received surgery, radiotherapy and chemotherapy immediately after the diagnosis. However, she died of pancytopenia and pneumonia six months after treatment.

Key words: Medulloblastoma, spine, metastasis, magnetic resonance imaging, hemorrhage

We report an unusual case of cerebellar medulloblastoma, which was detected incidentally on MRI examination of the spine with symptoms related to spinal cord compression by intraspinal metastasis. Bleeding in a primary or metastatic medulloblastoma is relatively rare.

CASE REPORT

A 4-year-old girl was referred to the emergency department with urinary retention and paraplegia. She had constipation for one week prior to the admission. She also suffered from progressive weakness which had progressed to complete paralysis of both legs within four days. On examination, she was alert, with sensory loss below the level of T10. Sensation was normal in both arms. The muscle power was grade 5 in both arms but 0 in both legs. Deep tendon reflexes were absent in both legs. The laboratory data was not contributory. Under the impression of spinal lesion, emergency spinal magnetic resonance imaging (MRI) study was performed.

The spinal MRI showed two intradural lesions, one in the cervicothoracic region extending from C3 through T3 and the other at the thoracolumbar junction extending from T9 through L1 (Fig. 1). The cervical spinal cord was enlarged with central hyperintensity on T2-weighted images. This lesion showed no enhancement after contrast media administration, suggesting edematous changes in the spinal cord. The T10 lesion with hyperintensity on T1-weighted images was an intradural extramedullary bulging mass with hemorrhage confirmed during surgery (Fig. 2). After use of intravenous contrast, the nodular thickened dorsal dural sheath was enhanced. In addition, a heterogeneously enhanced mass involving the vermis and the 4th ventricle was identified incidentally on sagittal images of cervical spine MRI (Fig. 3). There was abnormal nodular enhancement within the cisterns of the posterior fossa. Brain MRI examination revealed no other intracranial lesions.

Suboccipital craniectomy for the posterior fossa tumor was performed three days after admission, disclosing a tumor invading the vermis and cerebellar peduncle and spreading over the 4th ventricle and foramen magnum. The pathologic findings confirmed that it was a medulloblastoma. Exploration of the spine at the T10 level showed blood clots and discrete granular tissue on the surface of the cord. A lobulated mass protruded from the cord into the thoracolumbar subarachnoid space. Metastatic medulloblastoma was revealed by histopathologic examination.

Over the following months, this girl received radiotherapy and chemotherapy. Serial follow-up CT and MRI images showed shrinkage of the upper spinal cord lesion but progression of hydrocephalus. She died of pancytopenia and pneumonia six months after surgery.

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DISCUSSION

Common presenting features of medulloblastoma are abnormal gait, truncal ataxia and signs of increasing intracranial pressure, including headache, vomiting and papilledema. Medulloblastoma with initial presentation of spinal cord compression is rare. Only 0.01% of primary intracranial tumors had an initial clinical presentation of spinal cord or nerve root compression [1]. Of these, 61% were medulloblastomas, 16% were glioblastoma multiforme and 11% were anaplastic astrocytoma.

Metastasis of medulloblastoma can be found in both neuraxial and extraneural locations. Supratentorial metastasis is found in 14.6% of cases, intraspinal metastasis in 12.5% and systemic metastasis in 9.7% [2]. Medulloblastomas spread by local invasion, hematogenous dissemination, seeding along cerebrospinal fluid pathways, and, less likely, by lymphatic dissemination [3].

Intraspinal spread from primary intracranial tumors by seeding along cerebrospinal fluid (CSF) pathways is well recognized at autopsy. In the lumbosacral region, nodularity and irregularity of the thecal sac is common, as is diffuse thickening and adhesions of the nerve roots and irregular obliteration of the subarachnoid space. The high incidence of involvement of the lumbosacral region shows the effect of gravity on CSF-borne metastasis. Disseminated plaques and nodules are also more common in the dorsal region, supporting the theory of spread along CSF pathways, as the normal caudal flow of CSF is along the dorsal aspect of the cord [4]. As the thoracolumbar mass in our case was on the dorsal aspect of the spinal cord in our patient, dropped metastasis is suspected.

While metastasis of medulloblastoma throughout the spinal subarachnoid space is a well-known phenomenon, it is rare for medulloblastoma to have intramedullary spinal cord metastasis [3-6]. The pathway of spread is still unclear. Edelson et al. [5] reported that intramedullary metastasis may extend into the spinal cord from a deposit in the subarachnoid space. To our knowledge, there were two cases of intramedullary metastatic medulloblastomas before 1986 [3, 6] and the authors considered the possibility of tumor seeding via the central canal. One of them reported that the opening of the central canal due to hydrocephalus may allow seeding into the cord [6].

Spontaneous hemorrhage associated with primary or recurrent medulloblastomas unrelated to the operative procedure is rare. Fifteen patients with these tumors combined with spinal hemorrhage were reported [2]. The most common causes of bleeding in intraspinal tumors are ependymoma (64%), followed by neurinomas, astrocytomas, and other tumors. Djindjian et al. [7] reviewed the causes of spinal hemorrhage due to tumors in 50 patients and found no medulloblastomas. Therefore, due to the intratumoral hemorrhage in our case, the preoperative impression strongly suggested that this was a 4th ventricular ependymoma with intradural metastasis.

It is possible that some element of extension or invasion of the spinal cord occurred along vascular channels, as evidenced by the frequent and extensive perivascular cuffing by tumor cells seen along the deep intraparenchymal blood vessels of the spinal cord. One could postulate that blood vessel destruction and causes hemorrhage.
into the metastasis [3]. In order to have a better staging, we propose that once a posterior fossa tumor with cistern involvement is diagnosed, the patient should undergo a spinal survey.

REFERENCES

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臨床以急性癱瘓表現之小腦神經管胚細胞瘤
合併脊椎轉移：病例報告

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本篇報告一例罹患神經管胚細胞瘤的四歲女孩。其特殊處在於發病時以便祕，尿液滯留
及漸行性下肢無力等臨床症狀表現，並在懷疑脊髓受壓迫的情況下接受脊椎磁振造影的
檢查。結果發現脊椎內有腫瘤，同時也意外發現存在小腦的另一個病灶。病理切片證實此
病例為一小腦之神經管胚細胞瘤伴隨脊椎內的轉移。脊椎磁振造影時亦同時發現脊椎內的
病灶有出血現象，極為罕見。

關鍵詞：神經管胚細胞瘤，脊椎，轉移，磁振造影，出血