Ipsilateral cerebral atrophy with thalamic tumor of childhood

Case report

RYUNGCHAN KWAK, M.D., SHUN-ICHI SASO, M.D., AND JIRO SUZUKI, M.D.
Division of Neurosurgery, Institute of Brain Diseases, Tohoku University School of Medicine, Sendai, Japan

A case of cerebral tumor with ipsilateral cerebral hemiatrophy is presented and five previously reported cases are reviewed. The etiology of this entity is discussed on the basis of the symptomatological and clinicopathological findings noted in these six cases. It is suggested that ipsilateral cerebral hemiatrophy is due to thinning of the cerebral cortex with degeneration and disappearance of ganglion cells, demyelination in the subcortex, and destruction of axons. The mechanism proposed for ipsilateral cerebral hemiatrophy due to thalamic tumor is that thalamic tumor causes degeneration and disappearance of thalamic ganglion cells and nerve fibers, with consequent secondary Wallerian degeneration of afferent and projecting fibers from the thalamus as well as retrograde degeneration of efferent fibers, thus resulting in extensive atrophy of the cerebral cortex and subcortical tissue.

KEY WORDS □ ectopic pinealoma □ ipsilateral cerebral hemiatrophy □ thalamic tumor □ brain tumor in children

This report describes a case of brain tumor with ipsilateral cerebral hemiatrophy, and reviews five previously reported cases,\(^2\)\(^-\)\(^5\)\(^-\)\(^7\) three of them ours. An explanation is presented for the relationship between slowly growing thalamic tumors and ipsilateral cerebral hemiatrophy.

Case Report

In August, 1966, at the age of 8 years, this boy noticed slight paresis of the left arm. Seven months later he developed paresis of his left leg and he was admitted to the pediatric clinic of Tohoku University Hospital in March, 1967. Right-sided cerebral hemiatrophy was suspected on carotid angiogram and pneumoencephalogram, but an exact diagnosis was not made. He was discharged 1½ months later, but his neurological symptoms gradually became more severe. On March 6, 1969, the patient had a transient episode of stupor and was admitted to our clinic the next day.

Examination. Hemiparesis and pathological reflexes were noted on the left side, anisocoria (greater on the right than the left), loss of light reflex on the right side, slight paresis on the right oculomotor muscles, and horizontal nystagmus. There were no pathological findings in the cerebrospinal fluid. Cerebral hemiatrophy on the right side was suspected on carotid angiogram (Fig. 1 left), pneumoencephalogram (Fig. 1 right), and pneumotomogram. According to our pre-
Fig. 1. Left: Right carotid angiogram, anteroposterior view, shows slight shift of anterior cerebral artery from the left to right. Right: Pneumoencephalogram, anteroposterior view, shows slight shift to right side (tumor side) in the lateral and third ventricles. Air was apparent in the subarachnoid space, more on the right side than the left. Cerebral hemiatrophy on the right side was suspected.

Fig. 2. Left: Frontal section of brain at the level of the mamillary body. The tumor is located in the hypothalamus and thalamus on the right side with invasion of the capsula interna, nucleus lentiformis, and nucleus caudatus. Right: Frontal section through the trigone of the lateral ventricle. Dilatation of the ventricle system was marked on the right side. The right hemisphere was smaller than the left, and cerebral hemiatrophy of the right side is obvious. Arrow indicates tumor in the right hypothalamus.
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In a previous report, it was thought that cerebral hemiatrophy with the neurological symptoms on the opposite side indicated ipsilateral thalamic tumor, but tumor shadow was not obvious. The patient was discharged without surgical treatment. His condition deteriorated and he died of acute pneumonia on October 9, 1969.

Postmortem Examination. On autopsy, an invasive process was found that had destroyed the thalamus and hypothalamus on the right side (Fig. 2 left). Further diffuse cerebral atrophy and dilatation of the lateral ventricle was observed in the right cerebral hemisphere (Fig. 2 right). In other organs, acute bilateral hemorrhagic pneumonia and marked atrophy of the frontal lobe of the hypophysis, adrenal cortex, and thyroid gland were the main pathological findings.

On histological examination, the neoplasm was a two-cell-pattern pinealoma (Fig. 3 left) with giant astrocytes and perivascular round-cell infiltration (Fig. 3 right). In the right cerebral hemisphere, ganglion cells had partly disappeared or degenerated in the cortex (Fig. 4 upper left), and demyelination and destruction of axons (Fig. 4 upper right) were marked in the white matter of the temporal lobe (Fig. 4 lower left). In this area, proliferation of giant astrocytes and glial fibers was remarkable (Fig. 4 lower right). These pathological findings were in agreement with the microscopic diagnosis of cerebral hemiatrophy of the right side. There were no pathological findings in the pineal gland itself (Fig. 5).

Discussion

The six reported cases of brain tumor with overt ipsilateral cerebral hemiatrophy\textsuperscript{2-5,7} are summarized in Table 1. The onset of the disease was noted when the patients were between 8 and 14 years of age, with a mean of 11 years and 8 months. Duration of illness between onset and admission to the hospital ranged from 1 year and 2 months to 4 years, with an average of a little over 2 years and 1 month, showing a relatively chronic disease progression. Primary signs were marked decline of schoolwork in five cases, hemiparesis in four cases, and an episode of unconsciousness in one case, all of which were progressive throughout the course. The principal symptoms were hemiparesis, hemihypesthesia, character and emotional changes, deterioration of mental faculties, and behavioral abnormalities. It was interesting that none of the cases showed signs or symptoms of increased intracranial pressure before admission, and no papilledema was observed at the time of admission.

