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原　　著

Analysis of the diagnostic process in pediatric posterior fossa tumors

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後頭蓋窩腫瘍小児例の発症から診断にいたる過程に関する検討

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Abstract

Few focal neurological signs are seen in pediatric cases of posterior fossa tumor. Promptly establishing the diagnosis is thus often difficult in the absence of neuroradiological findings. The aim of this study was to evaluate the diagnostic process for children with posterior fossa tumors.
We retrospectively analyzed 27 children with posterior fossa tumor treated in our department. Diagnoses included medulloblastoma (n=11), pontine glioma (n=5), astrocytoma (n=3), ependymoma (n=2), and other tumors (n=6). The most common initial symptoms were headache, vomiting, and ataxia. Mean duration between symptom onset and radiological diagnosis was 1.9 months (range, 3 days to 7 months). Eleven patients were initially diagnosed with common cold or gastrointestinal infection by general physicians and pediatricians. Cases of posterior fossa tumor are difficult to diagnose based solely on general findings. However, ataxia in addition to repeated headache and vomiting is the key symptom in diagnosis. Physicians must bear in mind the possibility of posterior fossa tumors when examining children presenting with headache and vomiting.

要　　旨

小児後頭蓋窩腫瘍症例において症状出現から放射線学的診断が得られるまでの経緯，時間など過程に関する検討を行った。対象は当院で治療された15歳以下の小児後頭蓋窩腫瘍27例である。初発症状は，頭痛，嘔吐，ふらつきが高頻度であった。診断までの期間は３日から７ヶ月（平均1.9ヶ月）であった。放射線学的診断前に何らかの医療機関を受診した症例は11例で，全例感冒あるいは胃腸炎と診断されていた。小児後頭蓋窩腫瘍では，巣症状を欠き神経学的所見だけの診断が困難である。しかしながら頭痛，嘔気，嘔吐は最も頻度の高い診断の鍵となる症状であり，初診医は後頭蓋窩腫瘍の可能性も念頭に放射線学的検査を考慮しなければならない。

Key words : posterior fossa, tumors, children, diagnosis, ataxia

■和訳

小児後頭蓋窩腫瘍症例では巣症状に乏しく、神経学的検査だけでは診断が困難な場合が少なくない。今回われわれは症状出現から放射線学的診断が得られるまでの経緯、時間など過程に関する検討を行った。対象は当院で治療された15歳以下の小児後頭蓋窩腫瘍27例である。病理診断は髄芽腫11例、脳幹部神経膠腫５例、星細胞腫３例、上衣腫２例、その他６例であった。初発症状は、頭痛、嘔吐、ふらつきが高頻度であった。診断までの期間は３日から７ヶ月（平均1.9ヶ月）であった。放射線学的診断前に何らかの医療機関を受診した症例は11例で、全例感冒あるいは胃腸炎と診断されていた。小児後頭蓋窩腫瘍では、巣症状を欠き神経学的所見だけの診断が困難である。しかしながら頭痛、嘔吐、ふらつきは最も頻度の高い診断の鍵となる症状であり、初診医は後頭蓋窩腫瘍の可能性も念頭に放射線学的検査を考慮しなければならない。

■INTRODUCTION

Intracranial tumors are the most common solid neoplasm in children after leukemia, occurring in 2.5-4 per 100,000. Posterior fossa tumors account for approximately 60-70% of all brain tumors in children[1-4]. In treating these lesions, correct and prompt diagnosis represents a significant challenge.

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While tumors in the supratentorial regions show initial symptoms of hemiparesis, aphasia and seizure, few focal neurological deficits are seen in cases with posterior fossa tumor. General physicians and pediatricians have many chances to examine these patients before neurosurgeons. Pediatricians are involved in the daily medical treatment of children, but have few chances to deal with childhood brain tumors, particularly posterior fossa tumors. This lack of experience can lead to misdiagnosis and delays in reaching the correct diagnosis.

In this report, we analyzed the diagnostic process for children with posterior fossa tumors.

■MATERIAL AND METHODS

We retrospectively reviewed the medical charts of 27 children with posterior fossa tumors treated our department. Patient charts were reviewed for the following information: first symptom; referred first physicians; duration to established radiological diagnosis; presenting symptoms; and histological diagnosis.

■RESULTS

The most common initial symptoms were headache, vomiting and ataxia (Table 1). Mean duration between onset of symptoms and radiological diagnosis was 1.9 months (range, 3 days to 7 months) .Nine patients were diagnosed within the first month, 11patients within 2 months, 4 patients within 3 months, 2 patients within 4 months and 1 took 7months (Table 2).

Fifteen patients were referred to initial medical institutes prior to neuroradiological diagnosis. Initial diagnosis was common cold or gastrointestinal infection for 10 cases by pediatricians, and for1case by general physician (Table 3). Among these 11 cases, 7 presented with hydrocephalus, so headache and vomiting suggested to reflect high intracranial pressure due to hydrocephalus.

