Medulloblastoma in an elderly patient: A case report and literature review

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Abstract. Medulloblastoma is the most common central nervous system malignancy in children; however, it is significantly less common among adults, particularly elderly individuals. We herein report an unusual case of a 72-year-old woman who presented with progressive dizziness. Magnetic resonance imaging revealed a heterogeneous solid mass located near the right tentorium of the cerebellum. The patient underwent surgical removal of the tumor and microscopic examination revealed histological and immunohistochemical characteristics consistent with a diagnosis of medulloblastoma. To the best of our knowledge, this is the first complete report on the oldest medulloblastoma patient in China.

Introduction

Medulloblastoma is the most common central nervous system (CNS) malignancy in children; however, it is extremely uncommon among adults, comprising only 0.4-1% of CNS neoplasms in adults. Medulloblastoma is located in the cerebellar hemispheres and exhibits a male predominance in the adult population. The majority of affected adults (~63%) are aged 20-40 years, whereas occurrence in individuals aged >50 years is exceedingly rare (1). Several adult medulloblastoma cases have already been reported in China. Lin and Gao (2) have recorded the oldest patient to be aged 75 years, but lack clinical information regarding complaints, physical signs, imaging and pathology. To the best of our knowledge, the case presented herein appears to be the first complete report on the oldest medulloblastoma case in China. The aim of this study was to analyze the clinical characteristics, imaging findings, operative method and pathology, and provide specific information on adult medulloblastoma for clinicians.

Key words: medulloblastoma, elderly, cerebellum

Case report

A 72-year-old woman sought medical attention due to progressive problems with dizziness over 1 year, particularly when standing or walking, exhibiting a drunken gait. The severity of the dizziness increased during the month preceding admission and it was always present during daily activities. The patient reported nausea, vomiting and diaphoresis, gradually relieved after resting. The past medical history included hyperlipidemia. The physical examination revealed level nystagmus, positive Romberg's sign, and poor sense of vibration in the left arm compared with the right when testing with a tuning fork. The remaining cranial nerve, motor, and sensory examinations were normal. The preoperative magnetic resonance imaging (MRI) is shown in Fig. 1.

Surgical treatment and outcome. The operation was performed using the suboccipital midline approach, which included an inverted 'L'-shaped incision in the right occipital bone, removing a bone flap sized ~3x3 cm, from the midline to the transverse sinus.

During the operation, the lesion was found to be located in the inferior border of the tentorium of the cerebellum and upper surface of the right cerebellar hemisphere (Fig. 2). The boundary between the tentorium of the cerebellum and cerebellar tissue was clear. The lesion was sized ~2.4x2.0x2.0 cm, had a soft texture, gray color, and a rich blood supply.

The computed tomography (CT) scans before and after the operation are shown in Figs. 3 and 4, respectively.

Histological examination. The results of the pathological examination confirmed the tumor to be a medulloblastoma. The tumor cells were the classic type, and the structure was nodular. The cell types of medulloblastoma include 5 types: desmoplastic/nodular medulloblastoma; medulloblastoma with extensive nodularity; anaplastic medulloblastoma and large cell medulloblastoma (3).

The immunohistochemistry results (Figs. 5 and 6) were as follows: Cytokeratin (CK) (-), Neuronal Nuclei (+), oligodendrocyte transcription factor 2 (-), nestin (-), thyroid transcription factor-1 (-), neurofilament protein (NF) (-), B-cell lymphoma (Bcl)-6 protein (-), CD20 (-), CD3 (-), CD34 (-), CD56 (+), CD99 (-), CKAE1/3 (-), chromogranin A (+/-), cyclin D1 (-), glial fibrillary acidic protein (-/+), Ki-67 (+; >30%),

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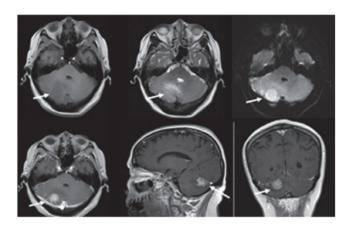


Figure 1. Magnetic resonance imaging (MRI) of the brain performed preoperatively revealed a lesion of the right cerebellum (top left, middle and righ panels: T1 long signal, T2 long signal and diffusion-weighted image high signal, respectively; arrows). MRI with contrast enhancement (bottom) revealed a heterogeneous solid mass (arrows) measuring ~2.4 cm in the anteroposterior diameter, with surrounding edema (left, middle and right panels: transverse, median and coronal planes, respectively).

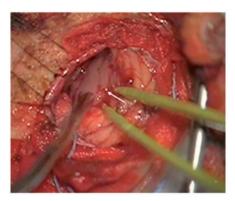


Figure 2. Intraoperative image: The tumor tissue is gray, whereas normal tissue appears yellowish.

