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CT and MR Findings of a Chordoid Meningioma: A Case Report[1]

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A 25-year-old woman with focal seizure, intermittent morning headache and vomiting for 2 years showed microcytic hypochromic anemia on peripheral blood smear and a 6 × 6 × 7.5 cm sized intracranial mass with cystic and solid portions at the right temporoparietal convexity on brain CT and MRI which was hypervascular on cerebral angiography. Histopathologic findings on light microscopy suggested chordoma, but it was confirmed as a chordoid meningioma by immunohistochemical study. The present case suggests that the diagnosis of chordoid meningioma shoud be considered in a juvenile or young adult who is presented with an extra-axial mass with typical location of meningiomas, findings of chordomas on light microscopy, and clinical findings of Castleman syndrome.

Index Words : Brain neoplasms, CT
Brain neoplasms, MR

INTRODUCTION

Meningioma is the most common nonglial primary brain tumor. The peak age is between 40 and 60 years of age, and only 1% to 2% of all meningiomas occur in children less than 16 years of age.

Kepes et al(1) reported 7 cases of unusual variant of meningioma, which morphologically appeared similar to chordoma and Glasier(2) reported a case of similar tumor in a 15-year-old patient. We report the radiologic findings of a chordoid meningioma, which is associated with lymphoplasma infiltrates and is sometimes associated with Castleman syndrome in young adult.

CASE REPORT

This 25-year-old woman was admitted to our hospital because of seizure, intermittent morning headache and vomiting for 2 years. Morning headache and vomiting were progressed with time. She also had tingling and cold sensation at left arm. Neurologic examination was normal. On CBC, hemoglobin(Hb) was 9.4 g/dl. Peripheral blood smear showed microcytic hypochromic anemia and neutrophilic leukocytosis suggesting iron deficiency anemia or inflammation. Total protein was 7.4 g/dl and albumin was 3.5 g/dl. On immunoelectrophoresis, arcs of IgG, A and M were increased, and polyclonality was observed. On the basis of these findings, polyclonal gammopathy due to tumor necrosis or inflammation was suspected.

On non-enhanced CT(Fig. 1), a 6 × 6 × 7.5 cm sized mass was noted at the right temporo-parietal convexity. Anterior one fourth of the mass was cystic and the remaining portion was solid with central necrotic area showing fluid-fluid level(dependent portion was more dense). Thickness of the solid portion was relatively uneven. The mass was relatively well demarcated with surrounding edema. Mass effect was also marked, showing compression of the right lateral ventricle, subfalcial herniation and partial obliteration of the right ambient cistern. On contrast-enhanced CT, a part of solid portion was slightly and inhomogenously enhanced. The demarcation between the mass and the brain parenchyme was better delineated.

On MRI(Fig. 2), anterior cystic portion revealed high signal intensity on long TR images and low signal int-

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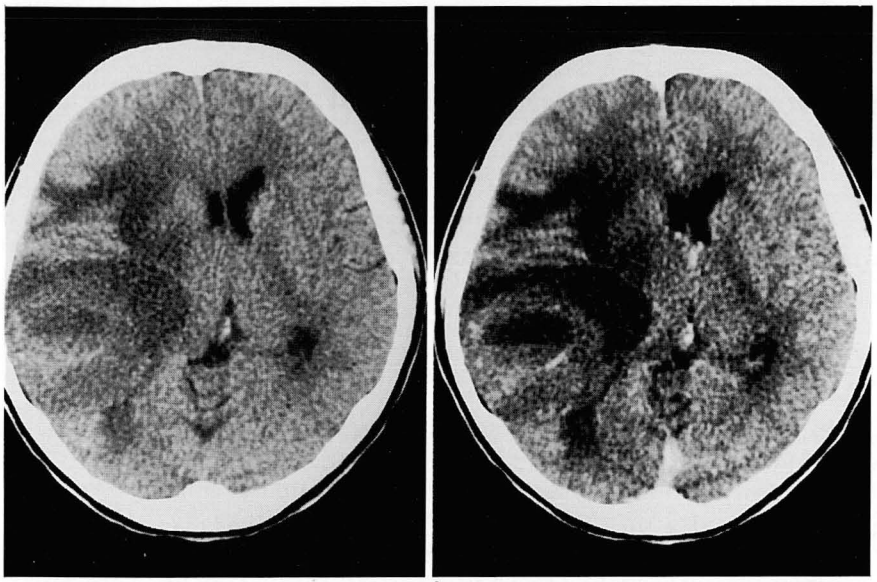

Fig. 1. CT findings.
a. Precontrast axial CT scan shows a huge mass with anterior cystic portion and posterior solid portion containing central fluid-fluid level in the right temporoparietal area. Surrounding edema is seen.
b. Postcontrast CT scan shows mild peripheral enhancement.