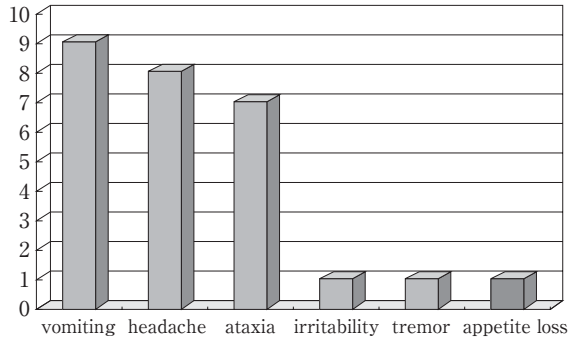

Table 1 Initial clinical symptoms

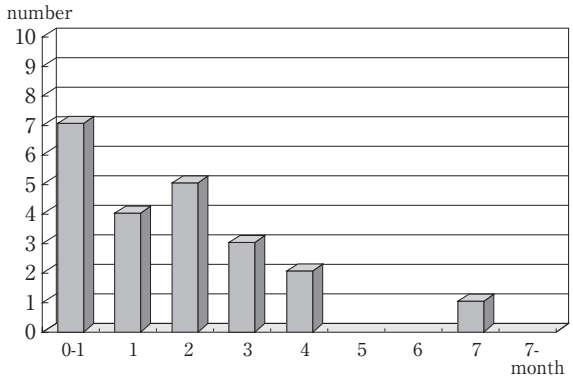

Table 2 Number of patients diagnosed within a certain time period after onset of initial symptoms.

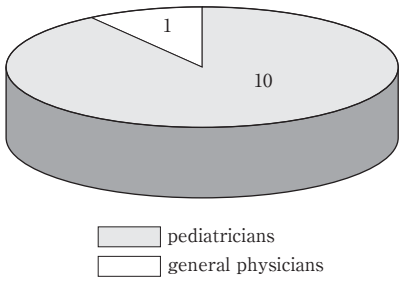

Table 3 Number of referred first physicians before the admission to our hospital.

■CASE PRESENTATION

Case1

A 4-year-old girl was referred to a general hospital due to loss of appetite and vomiting. A pediatrician diagnosed gastrointestinal infection and intravenous drip infusion was performed on admission. However, disturbance of consciousness developed and the patient fell into a coma, and was subsequently transferred to another hospital. Computed tomography (CT) revealed a hemorrhagic lesion in the 4th ventricle (Fig. 1), and she was transferred to our hospital. Emergent operation was performed, but she died without recovery.

Case2

A 14-year-old girl developed headache and repeated vomiting, and was referred to a general hospital. A pediatrician diagnosed as gastrointestinal infection and prescribed symptomatic therapy. However, vomiting did not resolve and gastoroendoscopic examination revealed no abnormalities. Four months later, magnetic resonance imaging (MRI) disclosed 4th ventricle tumor (Fig. 2), and she was referred to our hospital. On admission, the patient was unable to walk due to truncal ataxia. Surgical resection was performed and the pathological diagnosis was medulloblastoma.

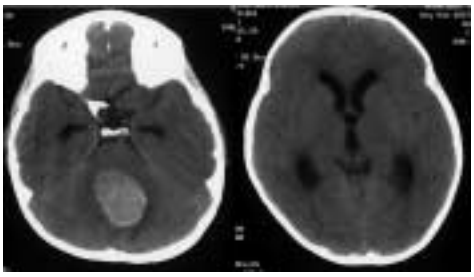

Fig. 1 CT scanning revealing a hemorrhagic lesion in the 4th ventricle.

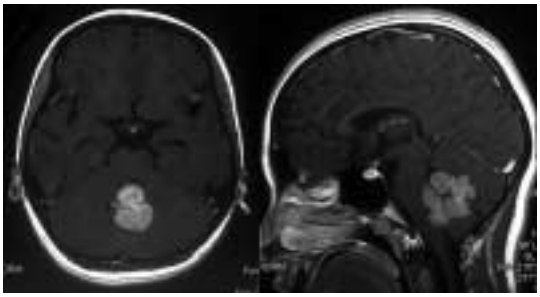

Fig. 2 MR imaging disclosing4th ventricle tumor.

■DISCUSSION

Pediatric brain tumors are the second-most common solid neoplasm after leukemia[1-4]. However, these lesions differ from adult brain tumors in several ways. The types of tumors encountered in children differ from those in adults. Location is in the posterior fossa region in approximately 60-70% of cases, and midline lesions are particularly common[1-4]. However, when treating this category of disorder, one of the most serious points is misdiagnosis and delayed diagnosis.

The mode of presentation depends on the age of the child, as well as the location of the tumor. Posterior fossa tumors manifest a variety of clinical symptoms, including headache, nausea, vomiting, ataxia, and cranial nerve symptoms depending on the tumor type. In general, cerebellar astrocytomas exhibit a long history of ataxia and symptoms of increased intracranial pressure secondary to non-communicating hydrocephalus. Medulloblastomas and ependymomas produce similar symptoms to cerebellar astrocytomas, but with swifter progression. Brainstem tumors present with facial palsy and extraocular muscle palsies, ataxia, and hemiparesis[1,4] Dörner mentioned the presence of torticollis in childhood posterior fossa tumors[5]. Torticollis has a wide range of differential diagnoses, but initial symptoms and signs tend to be nonspecific without focal neurological deficits in comparison with supratentorial tumors, leading to misdiagnosis and delayed diagnosis. Of course, infants and younger children cannot explain their symptoms by themselves.

Most patients were seen and referred by general physicians and pediatricians. These are professionals in the field of seeing patients on a regular basis and following physical development, but can be unfamiliar with brain tumors, and show a tendency to misdiagnose. Awareness of pediatric brain tumors within the population is low, and knowledge among general physicians and pediatricians is limited. This also leads to misdiagnosis and delayed diagnosis for pediatric posterior fossa tumors. In our series, 11 patients referred to initial medical institutes were diagnosed with common cold or gastrointestinal infection by pediatricians and general physicians.

Among subjective symptoms including statements from parents, feelings of dizziness and unsteadiness can be key to diagnosing posterior fossa tumor, representing truncal ataxia. Repeated, frequent headaches and vomiting in the absence of neurological focal signs should alert clinicians to the possibility of posterior fossa tumors, and thus the need for neuroradiological examinations such as CT and MRI without hesitation.

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