Extradural Extension of Primitive Neuroectodermal Tumor
—Case Report—

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Abstract
A 13-year-old boy presented with a primitive neuroectodermal tumor (PNET) with unusual extracranial extension. Precontrast computed tomography showed the tumor as a ring-shaped high-density area which was enhanced postcontrast, with a low-density center. Magnetic resonance imaging showed the tumor as a low-intensity area on the T<sub>1</sub>-weighted images with marked enhancement by gadolinium-diethylenetriaminepenta-acetic acid, and high intensity on the T<sub>2</sub>-weighted images. The central area appeared as high intensity on both images, suggesting free methemoglobin. The tumor was subtotally removed. Histological examination demonstrated PNET. Despite irradiation (20 Gy) to the surgical site, and further tumor removal, he died 6 months later. This case showed PNET can extend extracranially. Diagnosis and treatment of such a tumor located extracranially and intracranially require careful consideration.

Key words: extradural extension, primitive neuroectodermal tumor

Introduction
Primitive neuroectodermal tumor (PNET) was first described in 1973,<sup>2</sup> and about 150 cases have been reported since.<sup>1</sup>-<sup>10</sup> The common features include predominance in early life, clinical malignancy, occurrence in the cerebrum, grossly cystic and hemorrhagic with sharp borders, microscopically malignant and predominantly undifferentiated with evidence of focal attempts to differentiate along glial and neuronal lines, and prominent mesenchymal component.<sup>5</sup> The prognosis of PNET is extremely poor in spite of surgical and radiation treatment. The concept of PNET has been extended collectively to undifferentiated neuroepithelial tumors in infancy and childhood,<sup>2,10</sup> and there are no accepted criteria at present. Direct extracranial extension of PNET has been observed in only one case.<sup>11</sup> We report a 13-year-old boy with unusual extracranial extension of an intracerebral tumor with a histological diagnosis of PNET.

Case Report
A 13-year-old boy was admitted to our hospital with vomiting and convulsion on March 27, 1990. A plain skull x-ray film showed thinning and slight protrusion of the temporal bone. Precontrast computed tomography (CT) showed a ring-shaped high-density area, containing a low-density central area, in the right temporal lobe and midline shift. The pituitary stalk also appeared as a high-density area (Fig. 1 left). Postcontrast CT showed the ring-enhanced tumor and enhanced pituitary stalk (Fig. 1 right). The tumor appeared as a low-intensity area on the T<sub>1</sub>-weighted magnetic resonance (MR) images and high-intensity area on the T<sub>2</sub>-weighted images, and was markedly enhanced by gadolinium-diethylenetriaminepenta-acetic acid (Gd-DTPA). T<sub>1</sub>- and T<sub>2</sub>-weighted MR images showed the central area as a high-intensity area, suggesting free methemoglobin (Fig. 2). Coronal MR images showed that the tumor had penetrated the base of the middle fossa, extend-
ing to the infratemporal fossa, and a linear low-intensity area was visible in the tumor (Fig. 3). Cerebral angiography confirmed the middle meningeal artery at this site but did not show a clear tumor stain.

A right frontotemporal craniotomy was performed. The tumor was soft, necrotic, and grayish, and with relatively clear margins, but the pituitary stalk was macroscopically invaded by the tumor. The fluid within the tumor was previous hemorrhage. The tumor adhered to the dura at the base of the middle fossa, and parts of the dura and bone were destroyed. The intracranial lesion was subtotally removed. Temporal lobectomy was also performed. Oxycel and Biobond were used to repair the dural defect.

Histological examination of the surgical specimen showed aggregation of undifferentiated cells with small bodies and abundant chromatin, mitosis, focal necrosis, and structures resembling Homer Wright rosettes (Fig. 4). Some areas were rich in connective tissue. However, the center of the tumor had relatively little connective tissue. Immunohistochemical staining showed that a few cells were positive for glial fibrillary acidic protein and many were positive for vimentin. No cells stained positively for neuron-specific enolase or neurofilament. The histological diagnosis of PNET was based on these findings.

He received radiation therapy at the surgical site. However, irradiation could not be continued over 20 Gy because of general fatigue, nausea, and vomiting which was refractory to treatment. About 2 months postoperatively, symptoms of increased intracranial pressure recurred before radiation therapy was resumed. The residual tumor was again removed, but he died of development and dissemination of the tumor 6 months after admission. The histological findings of the specimen from the second operation
Discussion

The diagnosis in our case was based on the characteristics reported by Hart and Earle.5) The possibility of extracranial malignant tumor extending intracranially could not be ruled out completely, but the histological findings were characteristic of PNET.

PNET tends to be disseminated through the cerebrospinal fluid.1) Hematogenous extracranial metastasis of PNET has been observed.1,3,8) Only one case of direct extracranial extension has been reported, in which skull x-ray films showed erosion of bone involving the anterior fossa, extending posteriorly from the left median orbital rim to include both sides of the midline, sella, and sphenoid sinus.30) In our case, CT and MR imaging suggested extracranial extension which was confirmed at operation. Extradural extension of malignant gliomas is considered rare because the dura acts as a barrier.8,9) Orita et al.8) suggested that malignant gliomas extended extracranially by directly destroying the dura mater or passing through discontinuities where major vessels, cranial nerves, and spinal nerves penetrate the dura mater. In our patient, the MR imaging and operative findings suggested that the tumor had destroyed the dura, then extended extracranially from the foramen spinosum along the middle meningeal artery.

References


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