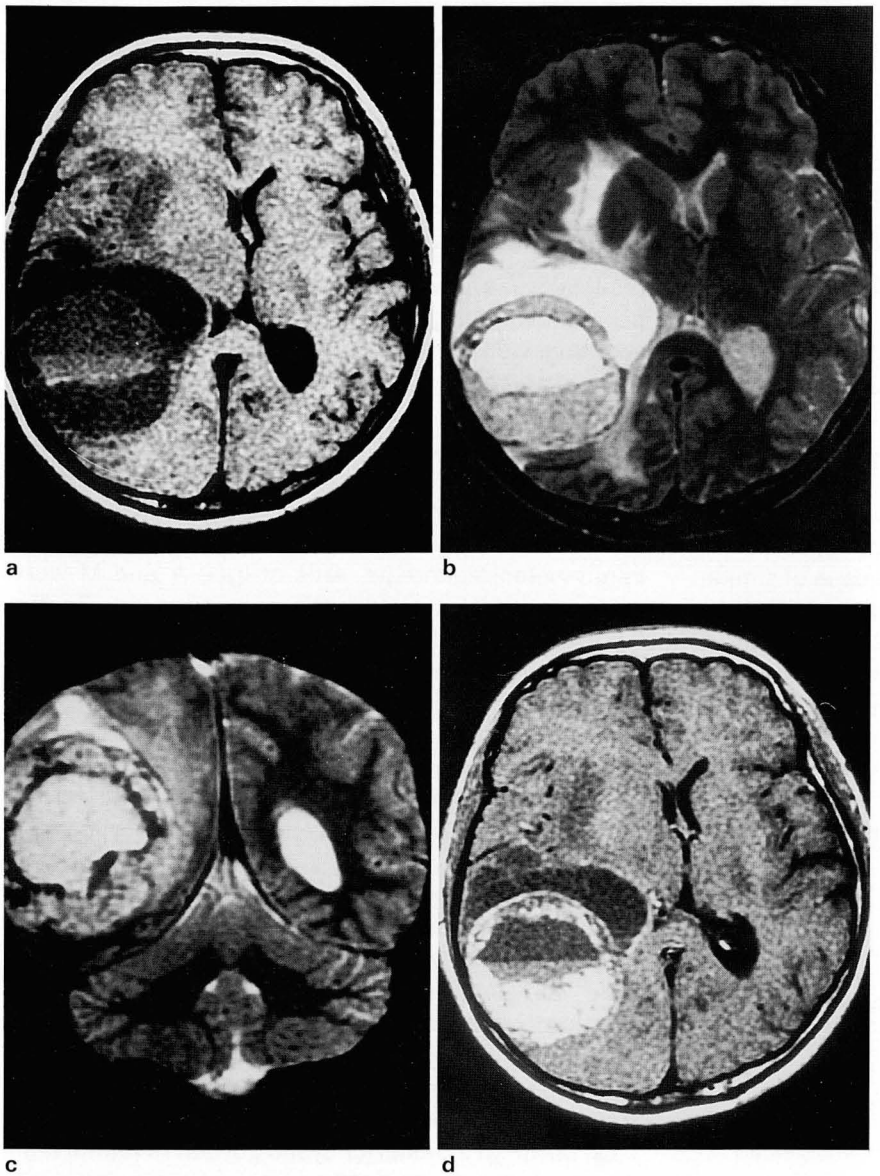

Fig. 2. MR findings.
a. T1-weighted image(300/15). Anterior portion of the mass reveals marked hypointensity. Posterior portion shows mild hypointensity with a central fluid-fluid level.
b. T2-weighted image(2200/90). Anterior portion of the mass reveals marked hyperintensity(secondary cyst formation). Solid portion of the mass shows slightly hyperintensity with a central fluid-fluid level.
c. Coronal T2-weighted image(2200/90) demonstrates multiple signal voids suggesting vascular structures.
d. Contrast enhanced axial image(300/15) shows a strong enhancement at the solid mass.

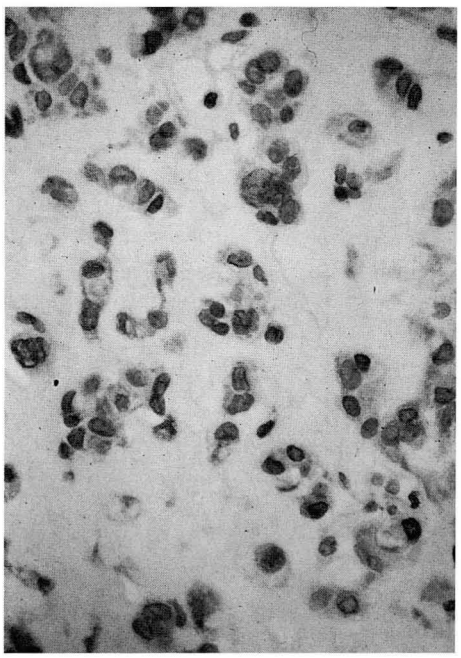

Fig. 3. Immunohistochemical study.
The cytoplasm of tumor cells with vacuoles stains positively for vimentin(immunoperoxidase stain for vimentin, ×200)

ensity on T1-weighted image. Several septations were also seen. Solid portion was mildly hyperintense on T2-weighted image and mildly hypointense on T1-weighted image compared with that of the gray matter. Multiple vascular signal voids suggesting hypervascularity were also seen.

