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Apoplexy in an intradural clival chordoma causing intraventricular bleed

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Abstract

Background: A few cases depicting apoplexy in a chordoma have been reported. Rarely, this intratumoral bleed may spillover into intracerebral or intraventricular regions.

Case Description: The authors report an intradural variety of clival chordoma presenting with apoplexy and spillover of blood into lateral ventricle. Clinical presentation, radiological scans, and relevant literature is also described.

Conclusions: In a stable case of clival chordoma, intratumoral bleed or apoplexy may cause rapid neurological worsening and warranting urgent surgical intervention.

Key Words: Apoplexy, chordoma, intraventricular bleed

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INTRODUCTION

Originating from the remnant notochord, chordomas are usually located in extradural bony structures of the skull base. These arise from spheno-occipital synchondrosis and slowly erode surrounding structures depicting locally invasive nature and propensity for aggressive recurrences. Vascular event or apoplexy may be a rare presentation of such a skull base neoplasm.[1,7] This report describes a case of skull base intradural chordoma with apoplexy, causing intraventricular bleed and deterioration of neurological symptoms attributable to bleed.

CASE DESCRIPTION

A 26-year-old male presented with complaints of diplopia for past 3 years and gradually progressive right-sided hemiparesis for 6 months duration. For the past 2 weeks, he was having a severe headache and worsening of sensorium. On examination, the patient was conscious but drowsy. He had left-sided 3rd and 6th cranial nerve palsies, and right-sided spastic hemiparesis. The patient was carrying magnetic resonance imaging (MRI) performed 6 months back, showing an extra-axial mass on the left side, arising from clivus [Figure 1a-e]. Computed tomography (CT) scan after recent clinical deterioration showed a hemorrhagic transformation of clival mass with spillage of bleed into left lateral ventricle [Figure 1f]. The patient underwent transsylvian approach after a left pterional craniotomy to reach the tumor. The tumor was arising from clivus, located intradurally, firm in consistency, grayish, lobulated, with evidence of blood clots with in tumor tissue, interspersed with yellow, and gelatinous material. Subtotal excision of the tumor was achieved due to hemodynamic instability, and histopathological examination was diagnostic of chordoma [Figure 2a and b].

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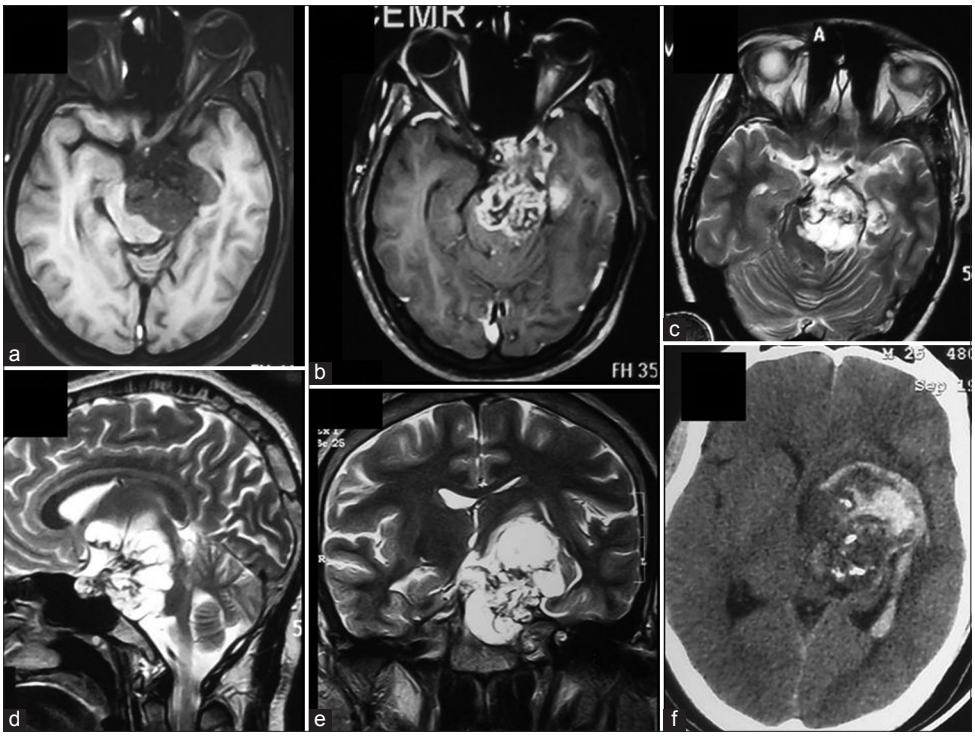

Figure 1: Magnetic resonance imaging scan, axial section of T1-weighted 1 (a), T1-weighted 1 with contrast (b) and T2-weighted 2 (c) showing an enhancing mass on the left side of clivus. Sagittal section of T2-weighted 1 (d) and coronal section of T2-weighted 1 (e) showing an extraosseous, intradural variety of chordoma, displacing pons and midbrain. Axial section, plain computed tomography scan (f) showing apoplectic chordoma with spillage of bleed into occipital horn of lateral ventricle