Ipsilateral cerebral hemiatrophy on the tumor side was evidenced by carotid angiography and pneumoencephalography (Fig. 1) and by exploratory craniotomy. Histopathological examination of the lesion revealed pinealoma in all six cases, of two-cell pattern in five cases and of one-cell pattern in

Fig. 3. \textit{Left:} Photomicrograph of the tumor, showing the two-cell pattern of a pinealoma. H \& E, \( \times 200 \). \textit{Right:} Photomicrograph showing giant astroglia in the surrounding tumor. Holzer, \( \times 100 \).
TABLE I

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Author, Year</th>
<th>Sex, Age on Onset</th>
<th>Period Onset to Admission</th>
<th>Symptoms*</th>
<th>Motor Hemiparesis</th>
<th>Characteristic and Emotional Change</th>
<th>Dementia</th>
<th>Abnormal Behavior</th>
<th>Other Symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Suzuki, et al., 1968</td>
<td>M, 11 yrs 9 mos</td>
<td>1 yr 5 mos</td>
<td>rt hemiparesis, schoolwork deteriorated</td>
<td>rt</td>
<td>no</td>
<td>yes</td>
<td>no</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>Kwak &amp; Suzuki, 1970</td>
<td>M, 13 yrs 4 mos</td>
<td>1 yr 6 mos</td>
<td>lt hemiparesis, schoolwork deteriorated</td>
<td>lt</td>
<td>careless, restless, superficial, impulsive, ready wit</td>
<td>yes</td>
<td>wandering, theft</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>Kwak, et al., 1974</td>
<td>M, 8 yrs 7 mos</td>
<td>2 yrs 6 mos</td>
<td>lt hemiparesis, schoolwork deteriorated</td>
<td>lt</td>
<td>no</td>
<td>yes</td>
<td>no consciousness disturbance</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>Kwak, et al., 1978</td>
<td>M, 9 yrs 2 mos</td>
<td>1 yr 2 mos</td>
<td>lt hemiparesis, schoolwork deteriorated</td>
<td>lt</td>
<td>careless, restless, talkative, superficial, ready wit</td>
<td>no</td>
<td>wandering, erotic behavior</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>Numabe &amp; Kamioka, 1963</td>
<td>M, 14 yrs</td>
<td>4 yrs</td>
<td>schoolwork deteriorated</td>
<td>rt</td>
<td>careless, restless, argumentative, violent, talkative, cheerful, superficial</td>
<td>yes</td>
<td>wandering, seizure incendiary, pass water, queer voice</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>Nakagawa, et al., 1973</td>
<td>F, 13 yrs</td>
<td>2 yrs</td>
<td>period of unconsciousness</td>
<td>lt</td>
<td>no</td>
<td>no</td>
<td>no</td>
<td></td>
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</tbody>
</table>

*None of the patients had symptoms of increased intracranial pressure.

The pineal body was noted to be normal in all three autopsy cases; ectopic pinealoma was also suspected clinically in the three remaining cases. The common site of tumor in this series was the thalamus and its surrounding areas.

The microscopically prominent features of the affected area included proliferation of giant astroglia within the tumor and in the adjoining areas (Fig. 3 right), general thickening of the cerebral cortex with degeneration and destruction of ganglion cells in the relatively superficial layer, a marked subcortical demyelination, disintegration of axons, and a marked proliferation of neuroglia and neuroglial fibers in this area (Fig. 4).

It has been reported by Greenfield, et al., that retrograde atrophy of the thalamus may
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<table>
<thead>
<tr>
<th>Examination</th>
<th>Tumor</th>
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<tr>
<td>Pressure of Lumbar Puncture (mm H₂O)</td>
<td>Radiological Findings</td>
</tr>
<tr>
<td>280</td>
<td>lt cerebral hemiatrophy</td>
</tr>
<tr>
<td>120</td>
<td>rt cerebral hemiatrophy</td>
</tr>
<tr>
<td>140</td>
<td>rt cerebral hemiatrophy</td>
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<tr>
<td>250</td>
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<tr>
<td>170</td>
<td>lt cerebral hemiatrophy</td>
</tr>
<tr>
<td>70</td>
<td>rt cerebral hemiatrophy</td>
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†BAR = BUdR-antimetabolite continuous intraarterial infusion-radiation therapy.

The histopathological findings noted in our Case 3, described here, seem to support our previously reported view on the pathogenesis of ipsilateral cerebral hemiatrophy associated with brain tumor.² That is: 1) thalamic tumor causes degeneration and disappearance of thalamic ganglion cells and nerve fibers; 2) secondary Wallerian degeneration of afferent and projecting fibers from the thalamus occurs, as well as retrograde degeneration of efferent fibers; thus 3) extensive atrophy of the cerebral cortex and subcortical tissue results (Fig. 6).

In slowly growing unilateral tumors of the thalamus, we think it is likely that unilateral cerebral hemiatrophy is brought about by the neoplastic growth in any kind of tumor. In

FIG. 5. Photomicrograph of pineal gland shows normal structure. H & E, × 400.

FIG. 6. Diagram of our speculated mechanism of ipsilateral cerebral hemiatrophy due to thalamic tumor.
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cases such as glioma in the thalamus with rapid growth, however, it seems that the patient's condition usually deteriorates and he succumbs to increased intracranial pressure before the development of cerebral atrophy.

References


Address reprint requests to: Ryungchan Kwak, M.D., Division of Neurosurgery, Institute of Brain Diseases, Tohoku University School of Medicine, 5-13-1, Nagamachi, Sendai, Japan.