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CASE REPORT

Primary intracranial choriocarcinoma presenting as a ring-enhancing lesion: A case report and review of literature

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ABSTRACT

We report here a rare case of primary intracranial choriocarcinoma without evidence of tumor elsewhere, presenting as a ring-enhancing lesion managed successfully in our institute (Grant Medical College and Sir J. J. Group of Hospitals, Mumbai, Maharashtra, India). A 22-year-old, right-handed housewife presented with complaints of headache, vomiting, generalized tonic–clonic seizures, and right upper limb weakness of short duration. On neurological examination, the patient was conscious, cooperative, well-oriented in time, place, and person. Visual acuity was 6/9 in both eyes. Fundus was showing grade 1 papilledema. Tone was increased on the right side. Power was decreased in the right upper limb as compared to the left. Reflexes were brisk in the right upper limb. Imaging was suggestive of a thick ring-enhancing lesion in the left parietal region suggestive of glioma or tuberculosis. Complete excision of the lesion was achieved. The diagnosis was confirmed by histopathologic examination of the lesion and β-human chorionic gonadotropin (β-HCG) levels. The patient recovered well postoperatively without neurological deficit. She was relieved of headache completely and there were no further episodes of vomiting or seizures. The right upper limb weakness improved to 4+/5. Choriocarcinoma is the most malignant lesion of all the gestational trophoblastic diseases. Primary brain choriocarcinoma presenting as a ring-enhancing lesion has not been reported in the literature. This diagnosis should be kept in mind, especially in young females of child-bearing age group. Complete surgical excision with adjunctive treatment is required to achieve prolonged remissions.

Key words: Choriocarcinoma, enhancing, intracranial, primary, ring

Introduction

Choriocarcinoma is a rare but the most malignant of all the gestational trophoblastic diseases. The most common site of origin is the body of the uterus; however, these lesions metastasize early to the lungs, liver, and less frequently to the brain.[1] Cerebral metastasis is known to present as hemorrhage with significant mass effect and morbidity.[2] Primary brain choriocarcinoma presenting as a ring-enhancing lesion without primary lesion is a very rare phenomenon. We report such a rare case of primary intracranial choriocarcinoma presenting as a ring-enhancing lesion with no evidence of disease elsewhere, and discuss the clinical, radiological, and surgical findings.

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Case Report

A 22-year-old, right-handed housewife presented to the hospital with complaints of headache of 15 days duration. Headache was generalized, intermittent in duration, aching in nature, associated with vomiting and visual obscurations, especially while changing positions. These symptoms aggravated in morning hours. There was also history of single

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episode of generalized tonic–clonic seizure 10 days back, after which the patient was started on anticonvulsants and there were no further episodes. Patient also noticed weakness of the right upper limb since last 8 days, which was gradual in onset and progressive in nature. Weakness was more significant in the left hand as compared to the left shoulder. There was also history of drowsiness since last 2 days.

On neurological examination, the patient was conscious, cooperative, and well oriented in time, place, and person with normal higher mental functions. Visual acuity was 6/9 in both eyes. Fundus was showing grade 1 papilledema on both sides. The rest of the cranial nerves' examination was normal. Nutrition and bulk was normal in all four limbs. Tone was increased on the right side. Power was 4+/5 in the right shoulder and 4/5 in the right elbow and right wrist. Grip was weaker on the right side as compared to the left side. Rest of the limbs had normal power. Sensory examination did not reveal any abnormality. Reflexes were brisk in the right upper limb, rest were normal. Gait was normal. Plain radiograph of the skull was normal.

Computed tomography (CT) scan [Figure 1] showed a lesion of 3.4 cm × 2.4 cm in size with well-defined margins and multiple foci of calcification in the left fronto-parietal region. The lesion was strongly enhancing inhomogenously on contrast administration with surrounding perifocal edema and mass effect suggestive of glioma or tuberculoma.

Magnetic resonance imaging (MRI) [Figure 2] scan was suggestive of a thick ring-enhancing lesion seen in the left high parietal region with perifocal edema with mass effect suggestive of granulomatous lesion, most likely tuberculosis.

CT angiography [Figure 3] showed a small lesion in the left high parietal region with increased vascularity.

