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Case Report

CEREBELLAR MEDULLOBLASTOMA IN AN ELDERLY MAN: AN UNEXPECTED FINDING

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SUMMARY – Medulloblastoma is one of the most common primary tumors of the central nervous system in children and quite uncommon in adult age. Clinically, medulloblastomas may be difficult to recognize in the elderly because of their rarity and histologic similarity to common metastatic tumors. Medulloblastomas arising earlier in life occur close to the midline, whereas those arising later in life are located more laterally within a cerebellar hemisphere. This case report presents an unusual appearance of medulloblastoma in an elderly man.

Key words: *Cerebellar neoplasms – diagnosis; Medulloblastoma – diagnosis; Medulloblastoma – therapy; Medulloblastoma – surgery; Adult*

Introduction

Medulloblastoma is usually considered to be a tumor of childhood, accounting for approximately 15% of all pediatric brain tumors[1,2], and according to the Connecticut Tumor Registry, for only 0.4% of all brain tumors in adults[1]. A significant number of cases have been reported in elderly patients[3]. Although medulloblastomas in adults are not rare, the onset after age 50 is exceptional[4]. Three large series have revealed that less than 25% of these tumors occurred in older adolescents and adults[5,6]. Recently, Giordana *et al.* have reported on adult patients to account for 34% of all medulloblastoma cases in their epidemiologic study[7]. According to literature reports, medulloblastoma shows a 1.5- to 2-fold male predominance[5,8,9].

In this case report, we present a 62-year-old man with a tumorous lesion in the superior cerebellar and pineal region compressing the aqueduct of Sylvius and producing subsequent hydrocephalus. A metastatic tumor or pinealoblastoma was suspected before the surgery.

Case Report

In February 2000, a 62-year-old man with medical history of cardiac arrhythmias and arterial hypertension experienced headache, occasionally followed by nausea and vomiting. The symptoms progressed with the development of gait disturbance and urgency incontinence. In July 2000, he was admitted to local hospital for sudden aphasia, right-sided hemiparesis, somnolence and papilledema. Noncontrast computed tomography (CT) scans revealed obstructive hydrocephalus, and a ventriculoperitoneal shunt was implanted. Postoperatively, magnetic resonance imaging (MRI) of the brain revealed a hypointense lesion in the superior cerebellar region expanding towards and consuming the pineal region. The lesion was hypointense on T1-weighted images and moderately hyperintense on T2-weighted images (Figs. 1, 2). The patient was admitted to our Department in August 2000. Neurologic examination performed on admission showed an oriented man with unsteady gait and vertigo. Coordination tests (finger to nose) showed right-sided dysmetria. Adiadochokinesia was also present. The rest of the neurologic examination was normal.

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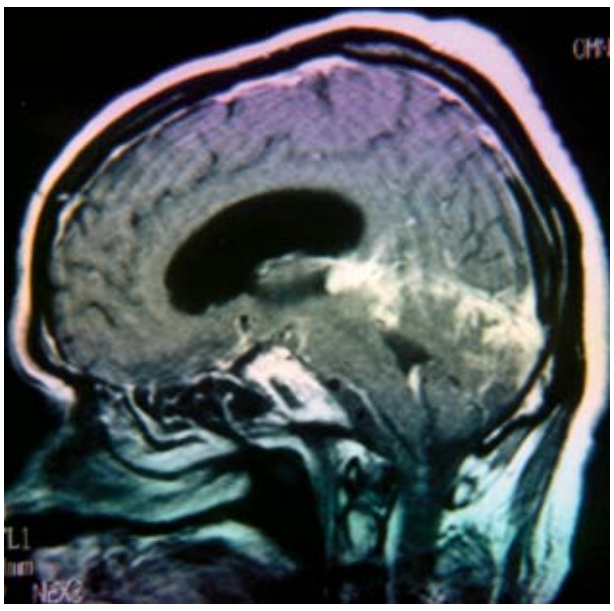

Fig. 1. Midsagittal postcontrast T1WI magnetic resonance image showing a large lesion expanding from cerebellar vermis towards pineal region.

