Holocord Astrocytoma

—Case Report—

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Abstract

A rare case of intramedullary holocord astrocytoma extending from the medulla oblongata to the conus medullaris is reported. A 27-year-old male who had been suffering from nuchalgia for a few years was admitted to our department because of lumbago and gait disturbance. Neurological examination revealed sensory disturbances in various locations, weakness of the right lower extremity, mild swallowing disturbance, and bowel and bladder difficulties. Magnetic resonance (MR) images and myelograms showed a long cystic lesion extending from the medulla oblongata to the thoracolumbar spinal cord. Gadolinium-diethylenetriaminepenta-acetic acid (Gd-DTPA)-enhanced images detected a solid tumor located at the Th6-7 level. Two syrinx-subarachnoid shunts were placed at the upper cervical and Th12 levels. The solid neoplasm at the Th7 level was partially resected and histologically diagnosed as astrocytoma. Differentiation between cystic lesions and solid masses in the spinal cord is difficult. The usefulness of Gd-DTPA-enhanced MR imaging in the diagnosis of holocord tumor and the pathogenesis of secondary syringomyelia are discussed.

Key words: holocord tumor, pan-spinal tumor, pan-medullary tumor, astrocytoma, intramedullary tumor

Introduction

Spinal intramedullary tumors affecting the entire cord are rare; few such cases have been reported since the first by Cushing in 1927.⁶⁾ Despite the advances in magnetic resonance (MR) imaging facilitating diagnosis, differentiation between solid masses and cystic lesions is still difficult. Moreover, there are still some problems in the treatment of this disorder. We report here a case of holocord astrocytoma topologically diagnosed by using gadolinium-diethylenetriaminepenta-acetic acid (Gd-DTPA)-enhanced MR imaging.

Case Report

A 27-year-old male was admitted to the neurosurgical department of The Jikei University Hospital on April 21, 1988, because of progressive lumbago, sphincter dysfunction, unsteady gait, and dyspha-

gia. He had suffered from nuchalgia for 5 years and loss of pain and temperature sensations on the left upper extremity for 9 months.

Sensory examination on admission demonstrated analgesia on the left side including the face and marked hypesthesia on the left upper and bilateral lower extremities and the pubes. He could not walk unaided due to weakness of the right lower extremity. The deep tendon reflexes were exaggerated in the bilateral lower extremities. Swallowing disorders and bladder and bowel dysfunctions were observed.

Plain spinal x-ray films were normal. Lumbar myelograms showed a defect in the contrast medium shaped like a rugby ball, 17 mm in diameter, and 50 mm in length at the Th12 level and partial block by the swollen cord above this level (Fig. 1). Suboccipital myelograms showed a tapering block at the upper thoracic level. T₁-weighted MR images demonstrated a low-intensity lesion extending from the medulla to the thoracic cord (Fig. 2), which was high intensity on the T₂-weighted images. Coronal MR images with a surface coil showed a solitary

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Fig. 1 Lumbar myelograms, showing a rugby-ball-like defect at the Th12 level (arrowheads) and partial block above this level (arrows).

Fig. 2 T₁-weighted MR images, showing a cystic lesion extending from the medulla oblongata to the thoracic spinal cord.

high-intensity lesion at the Th12 level. Under the diagnosis of an ependymoma of the conus medullaris accompanied by syringomyelia developing superiorly, an operation was performed.

A laminectomy from Th11 to L1 followed by a small incision in the swollen cord revealed a syrinx containing xanthochromic fluid. The syrinx continu-

ously extended rostrally but ended in a blind sac caudally. Biopsy of the cystic wall demonstrated proliferation of reactive astrocytes but no tumorous components. A shunt tube was placed between the syrinx and the subarachnoid space (Fig. 3). An occipital craniotomy and laminectomy at C1 were then performed to expose the swollen medulla oblongata and upper cervical cord. A small incision 2–3 mm in depth into the fasciculus gracilis revealed the large syrinx. A shunt tube was placed between the syrinx and the cervical subarachnoid space. There was no communication between the fourth ventricle and the syrinx, and no tumor was discovered.

Postoperatively, both syringomyelia and syringobulbia contracted. Postoperative Gd-DTPA-enhanced MR images revealed high-intensity, swollen spinal cord centered at the Th6 and 7 levels (Fig. 4). Since this was suspected to be a solid tumor, a second operation was performed.

After laminectomy from Th5 to 8, the yellowish, swollen spinal cord with abnormal vessels was ob-

Fig. 3 Operative photograph, showing the spinal cord exposed at the Th12 level. The syringomyelic cavity was opened, and a syrinx-subarachnoid shunt emplaced.

Fig. 4 Postoperative Gd-DTPA-enhanced MR image, showing intense enhancement of swollen cord at Th6–7.

Fig. 5 Operative photograph, showing a solid tumor in the spinal cord at Th6-7.

served. It was partially excised at Th7 (Fig. 5) and histologically diagnosed as astrocytoma.

