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Brain Tumors with Ipsilateral Cerebral Hemiatrophy

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Brain tumors with secondary ipsilateral cerebral hemiatrophy are so rare that only seven cases have been reported in the literature. Three new cases are presented and the clinical findings in all 10 cases are reviewed. The diagnostic value of computed tomography (CT) is emphasized. The authors conclude that nodular high-density lesions located deep in the cerebrum and showing slight contrast enhancement on CT may be associated with ipsilateral cerebral hemiatrophy in young male patients with slowly progressive hemiparesis, dementia, and personality change. In such cases, early diagnosis of germinoma and subsequent radiotherapy may prevent unnecessary surgery.

Secondary cerebral hemiatrophy is usually produced by trauma, vascular accident, infection and the like [1, 2]. It is very rare for a brain tumor to cause ipsilateral cerebral hemiatrophy. Thus far only seven such cases have been reported in the literature. We present three additional cases, review the clinical findings in all 10 cases, and discuss the characteristic course of the disease and the diagnostic value of computed tomography (CT). All reports are from Japan.

Case Reports

Case 1

A 13-year-old boy experienced dystonia of right upper and lower extremities 1 year before hospital admission with slowly progressive right hemiparesis, dementia, personality change, and diabetes insipidus. The pneumoencephalogram showed moderate dilatation of the left lateral ventricle and cortical sulci over the left cerebral convexity. Cerebral angiograms showed only minimal midline shift toward the left without evidence of tumor vessel or tumor stain. Precontrast CT demonstrated an ill-defined area of high density in the region of the left basal ganglia and thalamus, which was enhanced after the injection of contrast material (fig. 1A). The third ventricle was displaced to the left and the left lateral ventricle was dilated, suggesting left cerebral hemiatrophy. The possible diagnosis of germinoma was made by cerebrospinal fluid (CSF) cell culture, and whole-brain irradiation was administered (total dose, 50 Gy). After radiation therapy, left cerebral hemiatrophy was still noted but the abnormal high-density lesion was no longer apparent on postcontrast CT (fig. 1B). The patient was markedly improved and showed no signs of recurrence about 4 years after treatment.

Case 2

A 10-year-old boy had a 2 year history of slowly progressive right hemiparesis and mild mental retardation. Physical examination and laboratory studies were normal. A left carotid angiogram showed shift of the anterior cerebral arteries to the left, and the pneumoencephalogram showed marked dilatation of the left lateral ventricle, but neither revealed any evidence of brain tumor. Precontrast CT demonstrated a nodular high-density lesion in the left basal ganglia and hypothalamus (fig. 2A), which showed slight enhancement after injection of contrast material. Dilatation of the left lateral ventricle and left sylvian fissure was noted in addition to displacement of the third ventricle toward the left (figs. 2B and 2C). Exploratory craniotomy was performed 2 months after clinical presentation, and histologic diagnosis of germinoma was made. After radiation therapy (dose, 50 Gy), the abnormal high-density lesion appeared nearly resolved on postcontrast CT. Clinically, the patient's right hemiparesis was slightly improved.

Case 3

A 19-year-old boy had a 3 year history of slowly progressive right hemiparesis with a 1 year history of dysarthria. Intracranial pressure was normal. CT on first admission was negative. Precontrast CT about 15 months later (fig. 3A) demonstrated a nodular area of slightly increased density with a small focal low-density center in the region of the left hypothalamus, which enhanced after injection of contrast material (fig. 3B). The left sylvian fissure and left frontal horn were slightly dilated. The extension of the deep-seated tumor was much more clearly demonstrated by coronal CT (fig. 3C). Histologic diagnosis of germinoma was obtained by CSF cell culture. Whole-brain irradiation (dose, 40 Gy) was delivered. After irradiation therapy, the signs of left cerebral hemiatrophy were still present on postcontrast CT (fig. 3D), but the abnormal high-density lesion had completely disappeared.

