Suprasellar Benign Chondroma: A Case Report and Literature Review

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Intracranial chondroma is a rare benign tumor comprising only 0.2% of all intracranial tumors. A 27-year-old woman presented with visual dysfunction and headache. Brain computed tomography and magnetic resonance imaging revealed a suprasellar mass lesion with a calcified component. Gross total removal was achieved via a basal interhemispheric approach. Postoperatively, visual function improved to the normal range, and no recurrence was evident 4 years later. Histopathological examination confirmed the diagnosis of benign chondroma. Preoperative differentiation of chondromas from chordomas is clinically important, because of the different treatment and prognostic implications. The only effective treatment for chondroma is total surgical removal. We present a case of gross total resection of a suprasellar chondroma with reference to the literature.

Keywords: chondroma, suprasellar tumor, basal interhemispheric approach

Introduction

Chondromas usually arise from embryonic chondrocytic cell remnants in the small bones of the hands and feet, and consist of mature hyaline cartilaginous tissue.^{1,2)} Synchondrosis joints are found between the occipital and sphenoid bones, and between the petrous portion of the temporal bone and jugular process of the occipital bone. Intracranial chondromas frequently occur at the skull base of the parasellar region, clivus or cerebello-pontine angle.^{3–9)} Intracranial chondromas are rare benign tumors comprising 0.2% of all intracranial tumors.^{5,6)} In the clinical field, a chondroma resembles a chordoma and a chondrosarcoma in terms of radiographic images and pathological features.^{5,10)} Preoperative differentiation between chondromas and other tumors is thus often difficult.^{5,11)} However, differential diagnosis is also quite important, because the prognoses can be markedly different.^{5,12,13)} Here, we report the complete surgical resection of a suprasellar chondroma via the basal interhemispheric approach and present a review of the literature.

Case Report

A 27-year-old woman presented with a 1-year history of visual disturbance. At 26 years old, she had developed nephropsis. One year later, she had experienced slight headache, and magnetic resonance imaging (MRI) revealed a suprasellar tumor. She was then referred to our hospital. On examination, visual acuity was motus manus in the right eye and temporal hemianopsia in the left eye. Other neurological findings were intact. Pituitary function was normal.

Imaging Studies

Computed tomography (CT) showed a suprasellar mass lesion with a slightly calcified component (Fig. 1a). The mass lesion was located in the midline of the suprasellar region. Bone imaging revealed osteolytic changes and bony destruction of the posterior clinoid process (Fig. 1b). MRI showed the suprasellar mass lesion was hypointense on T1-weighted images (Fig. 2a), hyperintense on T2-weighted images (Fig. 2b) and a heterogeneous-honeycomb-enhanced lesion on gadolinium contrast-enhanced MRI (Figs. 2c and 2d). The tumor was attached to the diaphragma sellae and elevated the optic chiasm (Figs. 2a and 2b).

Operation and Postoperative Course

Because this tumor containing a calcified component was located behind the optic chiasm, we selected a basal interhemispheric approach to obtain a wide operating field and remove the calcified hard tissue radically. After dissection of the interhemispheric fissure, a dark-yellowish tumor was encountered. The tumor contained a large calcified component and a smaller gelatinous component. The calcified lesion had markedly elevated the optic nerve, however could be dissected from the nerve via a wide prechiasmatic window. The pituitary stalk was identified behind the tumor capsule. Gross total removal was achieved. The tumor origin was considered to be the dura of the posterior clinoid process, and some part of the dural attachment was left in place. The postoperative course was uneventful including the pituitary function. Visual symptoms improved to the normal range. MRI at 4 years postoperatively showed no recurrence (Fig. 3).

Histopathological Findings

Histopathological examination revealed lobular proliferation of chondrocytes with homogeneous small nuclei and lacunar structures indicating chondroid differentiation against a cartilaginous matrix background (Fig. 4). Tumor cells were arranged without tight cell-to-cell adhesion of

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the epithelial type and without physaliphorous cells associated with the chordoma. Cells did not show nuclei with increased chromatin and two small nuclei accompanied by the chondrosarcoma. No nuclear atypia or pleomorphism, or evidence of mitosis were seen.^{2,3,10} Immunohistochemistry disclosed positive findings for vimentin and negative findings for cytokeratin and epithelial membrane antigen.² Based on these findings, the pathological diagnosis was a benign chondroma.