Postoperative focal irradiation of 47.7 Gy was performed. The swallowing difficulties, nuchalgia, and back pain disappeared. The sensation and thermal paresthesia on the left upper extremity and trunk and weakness of the right lower extremity were improved. Nine months later, he returned to his employment.

Discussion

Spinal intramedullary tumors sometimes extend both superiorly and inferiorly along almost the entire cord. This has been known for a long time since the first report by Cushing⁶⁾ in 1927 followed by Horrax and Henderson¹⁵⁾ in 1939. Fischer *et al.*¹²⁾ reported two cases of ependymoma using the term "pan-spinal" ependymoma. Epstein and Epstein⁹⁾ reported three cases of astrocytoma and called them "holocord" tumors. All these cases had a cyst extending over several vertebrae. An unusual, pan-spinal intramedullary tumor, which was a solid mass not accompanied by a cyst, was termed as a "pan-medullary" tumor by Tedeschi *et al.*³²⁾ Thus, spinal intramedullary tumors growing along the entire cord are usually accompanied by a cyst and, regardless of the proportion of the solid mass to the cyst, are classified as mentioned above. According to Nikaido *et al.*²⁶⁾ and Tanaka *et al.*,³¹⁾ we have decided to use the term holocord tumor.

The incidence of holocord tumor in children has been reported to be 3-8% of all spinal tumors^{5,29)} and 8% of all intramedullary tumors³³⁾ and, including adults, 5% of all intramedullary tumors.¹⁴⁾ A summary of detailed reports on holocord tumors containing 24 cases, including ours, is given in Table 1. Such tumors usually occur in young patients (mean

age, 14.1 years) and in males (male to female ratio, 15:9). The initial symptoms include motor paresis and pain as in other spinal tumors. Examination may reveal irregular sensory impairment at any level of the spinal cord.

The tumor histology was ependymoma in eight cases, astrocytoma in seven, teratoma in five, lymphoma in one, small cell glioma in one, and ganglioglioma in one. In the teratomas, a large part of the tumor was located intramedullary. Interestingly, almost all the holocord tumors were accompanied by cysts regardless of the histological tumor type, and such cysts could be classified into two types. Type I consists of multiple small cysts within the tumor and is most frequently observed in teratomas and astrocytomas. Type II consists of a large cyst formed above and/or below the tumor mass, and the wall of this type cyst sometimes has Rosenthal fibers but no tumor cells. Liber and Lisa²⁰⁾ observed Rosenthal fibers also in non-neoplastic syringomyelia. Thus, these fibers are not a characteristic feature of the wall of the cyst accompanying tumors and, moreover, do not indicate the presence of tumor. At present, it is unclear whether the term syrinx or cyst should be used for liquid retention associated with tumor, but in general, the term cyst is preferred. Only large cysts with the wall containing no tumor cells, as type II, are termed secondary syringomyelia.

Although reports of holocord tumors are rare, secondary development of syringomyelia in cases of spinal intramedullary tumor is not rare, with the reported incidence of 16-58%.^{11,18,21)} Tannenberg suggested that this is due to retention of tissue fluid caused by pressure from the tumor.²¹⁾ Jonesco-Sisesti¹⁶⁾ reported different congenital causes for syringomyelia and the tumor. Russell²⁸⁾ stated that the syrinx results from a secondary effect of ischemia or tumoral hemorrhage. Liber and Lisa²⁰⁾ believed that the blockade of the perivascular space by the tumor cells hampers drainage of tissue fluid and forms the syrinx. Feigin *et al.*¹⁰⁾ advocated an important role of edema in the formation of the syrinx. However, it is difficult to believe that such a long syrinx could be formed only by the above mechanisms, so there must be some other mechanisms developing the syrinx. In regard to the primary cause of syringomyelia, Ball and Dayan²⁾ suggested that as the pressure in the spinal canal increases, the cerebrospinal fluid (CSF) extends into the medulla *via* the perivascular space, and Williams³⁵⁾ also emphasized pressure changes in the spinal canal. Nagahiro *et al.*^{23,24)} reported that such abnormal CSF circulation is likely to occur under conditions in which a tethering or subarachnoid block is caused by the tumor. A reported case