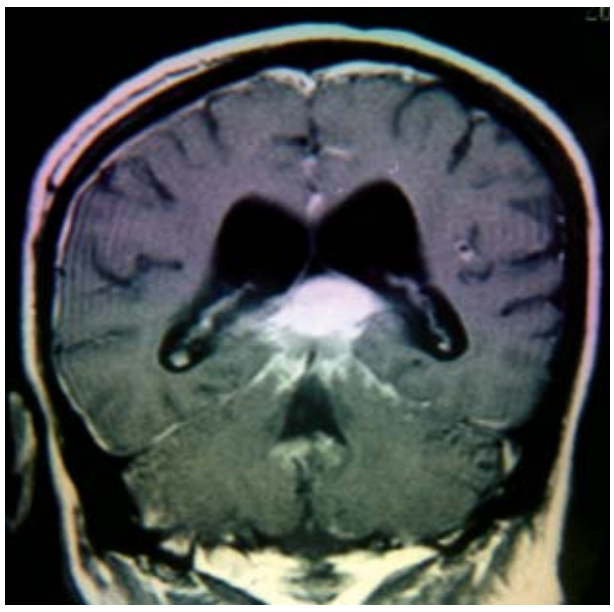

Fig. 2. Coronal postcontrast T1WI magnetic resonance image shows a midline, hyperintense lesion indicative of primitive neuroectodermal tumor (PNET) or pinealoblastoma.

The operation

An infratentorial supracerebellar approach was chosen upon preoperative MRI. The dura was opened by an Y-shaped incision. An operating microscope was brought into the field. The retractor was placed in the midline over the cerebellum, and cerebellar draining veins were identified, coagulated and divided. Deep veins and a solid, purplish, friable tumor were enveloped in a thickened arachnoid membrane. The arachnoid was opened. A portion of the tumor was removed and referred for neuropathologic frozen section diagnosis. After maximal reduction had been accomplished, hemostasis was completed. The dura was closed in a watertight fashion and there was no leak of cerebrospinal fluid.

At 48 h postoperatively, the patient experienced worsening of truncal ataxia and cerebellar mutism. Therefore, urgent CT scan was performed to show a minimal residual tumor and cerebellar edema. The patient received antiedematous therapy and his condition ameliorated. He was discharged from the Department on day 16 postoperatively, with persistent mild truncal ataxia and dysarthria, and started oncologic treatment. Two weeks after discharge from the Department, his condition deteriorated and he died suddenly from pulmonary embolism.

Pathohistologic examination

Histologically, the tumor was composed of small, undifferentiated, hyperchromatic cells with scanty cytoplasm. Cells were arranged as rosettes separated by prominent fibrous tissue (Figs. 3, 4).

Immunohistochemically, tumor cells were positive for synaptophysin and S-100 protein, showing neuronal differentiation of medulloblastoma, and negative for GFAP, vimentin, chromogranin-A, cytokeratin and HMB-45. Histochemical reactions and immunohistochemical analyses confirmed the diagnosis of desmoplastic medulloblastoma.

Discussion

Medulloblastoma is an uncommon tumor of the posterior fossa mainly found in children. Adult patients accounted for 15% to 36% of patients in some large series[10]. It may occur at any age, from neonates to the elderly, but is primarily a pediatric tumor[4]. More than 80% of adult medulloblastoma patients are diagnosed before age 40[11]. Medulloblastomas in adults differ from those in children in three ways: in adults, the tumor is more frequently located in the cerebellar hemispheres or cerebellopontine angles. Brain stem involvement is less common, and very

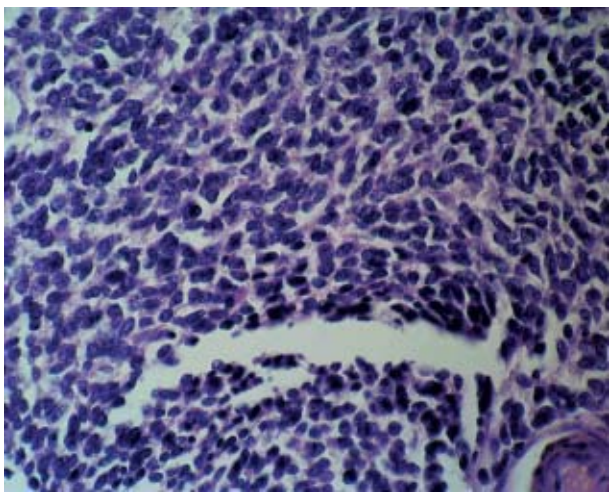

Fig. 3. Small, polymorphous cells of medulloblastoma forming few rosettes (HE, original magnification X200).

