Chordoma in the Sella Turcica
—Case Report—

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Abstract

A 75-year-old man presented with a rare case of chordoma in the sella turcica of the skull base. He had been treated for hypertension and chronic renal failure since 1990. Computed tomography detected a tumor in the sella turcica in 1994, but the patient had no clinical complaints and the serum pituitary hormone levels were normal. He died of disseminated intravascular coagulation, myocardial infarction, pulmonary infection, and multiple cerebral infarctions in 2000. At autopsy, the tumor in the sella turcica was 3.1 cm in greatest diameter and had compressed the pituitary gland posteriorly. Histological examination found oval cells and vacuolated short spindle-shaped cells which showed morphological changes similar to myxoma cells. The tumor was lobulated by narrow connective tissues. The tumor did not contain any cartilaginous tissue components, and was stained positively for epithelial membrane antigen but negatively for S-100 protein. The final diagnosis was chordoma. There was no association between the tumor and the cause of death.

Key words: intrasellar chordoma, skull base, autopsy

Introduction

Chordomas are tumors that are thought to originate from notochordal remnants and account for about 1% of all malignant bone tumors and 0.1–0.2% of intracranial tumors. Chordomas develop in the sacrococcygeal region in 49% of cases, in the clivus region of the skull base in 36%, and in the vertebrae in 15%, but chordomas rarely form in the sellar region. Chordomas are prevalent in middle-aged or older men. Here, we report a rare case of chordoma located in the sella turcica of the skull base.

Case Report

A 75-year-old man had been treated in another hospital for hypertension, chronic cardiac failure, and chronic renal failure since 1990. Computed tomography (CT) found an intrasellar tumor with a diameter of 2 cm in 1994. The tumor boundary was slightly obscure. The tumor appeared in homogeneous without calcifications. The tumor had expanded the sella turcica and partially destroyed the sella turcica, clivus, and planum sphenoidale (Fig. 1). We suspected that the lesion was a pituitary tumor, but the patient was only followed up without treatment because he was free from complaints and his serum
pituitary hormone (growth hormone, prolactin, adrenocorticotropic hormone, thyroid-stimulating hormone, follicle-stimulating hormone, luteinizing hormone) levels were normal.

He developed low-grade fever and cough on March 25, 2000. In addition, he complained of chest discomfort, which did not improve after sublingual administration of nitroglycerine, and began to show signs of respiratory acidosis. Furthermore, he had pneumonia, which was resistant to antibiotic treatment. He complained of chest pain on May 13, 2000. Electrocardiography and echocardiography indicated myocardial infarction in the inferior wall. His urine output was decreasing and comorbid conditions such as dyspnea were worsening, so he was transferred to Osaka Medical College Hospital on May 16. He was treated with hemodialysis and antibiotics, but he died on May 21 of multiple cerebral infarctions and disseminated intravascular coagulation. Autopsy was performed 3 hours after death.

**Autopsy Findings**

The intracranial tumor (3.1 × 2.2 × 1.7 cm) in the sella turcica had compressed the pituitary gland toward the posterior direction. The surface of the tumor was predominantly smooth and was yellowish-white (Fig. 2). Histological examination of the tumor found growth of oval cells or vacuolated short spindle-shaped cells and morphological changes similar to myxoma cells (Fig. 3A). The tumor was partitioned into lobules by narrow portions of connective tissues. The diagnosis was chordoma. No apparent cartilaginous tissue components were detected. Dura mater was located between the tumor and the pituitary gland, and no direct infiltration into the brain was found (Fig. 3B). Immunohistochemistry showed that the tumor was positive for epithelial membrane antigen (EMA) (Fig. 4A) but negative for S-100 protein (Fig. 4B).

Macroscopic and microscopic findings of the other organs included signs of sepsis, pyogenic meningitis, bacterial endocarditis (mitral valve warts), pyelonephritis, and microabscesses of the liver. In addition to the multiple cerebral infarctions, acute cardiac infarction of the inferior wall, lung hemorrhage (310 and 440 g), and chronic interstitial pneumonia were also noted.

**Discussion**

Only 22 cases of intrasellar chordomas have been
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reported since 1966.6) Chordomas usually grow slowly and are frequently found as relatively large tumor masses or are detected due to the signs of tumor compression. Patients with chordomas at the skull base will have symptoms such as headaches, visual impairment, upper lid ptosis, etc. Our patient had no clinical complaints and no infiltration of the tumor beyond the dura mater was detected at autopsy, so we suspect that the tumor had grown very slowly. Among the 22 reported cases of chordomas, only our patient was free of clinical complaints caused by the tumor.6)

Neuroimaging analysis is useful for the diagnosis of chordomas. Chordomas are characterized by bone destruction on CT in peritumoral areas and also by calcifications or bone fragments within the tumors.7,8) However, chordomas vary in imaging studies.8) T₁-weighted magnetic resonance (MR) imaging of chordomas usually shows relatively low signal intensities compared to the lipid tissues of the skull base surrounding the tumors, whereas T₂-weighted imaging shows high signal intensities. Calcifications or bone tissues appear as low-intensity areas in the tumor. CT is more specific for calcified masses such as chordoma than MR imaging.3) The differential diagnosis based on imaging studies includes the following: meningioma, neurinoma, metastatic tumor, plasmacytoma, giant cell tumor, sphenoid sinus cyst, nasopharyngeal carcinoma, chondrosarcoma, pituitary adenoma, and hemangioma. In the present case, the tumor was located in the sella turcica. Although CT clearly showed bone destruction, no calcification was detected in the tumor, and the boundary between the tumor and the pituitary gland was unclear. The differential diagnosis was difficult to establish in this case prior to death.

Histologically, chordomas contain physaliphorous cells with oval nuclei and eosinophilic cytoplasms containing many vacuoles which grow in chords or sheets within the myxoid matrix. Furthermore, abundant glycogen is detected within the tumor cells. Immunohistochemical examination shows that the physaliphorous cells are positive for epithelial cell markers such as cytokeratin or EMA. Although vimentin and S-100 protein have also been found,6) staining for S-100 protein was negative in our case for unclear reasons. A lesion incorporating interstitial formation of a cartilaginous matrix in addition to the properties of chordomas is called ‘chondroid chordoma’.4) The present case showed the typical histology of chordoma and did not contain any cartilaginous components.

A tumor without the typical histology of chordoma must be distinguished from chondrosarcoma, which can be graded histologically according to the degree of cellularity, pleomorphism, and mitosis.5) Chordoma contains epithelioid tumor cells forming chords or masses on a background of myxoid matrix. The presence of abundant glycogen in physaliphorous cells is important for differentiating chordoma from chondrosarcoma. Chondrosarcomas are positive for S-100 protein but negative for other epithelial cell markers.2) Since our case was positive for EMA and negative for S-100 protein, the immunohistochemical diagnosis excluded chondrosarcoma.

The prognosis for patients with chordomas is poor, because these tumors are less radiosensitive than chondroid chordomas, local recurrences frequently take place, and remote metastases are found in about 10% of cases.4) In the present case, the chordoma was located in the sella turcica and infiltrated into the bone, but the patient had shown no related symptoms during the 6 years following diagnosis. The chordoma was not considered to be related to

Fig. 4 Photomicrographs showing immunohistochemical staining of the tumor. Staining was positive for epithelial membrane antigen (A) but negative for S-100 protein (B). A: ×200, B: ×200.

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the patient’s death.

References


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