Discussion

Intracranial chondromas were first reported by Hirschfield in 1851.¹⁴ Approximately 15% of intracranial chondromas arise supratentorially from the dura mater, brain parenchyma, ventricle or parafalcine locations.^{4,6-9} Skull-base chondromas are usually divided into suprasellar, parasellar and clival types,^{1,3} which may arise from the residual primordial cartilaginous cranium, such as at the sphenopetrosal, petro-occipital and sphenopetrous synchondroses.^{1,3,4} Most skull-base chondromas are therefore located in the paramedian area.^{6,7}

In general, suprasellar tumors in adults are relatively common, such as giant pituitary adenoma, meningioma and craniopharyngioma. A midline suprasellar chondroma is a rare benign tumor among the primary intracranial tumors, with an estimated incidence rate of 0.2%, and is difficult to

preoperatively distinguish from other suprasellar tumors, such as chordomas, chondrosarcomas and craniopharyngiomas.^{2,5,10,11} CT reveals a lobularly circumscribed mass with regional calcification accompanied by erosion and destruction of the surrounding bone structures.^{6,7,15} Coronal CT with a bone window shows multiple small circular and curvilinear calcifications occurring in the myxochondroid matrix. Calcifications are recognized as irregular shapes in 60%, with

Fig. 3 Postoperative magnetic resonance imaging (MRI) shows successful removal. (a) Axial contrast-enhanced T1-weighted MRI. (b) Sagittal contrast-enhanced T1-weighted MRI.

Fig. 1 Computed tomography (CT). (a) Axial CT shows a suprasellar, isodense mass lesion with a calcified component. (b) Bone window sagittal CT reveals a bone-density component in the suprasellar portion.

Fig. 4 Histopathological findings (hematoxylin and eosin × 100). The tumor against the myxoid matrix background comprises cellular hyaline cartilage with irregular lobules, spindle cells and chondrocytes with homogeneous small nuclei.

Fig. 2 Magnetic resonance imaging (MRI). (a) Coronal T1-weighted MRI shows a suprasellar hypointense mass lesion elevating the optic chiasm. (b) Coronal T2-weighted MRI reveals hyperintensity. (c) Axial contrast-enhanced T1-weighted MRI shows a heterogeneously enhanced mass compressing the posterior part of bilateral cavernous sinuses. (d) Sagittal contrast-enhanced T1-weighted MRI shows a suprasellar, heterogeneously enhanced mass extending to the interpeduncular cistern and elevating the optic chiasm.

local bone destruction in 40%.^{2,6,7)} Chordomas have been reported to comprise 40–50% of lesions showing calcifications and 50–60% of those with bone destruction.^{5,10)} Similar calcified findings are also seen in patients with meningiomas or craniopharyngiomas. Bone destruction is often identified in both chondromas and chordomas, but less frequently than with chondromas.⁶⁾ Clinical symptoms in patients with suprasellar chondromas are always associated with visual dysfunction.^{1,15,16)} Differential diagnosis between skull-base chordomas and low-grade chondrosarcomas could be quite important. Chordomas also locally invade to the midline and parasellar skull base with similar clinical and radiological characteristics, given their origin from embryonic notochord remnants in the midline.^{5,10,11)} Pathological diagnosis is also very important, because the treatment strategies and prognoses differ.^{12,13)}

In patients with suprasellar benign chondromas, the only way to achieve a cure is to remove the tumor totally. No recurrence is expected following complete resection of the tumor, and the long-term prognosis is acceptable.^{14,17–19)} Past reports on surgical operations have described various approaches, including the trans-sphenoidal approach, trans-basal interhemispheric approach, extra-dural temporopolar approach, and anterior trans-petrosal approach.^{12,14,19)} In this case, we chose the basal interhemispheric approach to obtain a wide operating field surrounding the prechiasmatic window and to remove the calcified component behind the optic chiasm, although endoscopic transnasal surgery has recently been developed as an alternative surgical approach. The progression-free survival rate has been reported as 75% at 5 years and the overall survival rate as 93%.¹⁹⁾ In addition, visual dysfunction could be expected to improve after surgical resection. On the other hand, the 5-year overall survival rate for a patient with a chordoma is only 50–60%, because total removal of the tumor is difficult and tumor recurrence is frequent.^{5,10)} Radiation therapy is ineffective and malignant transformation to chondrosarcoma has been reported after partial removal.^{2,19)} Radiation therapy is therefore not recommended.

In conclusion, a suprasellar chondroma is a rare benign intracranial neoplasm. Differential diagnosis is quite important because of the possibility of clinically similar suprasellar neoplasms with different prognoses. Surgical resection is the treatment of choice. In our case, gross total removal was

achieved via a basal interhemispheric approach, followed by dramatic improvements in visual symptoms.

Conflicts of Interest Disclosure

The authors have no conflicts of interest to declare.

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