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Hypothalamic Hamartoma Associated with Precocious Puberty: Case Report

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〈국문초록〉

시상하부 과오종에 의한 사춘기 조발증 : 1예 보고

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배 원 경·김 표 년·김 일 영·이 병 호
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시상하부의 과오종은 비교적 드문 종양으로 대부분 6세 이전에 나타나며, 임상적으로는 성조숙과 간질발작을 나타낸다. 일반적으로 과오종은 크기가 작고 CT상 뇌실질과 같은 동종음영으로 보이기 때문에 진단에 어려움이 있다. 그러나 특징적인 병소의 위치와 MR 소견을 비교하므로서 과오종의 발견이 용이하다. 저자들은 수술로서 확진된 시상하부의 과오종과 특징적인 임상증상을 보인 8세 여아에서, 뇌혈관 조영상, CT 및 MR상을 종합 비교하여 그 진단방법과 각각의 특징적인 소견을 문헌고찰과 함께 보고하는 바이다.

—Abstract—

Hamartoma of the hypothalamic area is a well-recognized cause of central precocious puberty. We report a case of histologically proven hypothalamic hamartoma in a 8-year-old girl with precocious puberty. The CT showed an isodense, nonenhancing mass in suprasellar area, measuring 4.2X3.1cm, which, to our knowledge, seems to be the largest one of the published cases. On MR imaging, the signal intensity of the mass was homogeneous and isointense relative to gray matter on T1-, and hyperintense on T2-weighted images. The clinical and radiologic findings of the published cases of hypothalamic hamartoma are reviewed.

Index Words: Brain neoplasms, diagnosis
Brain neoplasms, CT
Brain neoplasms, MR
Hamartoma

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Introduction

A hamartoma is not a true neoplasm. It is a congenital malformation consisting of a tumor–like collection of normal tissue lodged in an abnormal location[1]. Reports of hamartoma of the hypothalamic region are relatively rare, approximately 90 cases have been reported in the world literature[2~8]. They are particularly interesting because of not only their symptoms but also the still mysterious mechanism that causes the symptoms[9]. The most common presenting symptoms associated with this lesion are isosexual precocious puberty and seiaures, including epileptic laughter(gelastic seizure). In most cases, the tumors originate in the area of the tuber cinereum and extend into the basal cistern: they rarely exceed 2 cm in diameter[2,6,9]. Histologically, they made up of nerve cells smillar to those seen in the tuber cinereum and adjacent hypothalamic structures. We are reporting a case of hypothalamic hamartoma, and describe pertinent CT and MR characteristics.

Case Report

A 8-year-old girl was admitted because of headache and frequent seizure of 2 year duration. The patient was born in a full-term spontaneous delivery. No family history of sexual precocity could be elicited. The parents and siblings were healthy and of average height. Physical examination revealed a well nourished and developed female with normal body proportions: 147cm tall and weighed 40kg. Her hight and weight greater than the 97th percentile for her age. Bilateral breast development (Tanner stage 2) was seen. No other physical or neurological abnormalities were identified. Bone age was 11 years compared with chronological age of 8 years. She had epileptic laughter 2 to 3 times a day during the first several days of admission. The endocrinological study revealed slight elevation of serum estradiol level(32.43 pg/ml), but levels of serum leutenizing hormone(LH), follicle stimulating hormone(FSH), testosterone,and prolactin were within normal range.

A skull series showed erosion of the tip of the dorsum sella. CT scan showed 42×31 mm sized isodense mass in the prepontine and suprasellar cisterns, and small cystic low density area in left posterior portion of this lesion. No enhancement was noted following contrast administration(Fig. 1). Bilateral carotid angiograms were normal. A vertebral angiogram showed marked posterior displacement of the basilar artery and anterior pontomesencephalic vein. No tumor blush was identified. MR imaging demonstrated a distinct mass occupying a large portion of the suprasellar cis-

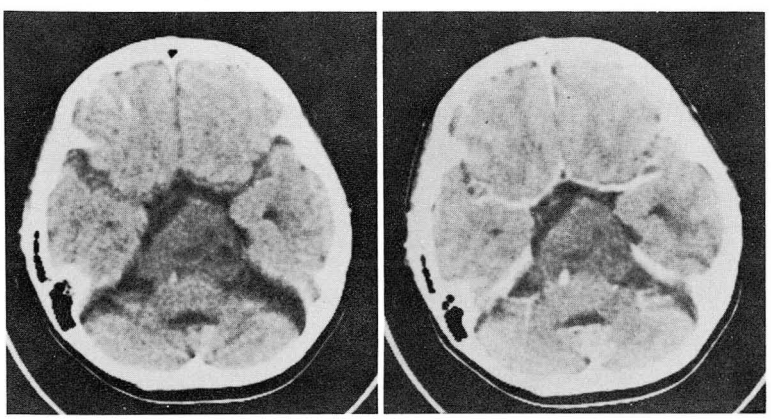

Fig. 1. Axial CT scan shows a large isodense mass in the basal cistern, and no evidence of enhancement in post-contrast scan.