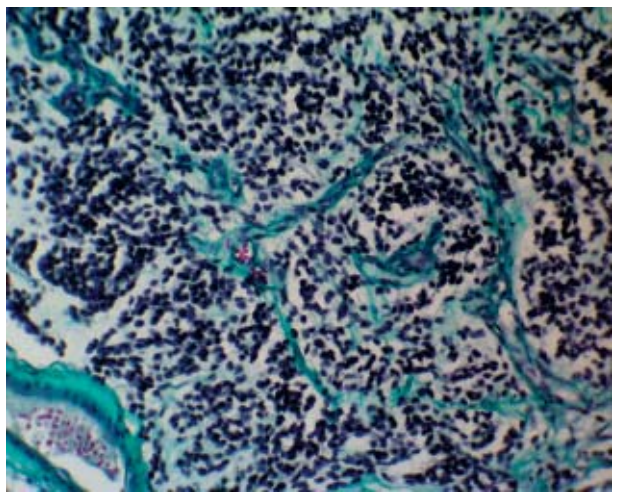

Fig. 4. Desmoplastic medulloblastoma. Prominent stromal fibrous tissue through the tumor (Mallory, original magnification X200).

late recurrences, e.g., after ten years, are recorded occasionally, virtually never in children. Medulloblastoma is thought to arise from primitive or pluripotential cells found in the germinative zone of the posterior medullary velum. These cells migrate upwards and laterally to form the external granular layer of the cerebellar hemispheres. If medulloblastoma arises from the external granular layer, neuronal differentiation might be expected in these tumors, which is observed in the majority of medulloblastomas. The prognostic significance of cellular differentiation, whether neuronal or glial, remains controversial[12].

Medulloblastoma can develop anywhere along the route of migration, medially or laterally. According to this hypothesis, the tumors that arise earlier in life occur closer to the midline, whereas those arising later in life are located more laterally within a cerebellar hemisphere[1,13,14].

We described a 62-year-old male patient with medulloblastoma of the superior vermis extending into the pineal region. We consider it as an exceptional occurrence of medulloblastoma, not only concerning the patient's age but also the extremely rare localization of the tumor, which is in discordance with the above hypothesis. In this case, pathohistologic examination revealed a desmoplastic variant of medulloblastoma, which may be more common in adults than in children[11]. Tumors that show a desmoplastic pattern on reticulum staining have been described as having a better prognosis than those with 'classic' histology, although other researchers found no difference in survival between the patients with and without desmoplastic features[15]. The prognostic value of tumor cell proliferation is limited. Recently, several investigators have evaluated loss of heterozigosity (LOH) on 17p chromosome as a prognostic indicator[16]. A correlation between LOH on 17p chromosome and poor therapeutic response and shortened survival time was proven[17,18].

Most studies show that total resection (rather than subtotal resection or biopsy) has a favorable impact on survival, however, without adjuvant therapy the tumor related mortality rate is almost 100%. Adults with medulloblastoma show 5-year survival rates of 46% to 78%[6,19]. Postoperative radiation therapy is always required and chemotherapy is often recommended, although its role is less certain.

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Sažetak

CEREBELARNI MEDULOBLASTOM U MUŠKARCA STARIJE ŽIVOTNE DOBI: NEOČEKIVAN NALAZ

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Meduloblastom je jedan od najčešćih primarnih tumora središnjeg živčanog sustava u dječjoj dobi, a rijedak u odraslih osoba. Meduloblastome koji se pojavljuju u odrasloj dobi teško je klinički prepoznati zbog njihove rijetke pojavnosti i histološke sličnosti s metastatskim tumorima. Meduloblastomi koji se pojavljuju u dječjoj dobi smješteni su bliže medijalnoj liniji, dok se oni koji se pojave u starijoj životnoj dobi nalaze lateralno unutar hemisfera maloga mozga. Prikazani slučaj predstavlja iznimno rijetku pojavu meduloblastoma u muškarca starije životne dobi, koji je zahvatio gornju površinu maloga mozga i pinealno područje. Većina istraživanja ukazuje na to da radikalan kirurški zahvat i poslijeoperacijsko zračenje imaju najbolji učinak na preživljenje.

Ključne riječi: *Cerebelarne neoplazme – dijagnostika; Meduloblastom – dijagnostika; Meduloblastom – terapija; Meduloblastom – kirurgija; Odrasla osoba*