Review and Summary of Clinical Findings

The clinical characteristics, tumor site, and histologic findings in 10 cases of brain tumor with ipsilateral cerebral hemiatrophy are summarized in table 1. All patients except one were young men. Disease onset occurred at 7–16 years of age. The average period from onset to clinical admission was about 2 years, indicating the relatively chronic clinical course of these tumors. The major clinical

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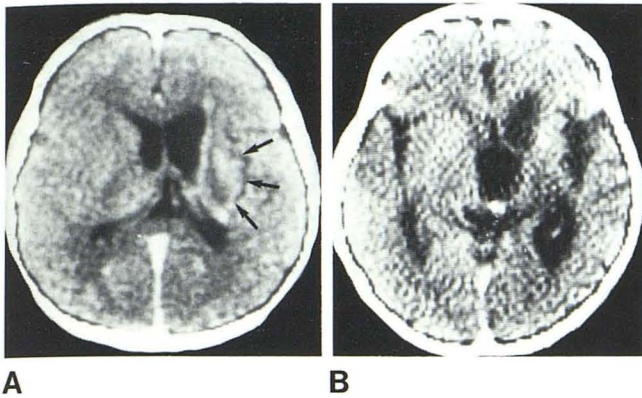


Fig. 1.—Case 1. Germinoma in 13-year-old boy. **A**, Contrast-enhanced CT. Nodular high-density lesion in left basal ganglia and thalamus (arrows). Ipsilateral shift of midline structures and dilated left lateral ventricle. **B**, Contrast-enhanced CT after radiation therapy (total dose, 50 Gy). Signs of left cerebral hemiatrophy still present, but abnormal high-density lesion has almost disappeared.

symptoms were hemiparesis, present in all 10 cases; dementia, present in seven cases; and personality change, present in five cases. No signs of elevated intracranial pressure were seen in any case. Tumors were located in the region of the basal ganglia, thalamus, and/or hypothalamus. Confirmation of the tumor histology was obtained by autopsy in three cases, by surgical operation in five cases, and by CSF cell culture in two cases. Histopathologically all tumors proved to be germinoma except one (an astrocytoma).

Cerebral angiography performed in all cases showed cerebral hemiatrophy but failed to demonstrate the tumor itself. Two of three cases diagnosed by exploratory craniotomy (which was performed because of progressive clinical deterioration and elevated CSF protein) died within 3½ years after onset of disease. The other patient shows no signs of recurrence 11 years after radiation therapy.

The last four cases reported, including the three current cases, could be diagnosed by CT. These patients are still alive after radiation treatment. The characteristic CT pattern in our three cases was the presence of a nodular high-density lesion with slight contrast enhancement, located deep in the cerebral hemisphere and

Fig. 2.—Case 2. Germinoma in 10-year-old boy. **A**, Precontrast CT. Nodular high-density lesion in left basal ganglia and hypothalamus. **B** and **C**, Contrast-enhanced CT. High-density lesion in left caudate head. Dilatation of left lateral ventricle and left sylvian fissure. Third ventricle displaced to left.