Table 1 Intramedullary holocord tumors

Author (Year)	Age	Sex	Location of tumor	Location of cyst	Tumor histology	Surgery	Result
Cushing (1927) ⁶⁾	8 yrs	F	medulla-conus	Th2-conus	ependymoma	total removal	improved
Horrax and Henderson (1939) ¹⁵⁾	20 yrs	F	medulla-conus	Th9-conus	ependymoma	total removal	improved
Lowenberg (1939)*	14 yrs	M	medulla-lumbar	multiple	astrocytoma	?	?
	26 yrs	F	medulla-conus	multiple	?	?	?
Black and German (1950) ³⁾	5 mos	M	medulla-conus	conus	teratoma	partial removal	died after 10 yrs
David <i>et al.</i> (1965) ⁷⁾	32 yrs	M	C1-L1	?	small cell glioma	total removal	no change
Mikagi <i>et al.</i> (1968) ²²⁾	6 mos	M	pons-conus	multiple	teratoma	—	died
Fischer <i>et al.</i> (1975) ¹²⁾	27 yrs	F	medulla-conus	upper part, lower part	ependymoma	?	?
	24 yrs	M	C2-L2	cervical, dorsal-lumbar	ependymoma	total removal	improved
Pickens <i>et al.</i> (1975) ²⁷⁾	6 wks	F	medulla-conus	multiple	teratoma	—	died
Kajita <i>et al.</i> (1975) ¹⁷⁾	4 mos	F	upper cervical-conus	multiple	teratoma	—	died
de Divitiis <i>et al.</i> (1978) ⁸⁾	31 yrs	M	C1-L1	Th6?-L1	ependymoma	total removal	improved
Albright and Byld (1980) ¹⁾	11 yrs	M	medulla-conus	medulla-C5, conus	ganglioglioma	total removal	improved
Nakamura <i>et al.</i> (1980) ²⁵⁾	6 yrs	F	pons-conus	—	ependymoma	—	died
Epstein and Epstein (1981) ⁹⁾	15 yrs	M	medulla-conus	medulla-C4, lower part	astrocytoma	subtotal removal	improved
	5 yrs	M	medulla-conus	medulla-C4, Th9-conus	astrocytoma	subtotal removal	improved
	4 yrs	M	medulla-conus	medulla-C4, Th4-conus	astrocytoma	subtotal removal	improved
Tedeschi <i>et al.</i> (1982) ³²⁾	12 yrs	F	C1-L2	multiple	astrocytoma	subtotal removal	improved
Nikaido <i>et al.</i> (1984) ²⁶⁾	2 yrs	M	C4-conus	C4-Th3, Th8-conus	astrocytoma	subtotal removal	died
Nagahiro <i>et al.</i> (1986) ²⁴⁾	23 yrs	M	medulla-filum	medulla-Th12	ependymoma	total removal	improved
Tanaka <i>et al.</i> (1986) ³¹⁾	14 yrs	M	medulla-filum	medulla-conus	ependymoma	total removal	improved
Landan <i>et al.</i> (1987) ¹⁹⁾	37 yrs	F	C2-conus	C2-conus	primary lymphoma	myelotomy	died
Urui and Oi (1988) ³⁴⁾	67 days	M	whole cord	?	teratoma	partial removal	no change
Present case	27 yrs	M	medulla-conus	medulla-Th5, Th9-conus	astrocytoma	partial removal	improved

*Cited from Tanaka *et al.*³¹⁾

of syringomyelia associated with extramedullary meningioma⁴⁾ suggests that the same mechanism involved with primary syringomyelia develops the syrinx associated with the tumor.¹³⁾

Radiological examination for holocord tumor includes plain spinal x-rays, which reveal the widening of the space between the pedicles of the vertebral arches and depressions in the vertebral bodies, and myelography, which shows enlargement of the spinal

cord. Imaging diagnosis has been greatly facilitated by the development of MR imaging. Tanaka *et al.*³¹⁾ investigated the MR findings for an ependymoma accompanied by a holocord cyst and differentiated the tumor from the syringomyelia by comparing the T₁ values. Slasky *et al.*³⁰⁾ found that, in the differential diagnosis between the spinal cord tumor and the syrinx or cyst, MR imaging following the administration of Gd-DTPA was useful. In our case, MR imag-

ing could easily identify the cystic lesion extending from the medulla oblongata to the cervical spine. However, a topological diagnosis of the solid mass was difficult. Since myelography demonstrated swollen spinal cord, the coronal images of the thoracic and lumbar spine using a surface coil were obtained but were unable to delineate the tumor mass. Only after administration of Gd-DTPA, was the actual tumor mass adequately identified.

Most ependymomas can be completely extirpated since the edge of the tumor is clearly defined. However, astrocytomas do not have clearly defined borders so that surgical treatment is still debatable. Epstein and Epstein⁹⁾ reported that although an extensive posterior myelotomy alone did not cause neurological deficit, excessive manipulation during the operation may cause permanent damage. They achieved success in conducting complete macroscopic extirpation using an ultrasonic surgical aspirator. Tedeschi *et al.*³²⁾ also reported successful complete macroscopic extirpation of solid astrocytoma. However, Guidetti *et al.*¹⁴⁾ reported that since the borders of most astrocytomas are unclear, complete extirpation is difficult and may frequently cause aggravation of neurological symptoms. In our case, since the border of the actual tumor was unclear, only laminectomy and partial extirpation were performed. Also, since no tumor cells were observed in the wall of the syrinx, complete extirpation of the syrinx was very likely to result in aggravation of the neurological symptoms, so a shunt tube was placed between the syrinx and the subarachnoid space.

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