

Pituitary Chondrosarcoma presenting as a sellar and suprasellar mass with parasellar extension: An Unusual presentation

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KEY WORDS

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ABSTRACT

Chondrosarcoma is a mesenchymal tumor composed of tumor cells producing cartilage. It is more common in older age and often affects the axial skeleton. We report a rare case of chondrosarcoma mimicking a sellar and suprasellar mass with parasellar extension. A 40 yr woman presented with decreasing visual acuity and headache. Magnetic resonance (MR) image revealed a cystic sellar and suprasellar mass with parasellar extension showing mild enhancing solid component. It favored the diagnosis of craniopharyngioma. The patient underwent trans-sphenoidal partial resection of the tumor resulting in removal of the sellar mass. However, the suprasellar mass could not be excised completely due to limited surgical field. The pathologic diagnosis was chondrosarcoma. Eight months after the operation, pterional approach was performed to remove the remaining mass. Intraoperative findings confirmed that the mass originated from dorsum sellae.

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Introduction

Most sellar and parasellar masses are of pituitary origin, only around 10% are non-pituitary. The differential diagnosis of non-pituitary sellar masses includes benign and malignant neoplasms, vascular, granulomatous and inflammatory lesions (1). Nearly 10% of such masses are cartilaginous tumors originating from the skull base; chordomas are more frequent than chondrosarcomas (2). "Chondrosarcoma accounts for 0.15% of all intracranial tumors. They are sub classified into the conventional (hyaline/myxoid), dedifferentiated, clear cell, and mesenchymal subtypes. In accordance with the current literature, the conventional type of

chondrosarcoma is the most common cartilage tumor to develop in the brain "(3).

However, the prognosis of these tumors is determined primarily by WHO histological grade. Chondrosarcoma is mostly divided into three histological grades, grade I (well differentiated), grade II (moderately differentiated), and grade III (poorly differentiated). The 5-yr survival rates for grade I, II and III chondrosarcomas of bone from all body sites are 90%, 81%, and 43%, respectively (4). Chondrosarcoma, which invades clivus or petrous bone, is not only difficult to access surgically but also hard to remove completely (5). Chondrosarcomas are frequently misdiagnosed as chordomas, which have a different prognosis. Differential diagnosis is very

important because, when treated with similar aggressive treatment strategies, chondrosarcoma has a much better prognosis than chordoma (6).

There have been many theories postulated about the occurrence of intracranial chondrosarcomas. Although most chondrosarcoma arise de novo, they are common in patients with Ollier's disease, Maffucci syndrome, Paget's disease and osteochondroma. One postulated theory for the development of intracranial chondrosarcoma highlights that while the bones of the skull vault develop primarily by intramembranous ossification, the bones of the skull base mature predominantly by endochondral ossification. Endochondral ossification is also responsible for the development of several sites in the mature skull including a large part of the petrous portion of the temporal bone, areas of the petro-occipital, sphenoccipital, and sphenopetrosal.

Synchondrosis Intracranial chondrosarcomas might develop from the chondrocytes within rests of endochondral cartilage that may be present in these areas. The primitive multifunctional mesenchymal cells involved in the embryogenesis of the skull base and temporal bone may be the source of these tumors. Intracranial chondrosarcomas develop from metaplastic mature fibroblasts (3, 4).

Although malignant tumors of sella are rare, they should be considered in the differential diagnosis of sellar tumors typified by pituitary adenoma, and chondrosarcoma is one possible candidate (7). We observed a rare case of chondrosarcoma originating from dorsum sellae, clinically mimicking the characteristics of craniopharyngioma or pituitary tumor.

Case-Report

A 40 yr female was admitted for evaluation and management of sellar and suprasellar mass with parasellar extension. The patient gave a 1-yr history of intermittent headaches and blurring of vision. Physical examination revealed bilateral

temporal hemianopia, reduction in visual acuity, and early optic atrophy. No evidence of diplopia, any cranial nerve palsy or galactorrhea was noted. Investigations showed normal serum thyroid stimulating hormone (TSH), luteinizing hormone (LH), cortisone, adenocorticotrophic hormone (ACTH), follicle-stimulating hormone (FSH) and prolactin (PRL). Magnetic resonance imaging (MRI) showed a large irregular hypo intense to cystic mass in sellar, suprasellar and right parasellar region measuring 3.5X3.0 cms with thick enhancing capsule and mild intrinsic enhancing solid component. Mass is encasing the pituitary gland and indenting the right medial temporal lobe. Optic chiasma was minimally compressed with possible destruction of roof and right lateral wall of right sphenoid sinus favoring the diagnosis of craniopharyngioma. (Fig. 1, A-B).