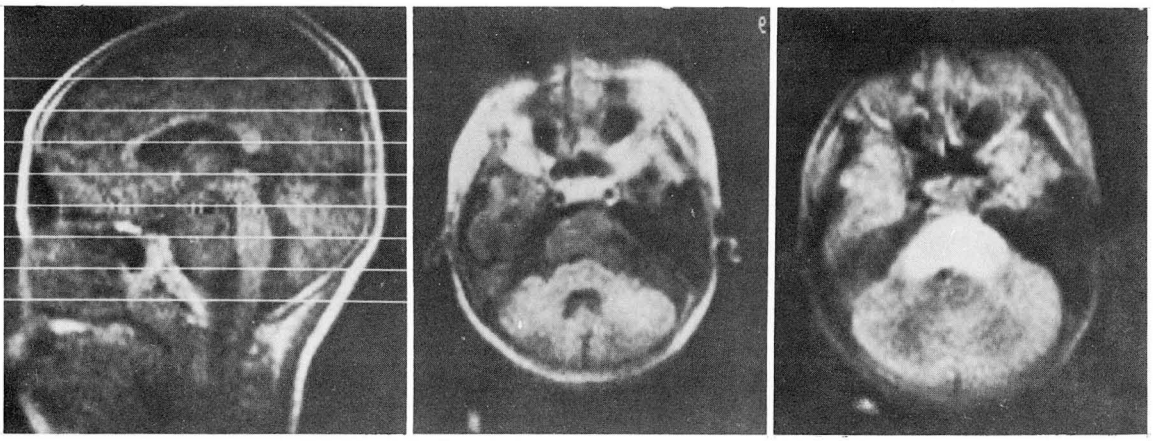

Fig. 2. T1-WI(TR 400msec, TE 30msec) show isointense mass, and homogeneous high signal intensity lesion on T2—WI(TR 1800 msec, TE 90 msec).

tern(Fig. 2). The mass was isointense to gary matter on T1–weighted images. The abnormal area was best demonstrated on T2–weighted images, appearing as a homogeneous, high signal intensity lesion.

Craniotomy was performed and a hamartoma was partially resected. Histologically, the tumor consisted of astrocytes and glial cells mixed with neurons, and focal minute calcification were noted in the tissue. No evidence of anaplasia was ween.

Discussion

In hypothalamic hamartomas, the nerve cells composing the malformation always resemble those of the tuber cinereum together with normal glial cells. In most cases this lesion has an anatomical connection with the tuber cinereum, usually by way of a distinct stalk[7,11]. Most of these lesions are small, with a diameter of a few millimeters to 1cm, rarely exceeding 2cm[2,6,10]. But size of the present case was about 4×3 cm, Which, to our knowledge, seems to be the largest one of the published cases of hypothalamic hamartoma. They are pedunculated growths with attachments to the posterior thalamus between the tuber cinereum and the mamillary bodies. They fill the free space between the optic chiasm and pons, and usually do not distort the thalamus or other parts of the base of the brain unless they are very large. These lesions occur with equal frequency in males and females. Symptoms occur early in life, before the age of 6 years in most published cases, and frequently before the age of 6 months[12]. Presenting clinical sings are isosexual precocious puberty, epileptic laughter, and mental changes. Patients with hypothalamic hamartoma may be divided into two clinical groups[13]. In the first group, secondary sexual characteristics predominate. Breast development, growth of pubic and axillary hair, and enlargement of external genitalia precede gonadal enlargement. The growth rate is increased and radiographic bone age is far ahead of chronological age. The hamartoma in these patients is usually less than 1.5cm in diameter. In the second group, sexual precocity may be absent. These patients present with gelastic seizures, intellectual impairment and psychic disturbances often correlating with a hamartoma greater than 2.0cm. The present case appears to be one of the second group, but revealed precocious puberty, increased growth rate, and gelastic seizure.

The diagnostic evaluation of precocious puberty has relied on neuroimaging studies to demonstrate or exclude hypothalamic hamartoma. A plain skull series is usually normal, although the tip of the

dorsum sella may show local erosion. CT scan usually shows isodense mass with obliterating or distorting the posterior suprasellar cistern[2,8,12,13]. There is usually no enhancement of the lesion following contrast administration. Before advent of CT, pneumoencephalography with midsagittal tomography in particular is a sensitive study in demonstrating these lesions[13~15]. A small 1cm lesion dose not require angiography unless there is some concern by its position that it could be an aneurysm missed on CT[7]. If the lesion is larger, or unusual, then angiography is indicated. The angiogram may show posterior displacement of the basilar artery and anterior pontomesencephalic vein, or a slight lateral displacement of the posterior communicating arteries. Abnormal tumor vessels or tumor blush is not expected. MR imaging is superior to CT in demonstrating this lesion[8,12]. On T2–weighted images, the high signal intensity of the tumor as compared to gray matter is striking and the tumor margins are sharply defined. T1–weighted images demonstrated a lesion isointense to gray matter. The present case showed same MR findings as mentioned above. MR imaging will undoubtly become the imaging modality of choice in the evaluation of precocious puberty and detection of hypothalamic hamartoma.

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