Chordoid glioma: CT and MR features

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A 50-year-old man with progressive headache and memory impairment was found to have a mass in the anterior third ventricle. Computed Tomography (CT) of the brain showed a hyperdense ovoid-shaped lesion in the suprasellar-hypothalamic region. Magnetic Resonance Imaging (MRI) revealed an intensely enhancing mass with an eccentric cyst in the anterior third ventricle near midline. Total resection was performed under the impression of third ventricle tumor, and chordoid gliomas was diagnosed histopathologically. To our knowledge, this is the first case of chordoid glioma reported in Taiwan literature.

Key words: Brain neoplasm; Brain, CT; Brain, MRI

The third ventricle chordoid glioma is pathologically characterized by a mixture of ependymal, piloid-like astrocytic feature and a myxoid stroma similar to myxopapillary ependymoma [1, 2]. It is an extremely rare tumor. In the literature, a variety of synonyms have been reported for this rare entity such as chordoid glioma [3-8], chordoid ependymoma of the lamina terminalis area [9], and tanycytoma [1, 2]. Here, we report the clinical presentation, radiological and pathologic features of the chordoid glioma in the anterior third ventricle. The differentiation of adult third ventricle tumors is discussed.

CASE REPORT

A 50-year-old man presented with headache and recent memory impairment, which was gradually for 3 years. The physical examination and clinical laboratory data including hematology and endocrine were remarkable. Computed tomography (CT) of brain demonstrated a well-circumscribed mass in the hypothalamic-suprasellar region on the pre-contrast scan (Fig. 1). The mass enhanced homogeneously after administration of contrast medium. The magnetic resonance

Figure 1. Unenhanced axial CT scan reveals a hyperdense mass (arrow) in the suprasellar-hypothalamic region.
image (MRI) showed a mainly solid mass with an eccentric cystic component in the anterior part of third ventricle. The lesion showed hypointense on T1-weighted, and hyperintense on T2-weighted image. The perifocal edema was present. After injection of Gadolinium, the lesion showed intense enhancement (Fig. 2a, 2b, 2c). A multi-voxel proton MR spectroscopy showed elevated choline peak intensity relative to creatine peak intensity in the solid part of the lesion (Fig. 2d). Total resection of the lesion was successfully performed. The histopathology showed a picture of cords and clusters of epithelioid cells within an abundant mucinous and loosely arranged vacuolated background of cells. In the immunohistochemical study. Most of the tumor cells revealed diffuse expression of vimentin and glial fibrillary acidic protein (Fig. 3). Furthermore, the vast majority of tumors showed focal coexpression of cytokeratins. According to the neuroimaging and pathological findings, a diagnosis of chordoid glioma of anterior portion of third ventricle was made.

The patient recovered well without significant neurological deficit after surgery.

Figure 2. The MR and MR spectroscopic findings in (a-d). Note the location of the mass in the anterior portion of third ventricle and intense enhancement. a. Axial T1-weighted SE (TR/TE: 350/30ms) MR images shows the solid component consisting of inhomogeneous signal intensity. b. Axial T2-weighted fast SE (1400/663ms) MR image reveals inhomogeneous signal intensity of the solid component with perifocal edema shown as hyperintensity around the mass. Note a hyperintense area adjacent to the mass, suggesting an eccentric cyst (arrow). c. Contrast-enhanced coronal T1-weighted SE (309/24ms) MR image shows homogeneous enhancement of the solid component without causing downward displacement on the optic chiasm. d. Representative transverse T1-weighted (344/20ms) MR image, with the chosen volume of interest for solid part of tumor and its measurement reveals a characteristic marked increasing in the amount of choline.
DISCUSSION

The term chordoid glioma of the third ventricle is first used to describe a rare and slowly growing neoplasm of uncertain histology, with chordoid appearance, occurring preferentially in middle-aged women [10]. Less than fifty cases have been documented in the literature and the epidemiology is not yet clear. The chordoid glioma in the anterior third ventricle could be better classified as chordoid ependymoma of the lamina terminalis area [9]. It has been named as tanycytoma [2] because of the tumor arising from the lamina terminalis, infundibular recess and median eminence, which were rich in tanyctye cells [1]. The tanyctic cells are generally seen in the primitive nervous system instead of the mature ependymal cells [11].