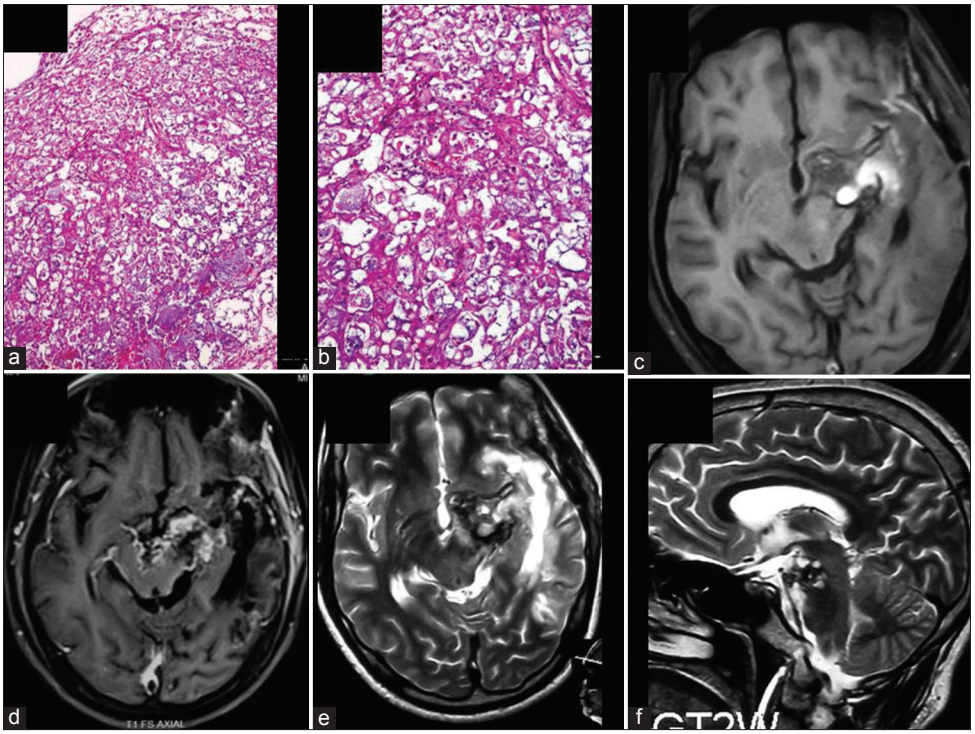

Figure 2: Histopathological photomicrograph (a) showing a cellular tumor arranged in chords and lobules in a myxoid stroma (H and E, ×200). (b) Tumor cells, large sized, having abundant, vacuolated cytoplasm indicating physaliphorous cell (H and E, ×400). Follow-up magnetic resonance imaging scan, axial section of T1-weighted 1 (c), T1-weighted 1 with contrast (d), axial section of T2-weighted 1 (e) and sagittal section of T2-weighted 1 (f) showing subtotal excision of tumor

Postoperatively, the patient had eventless recovery, and follow-up MRI scan showed residual lesion for which gamma-knife radiosurgery was administered [Figure 2c-f].

DISCUSSION

Chordomas usually present with gradually progressive clinical symptoms as these lesions slowly spread centrifugally to involve multiple cranial nerve palsies at the skull base. Occasionally, such a gradual deterioration may be hastened by apoplexy, taking clinicians by a surprise. The apoplectic chordoma may present with intratumoral bleed, subarachnoid bleed, and even intraventricular bleed.[1] The cause of such a phenomenon remains to be elucidated. Intratumoral hemorrhage in chordomas may result from the rupture of the thin-walled vessels or hemorrhagic infarction due to rapid tumor growth. Another mechanism suggests the destruction of the dural vessels due to tumor invasion resulting in intracranial bleed.[1] We support the former patho-physiology as the present CT scan shows blood interspersed in chordoma with spillage into the occipital horn of left lateral ventricle [Figure 1f]. Only a few cases of skull base chordomas with apoplexy have been reported, and only five cases of osseous intradural type presenting with apoplexy have been reported, whereas only two cases of extraosseous chordomas with bleed have been reported.[1-7] All cases of apoplectic chordomas had an intra-axial component and hence dural invasion by tumor is obvious. Apoplectic chordoma, as depicted in our report resembles that of pituitary apoplexy and the cause of bleed due to rupture of dural vessels seems unlikely.

CONCLUSIONS

Apoplexy in a chordoma is a rare phenomenon. Such an event may cause downward spiraling of the clinical status of an otherwise stable patient. The radiological scans, operative, and pathological findings support hemorrhagic infarction due to rapid tumor growth or rupture of the thin-walled vessels within the tumor.

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Conflicts of interest
There are no conflicts of interest.

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