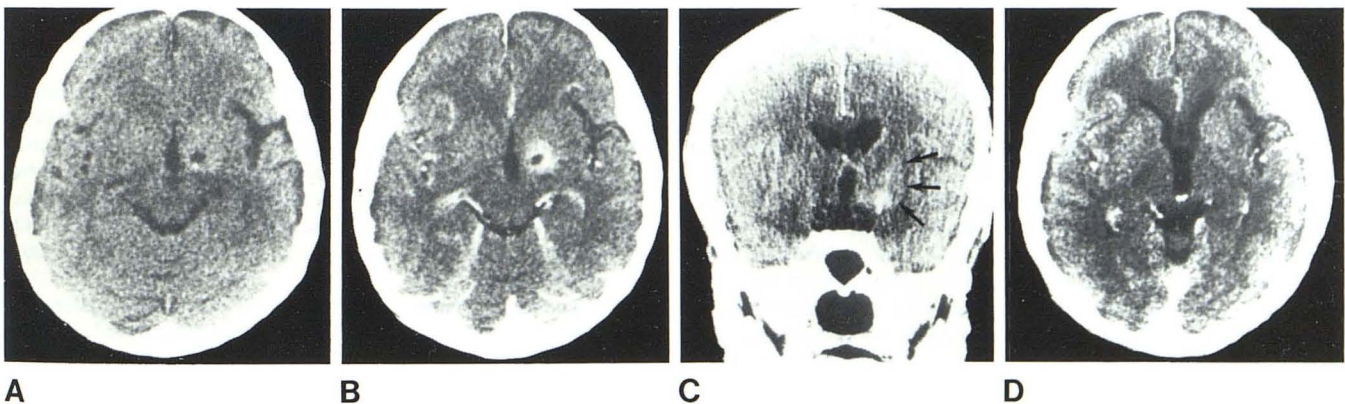
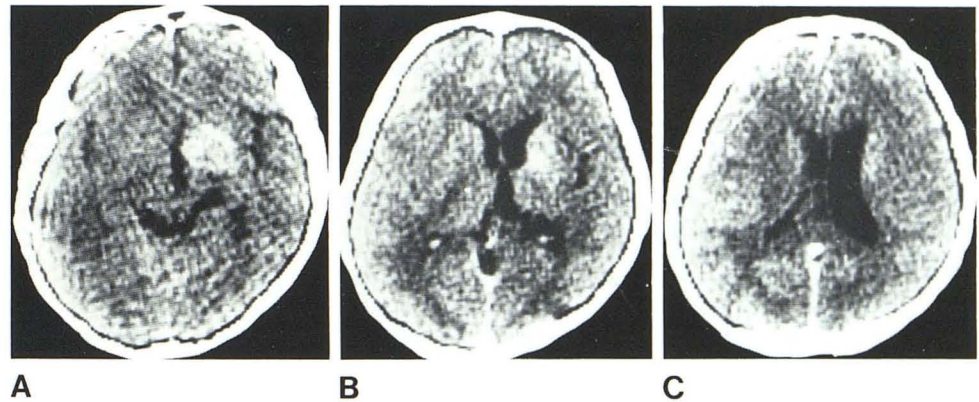


Fig. 3.—Case 3. Germinoma in 19-year-old boy. **A**, Precontrast CT. Nodular high-density lesion with small focal low-density center in region of left hypothalamus is enhanced (**B**) after injection of contrast material. Left sylvian fissure slightly dilated. **C**, Coronal CT shows extent of deep-seated

tumor (arrows). Ipsilateral displacement of third ventricle. **D**, Contrast-enhanced CT after radiation therapy (dose, 40 Gy). Signs of left cerebral hemiatrophy still present, but abnormal high-density lesion has resolved.

TABLE 1: Brain Tumors with Ipsilateral Cerebral Hemiatrophy

Case Reference No.	Age, Gender, Age at Onset (years)	Hemiparesis	Dementia	Personality Change	Hemiatrophy	Tumor Histology	Location			Confirmation
							BG	T	HT	
Numabe and Kamioka [3]	19, M, 14	R	+	+	L	Germinoma	+	+	...	Autopsy
Suzuki et al. [4]	14, M, 11	R	+	-	L	Germinoma	...	+	...	Surgery
Kwak and Suzuki [5]	14, M, 13	L	+	+	R	Germinoma	...	+	...	Surgery
Nakagawa et al. [8]	15, F, 13	L	-	-	R	Germinoma	+	+	+	Autopsy
Kwak et al. [6]	11, M, 8	L	+	-	R	Germinoma	...	+	+	Autopsy
Kwak and Suzuki [7]	10, M, 9	L	-	+	R	Germinoma	...	+	...	Surgery
Hamada et al. [9]	9, M, 7	L	+	+	R	Astrocytoma	+	+	+	Surgery
Maehara et al., Case 1	13, M, 12	R	+	+	L	Germinoma	+	+	+	CSF cell culture
Maehara et al., Case 2	10, M, 10	R	+	-	L	Germinoma	+	...	+	Surgery
Maehara et al., Case 3	19, M, 16	R	-	-	L	Germinoma	+	CSF cell culture