The patient underwent trans-sphenoidal partial resection resulting in removal of the sellar mass. Suprasellar mass could not be excised completely due to the limited surgical field. Histopathology revealed fragments of predominantly hyaline and focal myxoid cartilage. Cartilage was lobulated and cellular with accentuation of cellularity at its periphery. Chondrocytes showed hyperchromasia, binucleation and multinucleation with vacuolated cytoplasm. Cohesive nests and cords typical of chordoma with physaliferous cells were absent. The diagnosis of conventional chondrosarcoma, Grade 2/3 was made (Fig. 2, A-B). Immunohistochemistry showed negative staining for pancytokeratin and epithelial membrane antigen (Fig. 2, C-D), which was consistent with a grade 2 chondrosarcoma. Eight months later, a follow-up MRI showed that the suprasellar portion of tumor remained. Visual acuity showed slight improvement. The remaining mass was removed by pterional approach. Intra-operatively, the tumor displaced the pituitary gland laterally and the tumor was easily distinguished from pituitary gland. The histopathology revealed the same diagnosis of a grade 2 chondrosarcoma.

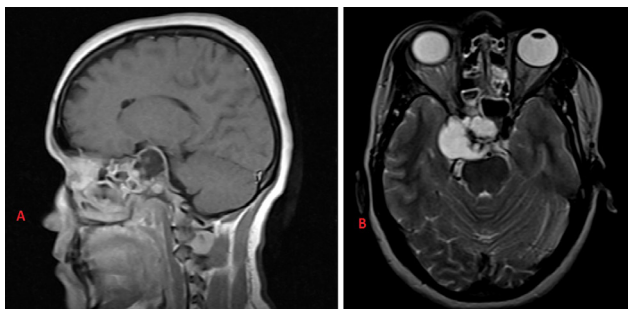


Fig. 1

MRI Brain showing a large irregular hypointense to cystic mass in supra and right parasellar region measuring 3.5x3.0 cm, with a thick enhancing capsule along with mild enhancing solid component.

(10, 11). Most patients suffer from visual problems mainly diplopia and headache. Most of the patients complaints are diplopia with impaired eyeball movement (51%) followed by headache (31%), but only 14% had decreased visual function. Less common presentations include hearing loss, dizziness, tinnitis, facial sensory disturbance, ataxia, and hemiparesis (10). Radiological examination usually shows bone destruction and variable calcification on CT imaging, involvement of neural and vascular structures on MRI, and mostly hypovascularity on angiography. Information on the endocrine

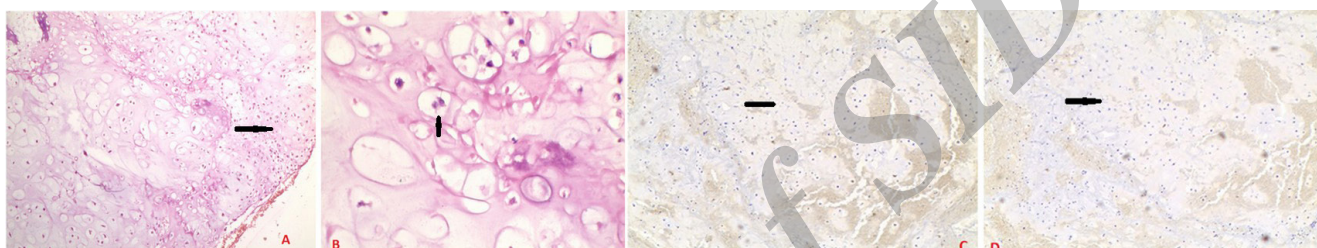


Fig. 2

(A): H&E section showing lobulated areas of cartilage with increase cellularity at the periphery (arrow). (100X), (B): H&E section showing chondrocytes exhibiting hyperchromasia, binucleation, multinucleation and vacuolated cytoplasm (arrow). (400X), (C): Immunohistochemistry showing negative staining for pancytokeratin (arrow). (100X), (D): Immunohistochemistry showing negative staining for epithelial membrane antigen (arrow). (100x)

Discussion

Most sellar lesions are either functioning or nonfunctioning pituitary adenomas. The tumors arising in this region may cause diagnostic dilemmas, as cartilaginous tumors can also occur in these locations rarely. In a previous series of 911 sellar masses, only 83 were of nonpituitary origin, and of these 11% were cartilaginous, mainly chordomas (8). Intracranial chondrosarcoma are even rarer, with only a few cases in the literature (1). Chondrosarcomas are malignant tumors of cartilage-forming cells that occur mainly in the axial part of the skeleton. These represent less than 5% of skull base tumors with ~75% arising in the parasellar region (9). Most common areas involved are petrosal bone, occipital bone, clivus, and sphenoid bone

function of patients presenting with such tumors is even sparser (1). The patient in this case was a 40 yr female admitted for evaluation and management of sellar and suprasellar mass with parasellar extension. There was a 1-yr history of intermittent headaches and blurring of vision. Physical examination revealed bilateral temporal hemianopia, reduction in visual acuity, and early optic atrophy. No evidence of diplopia, any cranial nerve palsy or galactorrhea was noted. Endocrinal investigations were normal. MRI showed a large irregular hypo intense to cystic mass in sellar, suprasellar and right parasellar region with thick enhancing capsule and mild intrinsic enhancing solid component. Mass was encasing the pituitary gland and indenting the right medial temporal lobe. Optic chiasma was minimally compressed with possible destruction

of roof and right lateral wall of right sphenoid sinus favoring the diagnosis of craniopharyngioma.

