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CASE REPORT

Sphenoidal Intracranial Chondrosarcoma

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ABSTRACT

A 40-year-old male presented with headache, complete loss of vision in the right eye and decreased vision of the left eye. On examination, fundoscopy revealed secondary optic atrophy in the right eye and pale temporal disc in the left eye. MRI brain revealed a soft tissue mass arising from sphenoid sinus, eroding the overlying bone and extending into the sellar and parasellar regions up to the third ventricle. The MRI scan favoured the diagnosis of craniopharyngioma, most likely, while CT scan showed sellar floor erosion and calcification which favoured a pituitary lesion. The mass was also compressing the optic apparatus. The patient underwent excision biopsy through subfrontal approach. The histopathology report revealed it to be a chondrosarcoma. Chondrosarcoma is a rare malignant mesenchymal tumour of cartilage-producing cells; only 7% tend to involve craniocervical region. Maximal surgical resection followed by proton radiation therapy have clinically shown promising outcome.

Key Words: Chondrosarcoma. Skull base. Intracranial. Sphenoidal.

INTRODUCTION

Chondrosarcoma is a rare malignant mesenchymal tumour of cartilage-producing cells. Only 7% of all the chondrosarcoma are of neurosurgical concerns. Furthermore, only less than 0.16% of all the brain tumours comprise of primary intracranial chondrosarcoma. Skull base is most often involved.2,3 Choroid plexus, dura matter and brain parenchyma are extremely rarely invaded.1 Surgical resection has found to be the preferred treatment modality. Maximal surgical resection followed by proton radiation therapy have clinically shown promising outcome.3-5 We present a case of 45-year-old male with a chondrosarcoma arising from sphenoid sinuses, eroding the skull base and invading the brain parenchyma, managed successfully by surgical resection.

CASE REPORT

A 40-year-old male was admitted through outpatient department with complaints of headache for 4 months, complete loss of vision of right eye for 2 months and decreased vision of left eye for 1 month. Pain was felt more in temporal regions but whole of the head was involved. It was sometimes associated with vomiting. The patient was non-alcoholic and non-smoker. Furthermore, there was no history of trauma. There was no significant family history for any cancer. The past and socioeconomic histories were also insignificant.

On examination, higher mental functions (HMF) were intact. Also speech and gait were normal. There was no perception of light in the right eye. In the left eye, the visual field defect was found to be left nasal hemianopia. Fundoscopy revealed secondary optic atrophy in the right eye and temporal pale disc in the left eye. Motor, sensory and cerebellar systems were completely normal. Respiratory, cardiovascular and abdominal examinations were insignificant. On systemic review, the patient was found to be impotent for few months.

Magnetic Resonance Imaging (MRI) scan revealed a soft tissue mass arising from sphenoid sinuses, eroding the overlying bone and extending into the sellar and suprasellar regions up to the third ventricle (Figures 1a-d). The tumour mass also showed heterogeneous enhancement with typical “honey-combing” appearance. Computed Tomography (CT) scan revealed stippled calcification in the tumour mass which is typical of chondrosarcoma. The histopathological examination proved it to be a chondrosarcoma.

The patient underwent excision biopsy through subfrontal approach. Bleeding got uncontrolled after resecting the tumour so sponge stones and cottonoids were used to pack the tumour cavity and were left there. Re-do surgery was performed 12 days after the first surgery to remove the homeostatic packs. The operated site was washed and then wound was closed. Patient was placed on appropriate antibiotics and made smooth recovery. He was discharged from hospital on the 21st postoperative day.

Histopathological examination of the specimen revealed chondroid neoplasm showing mild nuclear pleomorphism, and hyperchromasia with cells loosely set in extensively chondromyxoid stroma, features suggesting well-differentiated chondrosarcoma.

Postoperative CT brain revealed near total excision of tumour.