The unique histological characteristics of the chordoid glioma include irregular cords and cluster of eosinophilic-cytoplasm epithelioid cells within an abundant mucinous and often vacuolated background microscopically [7, 9]. The mass also has diffuse expression of vimentin, glial fibrillary acidic protein (GFAP) and focal co-expression of cytokeratins without presence of MIB-1 labeling immunohistochemically [4, 5, 9]. Microvilli were seen frequently with moderately developed intermediate junctions ultrastructurally [1]. Taken these findings into account, the histological findings of the tumor may resemble those of chordoid chordoma and chordoid menigioma microscopically, and the tumor could be differentiated by positive stain of GFAP immunohistochemically [12].

The distinctive imaging features of chordoid glioma are the typical suprasellar-hypothalamic location, ovoid shape, hyperdensity on non-contrast CT scan, and uniform intense enhancement after contrast administration of the tumor [10]. MR image allows the evaluation of tumor content, the exact location of tumor, and adjacent parenchyma involvement [2]. The high choline peak in our case on MR spectroscopy might reflect high grade of glioma or maybe normal in the ependymoma group [13].

Tumors involving the third ventricle can arise either intrinsically or extrinsically [10]. The differential diagnoses of an enhancing anterior third ventricle mass with eccentric cystic lesion in adult include chordoid glioma, menigioma, germ cell tumor, optic-hypothalamic pilocytic astrocytoma, and craniopharyngioma [2]. Meningioma and germinoma are rare in the anterior portion of third ventricle, but could present as hyperdense mass on non-contrast CT study. The pilocytic astrocytoma could be differentiated from chordoid glioma by the attenuation of the mass which is usually low in the former. Craniopharyngiomas are classified into the adamantinomatous and papillary types. The papillary type of craniopharyngiomas has solid and cystic components just like our case. But in the adamantinomatous type, the cystic component is hyperintense on both T1- and T2-weighted images due to proteinecious content. In addition, one can differentiate chordoid glioma from craniopharyngioma by the location of tumor epicenter.

In conclusion, chordoid glioma is a rare tumor with some indolent symptoms, Although this tumor is assigned as WHO grade II [2] because of benign

Figure 3. Histology of chordoid glioma in the anterior third ventricle. a. Photomicrograph shows large epithelioid tumor cells with abundant eosinophilic cytoplasm (arrow). The background consists of loosely arranged vacuolated cells (arrowhead). (HE stain, x400). b. Glial fibrillary acidic protein stain of chordoid glioma tumor cells shows strong, diffuse, cytoplasmic immunoreactivity.
pattern of the histological findings, complete resection is requested to avoid subsequent complications such as hydrocephalus and endocrine imbalance. The radiologist is often the first to suggest the diagnosis of chordoid glioma because of the characteristics of non-contrast CT finding. MRI may be indicated to identify the exact location and extension of the tumor.

REFERENCES

Chordoid glioma: 電腦斷層和磁振造影的影像表現

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我們報告一位五十歲男性有逐漸嚴重的頭痛和記憶喪失，診斷為chordoid glioma。頭部的電腦斷層表現出在蝶鞍上-腦下垂體的位置有一叢型高衰減值的腫塊。磁振造影則顯示此腫塊注射顯影劑後有高度顯影且位在第三腦室，腫瘤周圍有一個水囊，符合chordoid glioma的影像學特徵。手術後，組織病理學證實是chordoid glioma。依據我們所知，這是台灣文獻中有關chordoid glioma的第一篇個案報告。

關鍵詞：腦瘤，腦部，電腦斷層，腦部，磁振造影