The main differential for chondrosarcoma is chordoma. They can be distinguished by histopathology and immunohistochemistry (IHC), the prognosis of chondrosarcomas is more favorable than chordomas, with even better results on longer follow-up (4-6, 11, 12). Therefore, it is imperative to distinguish between both these entities to plan further treatment. In Histopathology of chordoma, there are present cords and nests of typical physaliferous cells.

On IHC, chondrosarcoma and chordoma cells are positive for S-100 protein; chondrosarcoma cells, however, are negative for pan-cytokeratin markers and epithelial membrane antigens, unlike chordoma cells, which are positive for both (13, 14). Patient in this case was diagnosed as chondrosarcoma on histopathology. Immunohistochemistry showed negative staining for pancytokeratin and epithelial membrane antigen, which was consistent with a grade 2 chondrosarcoma.

Chondrosarcoma is classified histologically from I to III of Evan's classification by nuclear size, differentiation and nuclear pleomorphism. Chances of recurrence increase with the grade of the tumor (15). The grade of the tumor in this case was II.

Chondrosarcomas of the skull base are usually slow growing low-grade malignancies that rarely metastasize outside the skull but expand locally to compress adjacent structures (1). Most important predictor of long-term outcome is the extent of resection of the initial tumor. Local recurrence rates of 53% have been reported. In 80% of recurrent cases, the initial resection was subtotal. However, the anatomical location of the tumors renders complete resection extremely difficult and hazardous to achieve. So, an adjuvant treatment such as postoperative radiotherapy must be considered. The conventional radiotherapy does not seem to be effective as chondrosarcomas are relatively radio-resistant (1). Results that are

more promising have been obtained with the use of proton beam irradiation following surgical resection. However, treatment-related visual impairment or pituitary insufficiency has been documented. Using this as adjuvant treatment, 5- and 10-yr local control rates of 99% and 98%, disease-specific survival rates of 99%, and 3-year local control rates of 94%, have been achieved (1,16). This patient underwent trans-sphenoidal partial resection resulting in removal of the sellar mass. Suprasellar mass could not be excised completely due to the limited surgical field. Eight months later, a follow-up MRI showed that the suprasellar portion of tumor remained. Visual acuity showed slight improvement. The remaining mass was removed by pterional approach.

Many of the previous studies have reported these tumors to be well differentiated (1, 2) but in our case, chondrosarcoma was classified as moderately differentiated (grade 2/3) according to the histological features.

The prognosis of patients with intracranial chondrosarcoma is strongly influenced by several significant factors. These factors include the use of postoperative adjuvant radiation therapy, pathological pattern, previous treatment (surgery or radiation therapy) and extent of tumor removal. However, local recurrence is considered by many to be the most significant predictor of mortality in these patients. Chondrosarcoma patients treated with surgery alone demonstrate a higher 5- year mortality rate, while the implementation of postoperative adjuvant radiation therapy reduces this mortality rate dramatically. Meticulous surgical resection together with postoperative radiotherapy provides the best long-term outcomes for these patients (3).

Stereotactic radiosurgery uses single high dose of concentrated radiation delivered with great precision due to stereotactic mapping and sophisticated radiation technology. It ensures that at the perimeter of the stereotactically mapped target the dose falls rapidly, thereby

sparing adjacent normal structures from such high doses. It has been used to treat skull base chondrosarcoma and seems to be of particular benefit when the irradiated tumor volume is small. Chemotherapy is not an effective form of adjuvant treatment for chondrosarcomas (1, 17).

Conclusion

This case illustrates that chondrosarcoma, a very unusual tumor to arise in the sella, can exhibit clinical, endocrinological, and radiological features similar to those of the more commonly encountered lesions in this region. Chondrosarcoma, chordoma, craniopharyngioma and pituitary tumors are very different from one another in surgical approach, treatment, and prognosis. Therefore chondrosarcoma should be included for differential diagnosis of sellar and suprasellar tumors. The unusual consistency of the tumor should alert the neurosurgeon to perform as near-complete resection as possible, because this is the most important predictor of outcome. Regular and prolonged follow-up is necessary to monitor further tumor recurrence.

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Conflict of interest

The authors declare that there is no conflict of interests.

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