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DISCUSSION

Chondrosarcoma is a rare malignant cartilaginous tumour. It is rare in craniofacial region and comprises only 7% of all tumours of neurosurgical concern. Also primary intracranial chondrosarcoma are very rare comprising only less than 0.16% of all brain tumours. It most commonly involves skull base and usually found to be located in petrous portion of the temporal bone and the petro-occipital, sphenoid-occipital and sphenopetrosal synchondroses;1,2 brain parenchymal invasion is extremely rare,1 as is seen in this case. Grading of chondrosarcoma is particularly based on the appearance of cancerous cells under the microscope and the growth rate of the tumour itself. Several histological subtypes of chondrosarcoma have been reported in literature including conventional, mesenchymal, Clear-cell and de-differentiated subtypes.2,6 Grading system for conventional chondrosarcoma comprises of grade I (well-differentiated) through grade III (poorly differentiated).2 Most of the chondrosarcomas are of conventional subtype,8 while mesenchymal variant has found to be most malignant with strong tendency for intradural and cerebral growth.6

Chondrosarcoma resembles chordoma in clinical presentation and radiological features and can be misdiagnosed as such, but the two differ in behaviours and outcomes.7 Chondrosarcoma shows good prognosis when treated with a combination of microsurgical removal and proton beam irradiation, while chordoma shows no good prognosis despite the same aggressive management.8 Comparison of several imaging findings revealed no diagnostic features for either chondrosarcoma or chordoma, rendering the pre-operative diagnosis difficult on the basis of CT and MR imaging. Yet, these tumours show involvement of specific anatomical structures which might provide a clue for differential diagnosis and surgical planning.7

The signs and symptoms of brain stem and cranial nerve compression presented in cases of chondrosarcomas include headache, visual symptoms like diplopia, other oculomotor disorders depending on the location of the mass, tinnitus, dizziness etc.6 In this case, the signs and symptoms at the first manifestation were caused by mass effect and optic nerve compression.

Both CT scanning and MR imaging are usually necessary for the complete evaluation of chondrosarcomas because of the involvement of both soft tissue and bony structures at the skull base. CT imaging is required to evaluate intratumourous calcification and bone erosion. On MRI imaging, the tumour shows low to intermediate signal intensity on T1-weighted and high signal intensity on T2-weighted images. The chondrosarcomas show marked contrast enhancement in either a heterogeneous or homogeneous pattern.9 On T1-weighted images after gadolinium contrast injection, marked “septal” or “ring-and-arc” enhancement is typical for enchondromas and low-grade chondrosarcoma which corresponds to fibrous bands between the confluent cartilage lobules on the histologic analysis.9 On histological examination, chondrosarcoma shows a characteristic biphasic pattern composed of highly undifferentiated small round cells and islands of hyaline cartilage.8

Chondrosarcoma has been found to be minimally sensitive to conventional radiation and chemotherapy. Thus, radical excision has remained the main treatment modality for chondrosarcoma, except for inoperable cases.1 But the recurrence rate of the tumour is found to be high when the patients are treated with surgery alone.2,4 Since tumour control has not been achieved by surgical treatment alone, local recurrences after surgical treatment have created a need of some adjuvant therapy.3,4 Combined surgical and post-operative proton radiation therapy have shown promising results with regard to tumour control.2,4,5 According to a recent study, a 5-year recurrence rate for the patients of chondrosarcoma, who were treated with surgery alone, was found to be 44% which has dramatically reduced to 9% after the addition of adjuvant radiation therapy.2 The concept of combined maximal surgical resection and adjuvant proton beam irradiation is now widely accepted but further controlled studies would help in establishing its definite role.

Modality of treatment, treatment history, histological subtype, and histological grade of the tumour are the aspects found to influence the recurrence-free survival of the patients.10 High grade chondrosarcoma and
mesenchymal subtypes have shown a bad prognosis and high mortality rates and also more recurrences. To the best of authors’ knowledge, this is the first reported case of its type from Pakistan. We, therefore, recommend concerned entities to include sphenoidal intracranial chondrosarcoma in the differential diagnoses of skull base, sellar and parasellar tumours. This case report would be of interest to not only a particular speciality like neurosurgery as seen in this case but also it would have a broader impact across many areas of medicine as ENT and ophthalmology because early manifestations of such tumours may be related to eye, ear and/or nose.

REFERENCES


