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Case Reports

Primary medulla oblongata germinoma – an unusual posterior fossa tumors in young adults

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ABSTRACT

We encountered 2 patients with germinoma arising from the medulla oblongata in whom preoperative radiological diagnosis was difficult. A 30-year-old woman presented due to aspiration pneumonia caused by bilateral lower cranial nerve palsies, and a 24-year-old man presented with headache caused by obstructive hydrocephalus. In both patients, there was a midline tumor that extended from the lower part of the fourth ventricle to the C1 lamina level. It was well-demarcated and homogeneously enhanced tumor with a slightly high density on plain CT scan. The preoperative diagnosis for both patients was ependymoma. The former patient had persistent lower cranial nerve palsies due to brain stem injury after tumor resection. Both patients achieved complete remission with adjuvant therapy. Fewer than 10 cases of germinoma affecting the medulla oblongata have been reported. Radiological findings resembling those of the pineal region germinoma were observed in the two patients reported here. We would like to stress the importance of remembering germinoma when making a preoperative differential diagnosis of fourth ventricular tumors in young adults.

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1. Introduction

We recently encountered two young adults who had tumors of the fourth ventricle. A preoperative diagnosis of ependymoma was made in both cases and resection was performed according to the intraoperative pathological diagnosis. In both patients, the final diagnosis was germinoma and complete remission was achieved by adjuvant therapy. We present here the neuroradiological features of germinoma arising from the medulla oblongata.

2. Case report

2.1. Patient 1

A 30-y-old woman presented with a 2 month history of a gradually worsening swallowing disturbance. On admission, she had a left cranial nerve (CN) VI palsy and a bilateral palsy of CNs IX, X, and XII. Plain CT scans showed a tumor without calcification in the fourth ventricle that had a slightly high density (Fig. 1A). An MRI showed a homogeneous tumor with a slightly low signal intensity on T1-weighted (T1WI) MRI while it had a heterogeneous high signal intensity on T2WI, and strongly compressed the medulla oblongata (Fig. 1B,C). Sagittal images showed the tumor as heterogeneous, weakly enhanced by gadolinium-diethylenetriamine

penta-acetic acid (DTPA), arising in the hypoglossal triangle and extending into the cisterna magna (Fig. 1D). The tumor was pinkish, soft, and not highly vascular (Fig. 1E). Unlike the tumor border, which was well-defined, the border with the medulla oblongata was not. The intraoperative pathological diagnosis was a highly malignant tumor, so we attempted a maximal tumor resection. However, the final pathological diagnosis was germinoma (Supplementary Fig. 1A and Fig. 1B). Chemotherapy and focal irradiation were administered, and complete remission was confirmed by imaging at 1 year postoperatively (Fig. 1F). Her cranial nerve palsies showed some improvement, but there was residual palsy of CN VI and VII.

2.2. Patient 2

A 24-y-old man presented with a 2 week history of headache and nausea, and a tumor in the fourth ventricle was diagnosed. On admission, his headache was resolved and there were no obvious neurological deficits. There was a tumor in the lower part of the fourth ventricle with a slightly high density on plain CT scans (Fig. 2A). An MRI of a clearly demarcated tumor showed isosignal intensity on T1WI and a slightly high signal intensity on T2WI. It showed weak homogeneous contrast enhancement, and extended into the cisterna magna (Fig. 2B–D). The tumor was a soft, reddish brown mass that was prone to bleeding. Although the border of the tumor was well-defined, the border with the medulla oblongata was not (Fig. 2E). Because an intraoperative pathological diagnosis of germinoma was made, 80% of the tumor was extirpated. The

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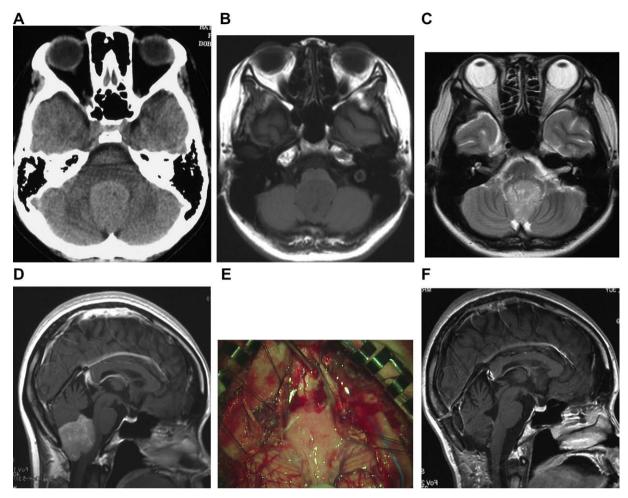


Fig. 1. (A) A precontrast axial CT scan on admission showing a round, slightly high density tumor occupying the fourth ventricle. (B, C) Axial T1-weighted (B) and T2-weighted (C) MRI showing the mass occupying the lower half of the fourth ventricle and compressing the lower part of the brain stem. (D) Sagittal T1-weighted MRI. The tumor contained a small cyst (not marked) and was homogeneously enhanced by gadolinium-diethylenetriamine penta-acetic acid (Gd-DTPA). (E) An intraoperative photograph showing that the pinkish, soft midline tumor split the cerebellar tonsils and extended downward to the C1 lamina level. (F) Sagittal T1-weighted MRI after intravenous injection of Gd-DTPA at 1 year postoperatively, showing complete remission of the tumor.

final pathological diagnosis was germinoma (Supplementary Fig. 2A and Fig. 2B), so chemotherapy and focal irradiation were administered. Complete remission was achieved at 8 months post-operatively, without any symptoms (Fig. 2F).

3. Discussion

Intracranial germinoma is a tumor characterized by its location and by the age of onset.¹ Such characteristics mean that it is difficult to diagnose germinoma accurately when the location or age of onset is atypical. Unlike for other germ cell tumors, there is no available marker to measure, so the imaging findings are vital. There are only 7 previous reports of germinoma originating in the medulla oblongata.^{2–8} Intracranial germinoma develops predominantly in the pineal region in men, but in the neurohypophysis in women.¹ Including our report, medulla oblongata germinoma has been reported in 6 women and 3 men. The mean age of onset of intracranial germinoma is reported to be 16.1 years,¹ but the 9 medulla oblongata germinomas developed slightly later (mean 26.9 years).

Intracranial germinoma is revealed as a high density tumor on plain CT scans.¹ The tumor margins are usually clear on MRI. The signal intensity is almost equivalent to that of the grey matter on T1WI, and is similar to the grey matter, or is a slightly lower inten-

sity on T2WI.¹ Contrast enhancement is often homogeneous and intense on both CT scans and MRI. These imaging characteristics were largely present in the reported cases of medulla oblongata germinoma, including the two cases reported here (Table 1). This tumor is reported to develop above or below the midline from the lower fourth ventricle to C1 level and its progression pattern differs from that of ependymoma, which extends along the brain stem. Also calcification of germinoma has not been reported.

In summary, germinoma should be suspected in young women in their 20s and 30s with a midline tumor that extends from the medulla oblongata into the fourth ventricle and descends to the cisterna magna. The possibility becomes higher when the mass has a slightly higher intensity on plain CT scans, no calcification or peritumoral edema, and homogeneous enhancement is detected.

4. Conclusion

Imaging findings of medulla oblongata germinoma were similar to those for germinoma originating in the pineal region, and imaging may have an important role when deciding the appropriate therapeutic strategy.

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