Midline Cerebellar Medulloblastoma in a Seventy-One-Year-Old Patient

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ABSTRACT: Background: Medulloblastoma is the most common malignant central nervous system tumour in children but, in contrast, quite rare in adults. Hemispheric, rather than midline, cerebellar medulloblastomas are more common in older children and adults. We present the unusual case of a 71-year-old man who presented with a fourth ventricular mass that proved to be a medulloblastoma. Methods: A 71-year-old man presented with progressive balance problems, slurred speech and double vision. A CT scan of the brain revealed a hyperattenuating, partially calcified, avidly enhancing mass within the fourth ventricle. Diffusion weighted MRI showed restricted diffusion within the mass. The patient underwent a midline suboccipital craniotomy and a subtotal resection was achieved. Results: Histological examination showed a densely cellular neoplasm composed of small cells with a tendency towards neuroblastic rosette formation. Most cells were strongly positive for neuron-specific enolase and synaptophysin. Ultrastructurally, tumour cells showed evidence of neuronal differentiation. These findings were consistent with a classical medulloblastoma. Conclusion: Adult medulloblastoma should be considered in the differential diagnosis of a partially calcified hyperattenuating mass within the fourth ventricle.
The patient described nausea and vomiting, but denied headache, urinary incontinence, or sensory symptoms. His past medical history was significant for hypertension and polycythemia rubra vera, which had been treated successfully with phlebotomy and hydroxyurea. Physical examination revealed bilateral sixth nerve palsies, dysarthria, dysmetria greater in the right arm than the left, and gait ataxia. The remaining cranial nerve, motor, and sensory examinations were normal.

A CT scan of the brain revealed a hyperattenuating, partially calcified, avidly enhancing mass within the fourth ventricle (Figure 1). An MRI scan of the brain demonstrated a mass within the fourth ventricle with heterogeneous signal characteristics (Figure 2A). Diffusion-weighted imaging showed restricted diffusion within the mass (Figure 2B). The differential diagnosis included ependymoma, atypical meningioma, choroid plexus papilloma, and metastasis. A metastatic screen including a chest radiograph, abdominal ultrasound, and liver function tests was negative.

The patient underwent a midline suboccipital craniotomy with ultrasound guidance for resection of the tumour. The tumour was situated within the fourth ventricle, had invaded the floor, and was purple-grey, soft in consistency, and highly vascular. Only a subtotal resection could be achieved due to the tumour’s vascularity, which hindered adequate visualization of the tumour margins. Histological examination showed a densely cellular neoplasm composed of small cells with a high nuclear-to-cytoplasmic ratio, hyperchromatic nuclei and a tendency towards neuroblastic rosette formation (Figure 3A). Most cells were strongly positive for neuron-specific enolase (Fig. 3B) and synaptophysin (Figure 3C). Ultrastructurally, tumour cells were poorly differentiated (Figure 4A) and showed evidence of neuronal differentiation in the form of aggregates of microtubules in the cytoplasm and cell processes (Figure 4B). These findings were consistent with a classical medulloblastoma.

Postoperatively, the patient’s neurological examination was not significantly changed. An MRI of the entire spine was performed for staging purposes and was negative. He went on to receive 3600 cGy of external beam radiation therapy to the craniospinal axis in 20 fractions with a boost to the posterior fossa of 1800 cGy in 10 fractions. Initially, treatment was tolerated well with only some mild nausea and vomiting, but the patient went on to develop subacute radiation encephalopathy eight weeks after the completion of radiation therapy, presenting with expressive dysphasia and worsened cerebellar symptoms. A follow-up MRI at this time revealed that the residual portion of the tumour within the right vermis had decreased moderately in size. The appearance of the tumour was stable at 24 weeks after the completion of radiation therapy and the patients symptoms had improved.

**DISCUSSION**

Medulloblastoma is rare in patients older than 50 years of age. They are thought to be derived from remnants of the external granular layer of the cerebellum, cells of which may have a predisposition to neoplastic transformation early in life.4
The oldest patient on record with clinical information was 73 at the time of diagnosis and had a cerebellar hemispheric medulloblastoma. To our knowledge, we report the oldest patient with a medulloblastoma of the fourth ventricle. Mechanisms of pathogenesis late in life are currently not known.

The signature imaging feature of medulloblastoma on non-contrast CT is hyperattenuation, reflecting the dense cellularity of these tumours on histological analysis. The dense cellularity of these tumours also results in restricted diffusion of free water, as illustrated via diffusion-weighted MRI. Treatment should consist of gross total resection if possible, early postoperative MRI for prognostication, and craniospinal radiation. Adjuvant chemotherapy may be useful in patients thought to be at high risk of recurrence, but the available evidence has not clearly demonstrated benefit. Collin’s Law states that the period of risk of recurrence for embryonal tumours is nine months plus the age of diagnosis. According to this rule, our patient would certainly be at risk of recurrence for the remainder of his life. The application of Collin’s Law, however, is only valid if theories of childhood medulloblastoma pathogenesis hold true for older

**Figure 2:** (A) Sagittal FLAIR sequence shows a mass within the fourth ventricle with heterogeneous signal characteristics. (B) Axial diffusion-weighted sequence demonstrates restricted diffusion within the tumour.

**Figure 3:** (A) The tumour is densely cellular and composed of small cells with round or ovoid nuclei and small amounts of cytoplasm arranged in clusters or solid sheets. There is a tendency for the tumor cells to aggregate around eosinophilic fibrillar areas hematoxylin and eogin (H+E). The majority of tumour cells show strongly positive cytoplasmic staining for neuron-specific enolase (B) and synaptophysin (C). A: 200x; B: 200x; C: 400x
patients. In any case, close follow-up with serial MRI for older patients with medulloblastoma is clearly warranted.

We conclude that adult medulloblastoma should be considered in the differential diagnosis of a partially calcified hyperattenuating mass within the fourth ventricle.

REFERENCES


Figure 4: (A) Electron micrograph showing closely packed undifferentiated cells with scant cytoplasm, paucity of organelles, peripherally condensed chromatin and advanced degenerative changes. (B) Dense aggregates of microtubules are present in the cytoplasm of tumour cells. Bars = 1 µm.