

# An Adult Case of Medullomyoblastoma Extending to the Supratentorial Area

Fatih BAYRAKLI \*, Selçuk PEKER \*\*, Özlem YAPICIER \*\*\*, Necmettin PAMİR \*\*

Marmara Üniversitesi Tıp Fakültesi, Nöroşirürji Bilim Dalı\*, Acıbadem Hastanesi, Nöroşirürji Bölümü\*\*, Marmara Üniversitesi Tıp Fakültesi, Patoloji Bilim Dalı\*\*\*, İstanbul

✓ A rare case of adult medullomyoblastoma with unique location presented. The patient was admitted to our clinic with the history of progressive balance disorder, nausea and vomiting. Disequilibrium had started 2 months earlier. The patient underwent a craniotomy and histopathologic examination revealed a biphasic tumor. Few definite strap cells with striation were identified in the tumor. The diagnosis was medullomyoblastoma. Histopathological evaluation is crucial for diagnosis of MMB. Tumor resection is the first step of treatment.

**Key words:** Medullomyoblastoma, posterior fossa tumors, primitive neuroectodermal tumors, cranial surgery

## Erişkinde Supratentoryel Uzanım Gösteren Bir Medullomyoblastoma Olgusu

✓ Makalemizde nadir rastlanan erişkin hastada medullomyoblastom olgusu sunulmuştur. Hasta, kliniğimize iki ay önce başlayan ilerleyici denge bozukluğu, bulantı ve kusma şikayetleri ile kabul edilmiştir. Hasta opera edilmiş ve histopatolojik tetkik sonucu bifazik tümör sonucunu ortaya koymuştur. Tümör içerisinde çizgili kayış şeklinde hücreler görülmüştür. Tanı medullomyoblastomadır. Medulloblastoma tanısı için histopatolojik değerlendirme önemlidir. Tümörün cerrahi olarak çıkarılması tedavinin ilk basamağıdır.

**Anahtar kelimeler:** Medullomyoblastoma, posterior fossa tümörleri, primitive neuroectodermal tümörleri, cranial cerrahi

Medullomyoblastoma (MMB) is a rare childhood brain tumor first described by Marinesco and Goldstein in 1933. MMB invariably occurs in the cerebellum and often involves the cerebellar vermis. It typically affects children under 10 years of age. The majority of patients present with symptoms of increased intracranial pressure (5). Tumor is classified as WHO grade IV and is defined histopathologically as a combination of primitive neuroectodermal and myoblastic elements (7).

In the current case, we report the fifth adult case of MMB in literature; this is also the first case showing supratentorial extension.

## CASE REPORT

A 31-year-old woman presented to our clinic

with progressive balance disorder, nausea and vomiting. The disequilibrium had started 2 months earlier. History of patient revealed that she had undergone three midline suboccipital operations and craniospinal irradiation for posterior fossa tumor which their pathological examination results reported as medulloblastoma in another health center. Initial presentation was two years ago. Operation notes did not reported any tentorial involvement. Preoperative T1 weighted cranial magnetic resonance imaging (MRI) studies showed a heterogeneously contrast-enhancing mass in the right cerebellar hemisphere, extending through the supratentorial area via the tentorium. A cystic component of the tumor can also be seen in the posterior fossa (Figure 1a). Using old incision midline approach performed and gross total resection of the lesion was achieved. Early-postoperative mag-

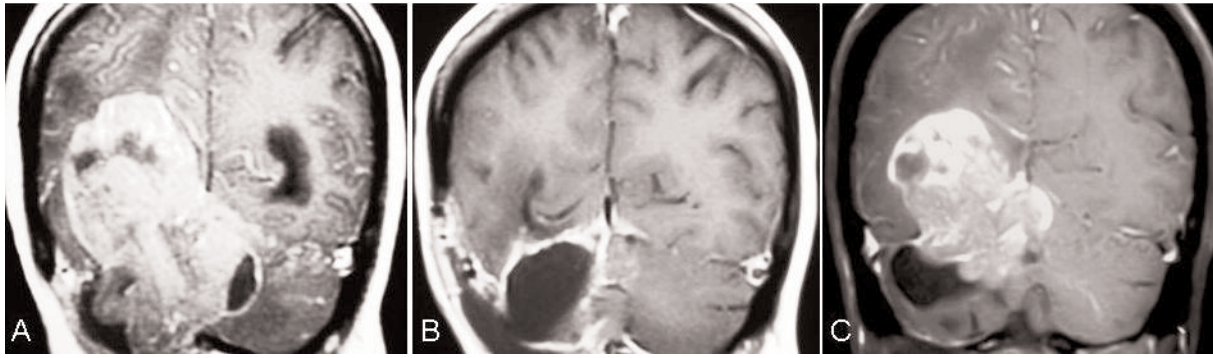


Figure 1. (a) A T1-weighted coronal magnetic resonance image shows the tumor enhanced with contrast; (b) Early postoperative imaging demonstrates gross total removal; (c) Repeat imaging at 2 months post-surgery revealed aggressive recurrence.