Note.—BG = basal ganglia; T = thalamus; HT = hypothalamus; R = right; L = left; + = present; - = absent; CSF = cerebrospinal fluid.

associated with signs of ipsilateral cerebral hemiatrophy (dilated ipsilateral lateral ventricle and sylvian fissure as well as midline shift toward the tumor side).

Discussion

In 1963, Numabe and Kamioka [3] reported a case of a 19-year-old boy with a 4 year history of right spastic hemiparesis, dementia, and emotional instability. Autopsy revealed an invasive ectopic pinealoma located in the left internal capsule and basal ganglia with left cerebral hemiatrophy. Suzuki, Kwak, et al. [4-7] reported four cases of ectopic pinealoma with ipsilateral cerebral hemiatrophy from 1968 to 1975. In 1973, Nakagawa et al. [8] reported a case of a 15-year-old girl with ectopic pinealoma associated with ipsilateral cerebral hemiatrophy, confirmed by autopsy. In 1977, Hamada et al. [9] published a case report of ipsilateral cerebral hemiatrophy and precocious puberty produced by astrocytoma. The patient was a 9-year-old boy with a 2 year history of left-sided motor weakness and mental retardation. The tumor appeared on CT as a high-density area involving the right thalamus, internal capsule, and caudate head. Hamada et al. did not offer specific comment on the use of contrast-enhanced CT, but they emphasized the value of CT in the diagnosis of such tumors and pointed out the histologic difference between their case (astrocytoma) and the other reported cases (pinealoma).

It is difficult to explain the mechanism of cerebral hemiatrophy secondary to brain tumors because of the dearth of evidence from autopsies. In 1974, Kwak et al. [6] presented an autopsy case of ectopic pinealoma and discussed the histopathologic correlation between the brain tumor and the ipsilateral cerebral hemiatrophy. In this case, postmortem examination showed the so-called two-cell-pattern pinealoma located in the hypothalamus and thalamus with invasive extension into the internal capsule and caudate head. In addition to this neoplastic process, degeneration and destruction of ganglion cells, demyelination, and damage to subcortical white matter were noted. The author suggested that hemiparesis was caused by damage to the internal capsule; personality change, by disturbance of the so-called emotional circuit in the hypothalamus, thalamus, and hippocampus; and dementia, by diffuse organic damage to the cerebrum. They hypothesized that the ipsilateral cerebral hemiatrophy was induced by Wallerian (orthograde) and retrograde degeneration resulting from damage to the thalamic ganglion cells and the afferent and efferent nerve fibers, secondary to invasive tumor in the thalamic region.

We agree with this hypothesis, but must speculate as to why invasive glioma in the same location does not cause ipsilateral

hemiatrophy as does germinoma. The presumable reason is the difference in age of the patients typically affected by these two types of tumor: germinomas are commonly seen in childhood, whereas thalamic gliomas occur in older patients. The case of astrocytoma with ipsilateral cerebral hemiatrophy reported by Hamada et al. [9] is an exception, since it occurred in a 9-year-old boy.

In summary, brain tumors with ipsilateral cerebral hemiatrophy are usually seen in young men, with disease onset occurring at 7-16 years of age; the clinical course is relatively chronic; and the major symptoms are hemiparesis, dementia, and personality change, without any signs of elevated intracranial pressure. It is quite interesting that all reported cases are Japanese. The probable reason for this is the high incidence of germinoma in Japan, where germinoma is more than three times more common than in other countries [10].

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