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of a right cerebellar lesion.

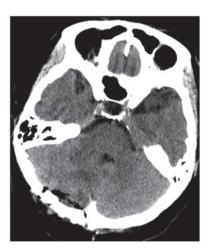


Figure 4. Computed tomography scan after the operation, showing no hemorrhage or effusion.

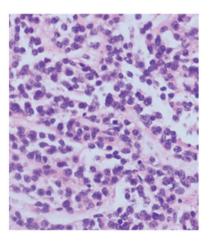


Figure 5. Pathological characteristics of medulloblastoma (hematoxylin and eosin staining; magnification, x400). The tumor cells were the classic type and the structure was nodular. On microscopic analysis, small basophilic tumor cell exhibited diffuse growth, and were arranged in trabeculae, sheets and nests. The cells had round or oval nuclei with deeply stained chromatin, high nuclear:cytoplasmic ratio, with mitotic figures and partial necrosis.

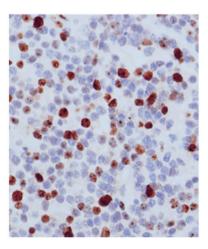


Figure 6. Medulloblastoma Ki-67 staining (~30%); magnification, x400.

multiple myeloma oncogene 1 (-), neuron-specific enolase (+), S-100 (-), synaptophysin (+), Bcl-2 (+), vimentin (-) and reticular fiber antibody (-). Based on the microscopic findings, the medulloblastoma was classified as grade IV according to the WHO classification (3).

Figure 3. Computed tomography scan before the operation showed no signs

Therapeutic method. The patient's condition significantly improved after the operation. The patient was treated with

radiotherapy of the posterior cranial fossa, the whole brain and the spinal cord (the whole-brain and spinal cord dose was 26-36 Gy, whereas the dose to the posterior cranial fossa without the brain stem was 50-60 Gy. The patient is currently followed up by mail or telephonical communication.

Discussion

Medulloblastoma is a CNS malignancy originating from the neuroepidermal layer of the cerebellum, first presented by Cushing and Bailey in 1925. This malignancy is the most common CNS tumor in children, and is commonly located in the fourth ventricle, cerebellum and vermis. However, only 9 cases of medulloblastoma in adults (aged >65 years) have been reported outside China (4), with an onset between 65 and 88 years (mean age, 77.8 years); 7 cases (77.8%) occurred in the cerebellar hemisphere and 2 cases (22.2%) in the cerebellar vermis. One case (11.1%) was located in the fourth ventricle, whereas the cerebellar hemisphere was the most common location in elderly patients. Our findings were similar to those of previous cases regarding the onset and location of adult medulloblastoma. Koeller and Rushing (1) described undifferentiated cells in the posterior medullary vellum that are close to the midline early in life, but exhbit lateral and superior lateral migration with aging. Gupta et al (5) reported that adult medulloblastoma originates in a primordium of the granule cell layer of the cerebellum, which is located on the surface of submeningian molecular layer, and its cells disappear gradually within 18 months after birth. This theory of biological origin may explain the predilection site at the dorsal surface of the cerebellar hemisphere in adults. Thus, medulloblastoma should be taken into account in parenchymatous tumors of the cerebellar hemispheres.

The main clinical manifestations of medulloblastoma are increased intracranial pressure and cerebellar ataxia. If the nuclei of the brain stem become damaged, the patients present with gait disturbance, diplopia and anesthesia. If the tumor involves the fourth ventricle, it may cause obstructive hydrocephalus. The clinical symptoms were not severe in our patient, which was related to the location of the tumor.

The imaging characteristics of adult cerebellar medulloblastoma are closely associated with the tentorium of the cerebellum or meninges. The CT images showed isodensity or marginally high density, on a background of edema, with a clear boundary. MRI imaging revealed mildly long T1 and T2, with a clear boundary, which may be associated with the cellular composition of the tumor (6). However, through careful observation, particularly after enhancement, all levels and various angles of observation revealed a difficulty in differentiating between the tumor and the cerebellar parenchyma (7). Due to the lack of knowledge of non-cerebellar vermis medulloblastoma, the misdiagnosis rate is high. Our patient was initially misdiagnosed with meningioma, vascular tumor and metastasis.

The origin, pathological subtype, clinical treatment and prognosis of adult medulloblastoma patients differ compared with those in children; in adults, the postoperative 5-year survival rate was ~64.9-81.0% and the postoperative 10-year survival rate ~52.0-62.0%, with a median survival time of 8.1-17.7 years (8). Some studies report that older medulloblastoma patients may have a better prognosis (9). With the aging of the general population, there may be more similar cases reported in the future.

Acknowledgements

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