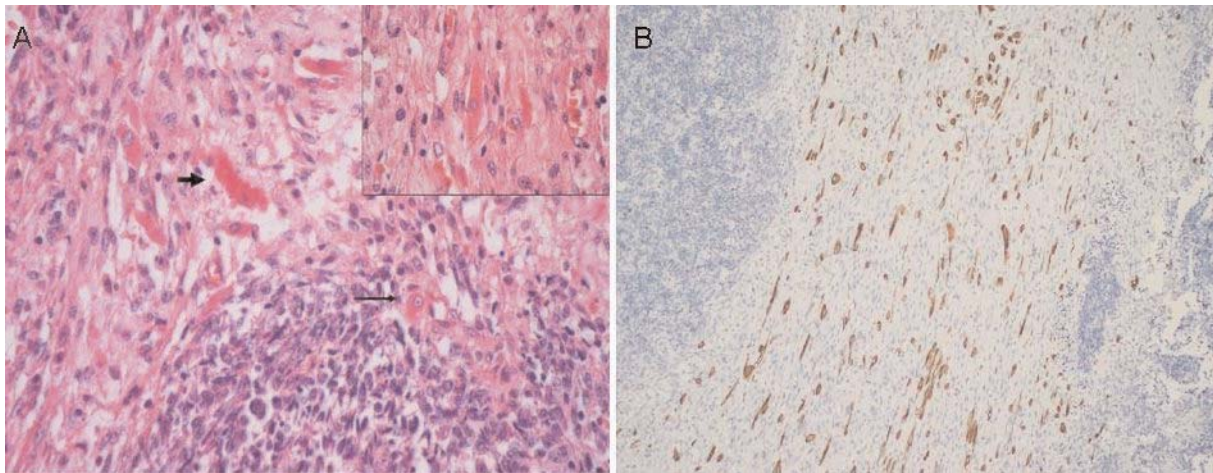


Figure 2. (a) The surgical specimen showed the biphasic pattern typical of medullomyoblastoma, with myogenic cells and primitive neuroectodermal cells. The myogenic cells are either in blast form (long arrow) or in strap cell (short arrow) configuration (H&E, x400). The inset shows well-formed cross-striations in a myogenic cell (H&E, x1000); (b) Myogenic cells expressing desmin (Immunostaining for desmin, x100).

netic resonance imaging indicated complete excision (Figure 1b), but 2 months later, control cranial MRI revealed a recurrent tumor showing similar radiological features with preoperative lesion (Figure 1c). Patient rejected postoperative chemotherapy treatment.

Histopathologic examination revealed a biphasic tumor composed of solid sheets of small undifferentiated cells associated with bundles of spindled cells containing abundant eosinophilic cytoplasm. The tumor consisted mostly of small round cells with minimal cytoplasm, hyperchromatic and anaplastic nuclei. Tumor cells also showed elongated and carrot-shaped nuclei associated with occasional Homer-Wright rosettes. Frequent mitotic figures were seen in the tu-

mor. Some tumor cells revealed relatively abundant eosinophilic cytoplasm and eccentrically located nuclei that resembled rhabdomyoblastic differentiation. Few definite strap cells with striation were identified in the tumor. Immunohistochemical study was positive for desmin, synaptophysin, and chromogranin. The histopathological diagnosis was MMB (Figure 2).

## DISCUSSION

MMBs arise exclusively within the infratentorial compartment. Previous literature reports had documented cases involving the cerebellar vermis/4th ventricle (1-9,11), including 3 tumors that invaded the brainstem (4,8) and 2 tumors with extension to the cerebellopontine angle (8, 10); 5 tu-

mors that were limited to the cerebellar hemispheres<sup>(5)</sup>; 3 tumors involving both the hemisphere and the vermis, including 1 tumor with brainstem invasion<sup>(5)</sup>. In our case, the tumor showed cerebellar hemispheric involvement with supra tentorial extension that make this case unique for involvement in a location other than posterior fossa in reported MMB cases in the literature.

Of the patients previously described in the literature, approximately 90 % were under 10 years of age, and only 4 were adults<sup>(5,8,9,11)</sup>. Our case represents the fifth adult MMB case in literature.

There are a few reports in literature that neuro-radiologically evaluated by MRI<sup>(2,3,5,7,9,10)</sup>. Described lesions showed different radiological views such as isointense to cortex on T1W and hyperintense on T2W, multiloculated cystic vermian tumor and exhibited variable enhancement and areas of necrosis<sup>(2,3,5,7,9,10)</sup>. Our case showed heterogenous contrast enhancement with cystic component in posterior fossa and extension to supratentorial area.

Diagnosis of MMBs mainly depends on histopathological evaluation. Presented cases in literature have been biphasic, containing primitive neuroectodermal and rhabdomyoblastic components by definition<sup>(5)</sup>. Various other cellular components or lines of differentiation have been described in MMB, including cells that exhibit neuronal/ganglionic differentiation, or glial/astrocytic differentiation, heterologous elements and, pigmented/melanotic cells<sup>(5)</sup>. Occasional case reports mention discrete islands of rhabdomyoblastic components<sup>(5,8)</sup>. There were also reports that describing the intermixing of these two cellular components as in our case<sup>(2,5)</sup>.

Treatment strategy for patients with MMB consists of surgical resection followed by adjuvant chemotherapy and radiotherapy. Jaiswal et al.<sup>(6)</sup> reported a case of MMB with long survival who after gross total tumor resection received craniospinal radiation and combination chemotherapy. During his 11 year follow up period he was

asymptomatic. Helton et al.<sup>(5)</sup> reported a series of MMB including 6 cases. They applied craniospinal radiation and chemotherapy as initial treatment in 5 patients. Two of them were tumor free but the other 3 patients showed local recurrence and leptomeningeal disease. One patient who received only craniospinal radiation for initial treatment, took chemotherapy after local recurrence was tumor free. Our patient had undergone surgical operation four times and craniospinal irradiation but after the fourth operation the clinical course of tumor was aggressive.

In conclusion, neuroradiological and, histopathological features of an adult female with MMB showing supratentorial extension are described and discussed herein. Histopathological evaluation is crucial for diagnosis of MMB. Tumor resection is the first step of treatment. To draw a certain conclusion about adjuvant therapy's effectiveness on MMB, larger series with longer follow up periods are needed.

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