The upper supernatant portion of the central necrotic area of the solid mass showed slightly low intensity on T1-weighted image and very high intensity on long TR images. The dependent portion was isointense on T1-weighted image and hyperintense on T2-weighted image. Thin rim of low intensity was seen around the tumor margin on all sequences, particularly more clearly visible around the solid portion. Gd-enhanced T1-weighted image showed sand-and-pepper like strong enhancement at the solid portion.

Angiography revealed hypervascularity supplied by the right middle meningeal artery and tumor staining.

At surgery, the anterior cystic portion was aspirated at first, and the fluid was the appearance of CSF. Solid tumor mass was well marginated, and hemorrhagic fluid was aspirated at the center of the mass. After removal of the mass, a clear outer surface of the cortex was seen, and the mass was revealed as a complete extraaxial mass.

Postoperative follow-up Hb level was 13 g/dl. Histopathologiclogic examination revealed irregular cords and nests composed of tumor cells, having pleomorphic nuclei with intra or extracellular vacuoles in myxoid stroma by light microscopy. Profound lymphoplasma infiltration surrounding blood vessels was noted around the tumor. Immunohistochemically, the cytoplasm of tumor cells with vacuoles was stained positively for vimentin(Fig. 3) but was negative for cytokeratin.

DISCUSSION

Chordoid meningiomas typically occur at the juvenile age. The present case is slightly older than the previously reported ones.

Determination of exact location of the mass, whether intra- or extraaxial, was difficult to evaluate on CT and MRI. Although the site of the tumor in the present case was the temporoparietal convexity, the tentorium or falx is the usual site(1, 2).

Fatty degeneration and hemorrhage are unusual features in the meningiomas(3, 4). Cystic meningiomas are also uncommon variants. Meningiomas associated with cysts were classified into five types by Worthington et al(5). Wasenko et al(6) divided the cystic component into three groups in practical value to the neurosurgeon. They may be (a)intratumoral cyst, (b) trapped CSF, (c)intraparenchymal cyst. In this present case, the anterior cystic portion of the mass was proved to be CSF loculation trapped between the tumor and brain. The mechanism of extratumoral cyst formation is not fully understood. The cyst may form as the result of a ball valve mechanism with the gradual accumulation of CSF in several sulci between the enlarging tumor and brain parenchyma(6).

In light microscopy of the present case, the histologic finding was of chordoma, showing myxoid degenerations of the intercellular substance and the clustering of cells against this mucoid background with many tumor cells displaying single or multiple intracytoplasmic vacuoles. However, the location of the mass was not of the chordoma. Immunohistochemically, the cystoplasm of tumor cells with vacuoles was stained positively for vimentin, but not for cytokeratin which is characteristically stained in chordomas.

On histopathologic study, our case showed lymphocytic-plasmacellular infiltration around the tumor. The patient preoperatively manifested hypochromic microcytic anemia and dysgammaglobulinemia. After the mass was removed, the blood picture was normalized. It appears that the peritumoral lymphoplasmacellular infiltrates, which may be regarded as reactive rather than primary cell proliferations, had brought about the type of systemic manifestations known as the Castleman syndrome(1, 7). Glasier et al (2) reported a case of chordoid meningioma without the findings of Castleman syndrome, but they did not describe whether or not there was the lymphocyticplasmacellular infiltrate around the mass.

The diagnosis of chordoid meningioma should be considered when an extraaxial mass is located compatible to that of meningioma with findings similar to chordoma on light microscopy and clinical findings of

the Castleman syndrome in a juvenile or young adult.

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척삭종양 수막종의 CT 및 MRI 소견 : 1예 보고[1]

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약 2년간 국소적 발작과 간헐적인 두통, 구토를 주소로 내원한 25세 여자환자가 말초혈액검사상 소구성 저색소성 빈혈을 보였으며 두부 CT와 MRI검사상 두개내의 우측 측두두정부에 낭성 및 고형성분의 종괴를 보였고 혈관촬영술상 고혈관성을 보였다. 병리조직학적 검사상에서는 광학현미경상 척삭종을 의심했으나 면역조직화학적 검사상에서 척삭종양 수막종으로 확진되었다. 사춘기나 젊은 성인에서 임상적으로 Castleman 증후군을 보이며 영상학적으로 수막종의 소견을 보이며 광학현미경상으로 척삭종의 소견을 보일 때 척삭종양 수막종을 진단할 수 있으리라 생각된다.