Patient was operated with left parietal craniotomy with total excision of lesion. The tumor was adhered to the undersurface of dura. The tumor was dark brown in color, necrotic, and solid with increased vascularity. There were feeders from the periphery. The brain was edematous and pulsating well. Lax duraplasty was done using pericranium. Tumor was excised *in toto*.

Postoperatively, the patient recovered well. She was relieved of headache completely and there were no further episodes of vomiting or seizures. The right upper limb weakness improved to 4+/5. The blurring of vision also improved gradually. There was no new neurological deficit postoperatively.

Postoperative CT scan [Figure 4] showed complete excision of the lesion.

The patient was discharged on 10th postoperative day without any neurological deficit.

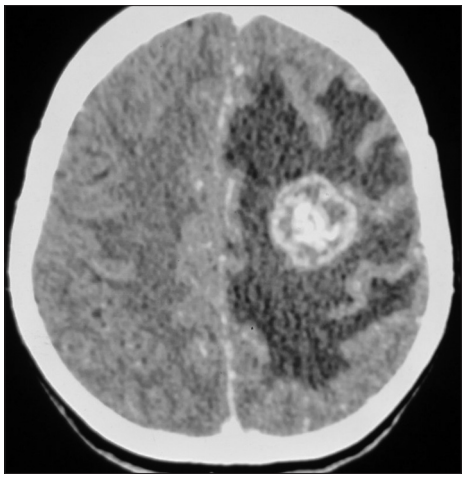

Figure 1: CT scan (axial view) showing a lesion of 3.4 cm × 2.4 cm in size with well-defined margins. The lesion is strongly enhancing inhomogenously on contrast administration, with surrounding perifocal edema and mass effect

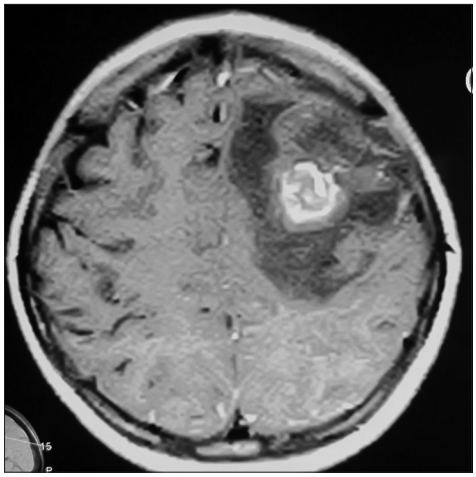

Figure 2: MRI (axial view) scan showing a thick ring-enhancing lesion seen in the left high parietal region with perifocal edema with mass effect

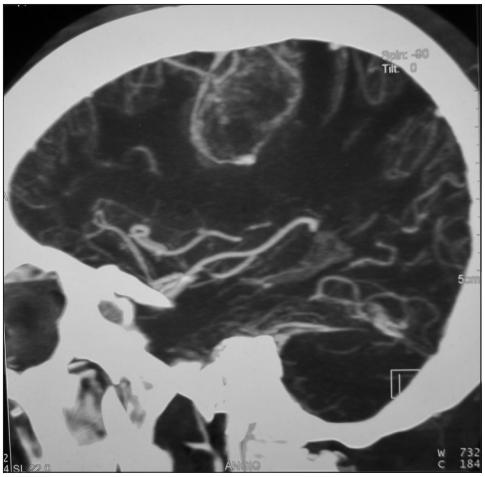

Figure 3: CT angiography showing a small lesion in the left high parietal region with increased vascularity

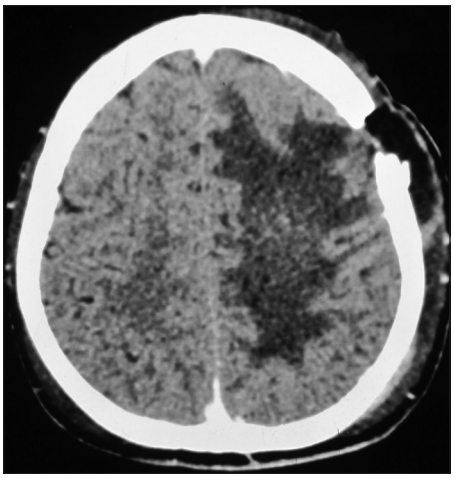

Figure 4: Postoperative CT scan showing complete excision of the lesion

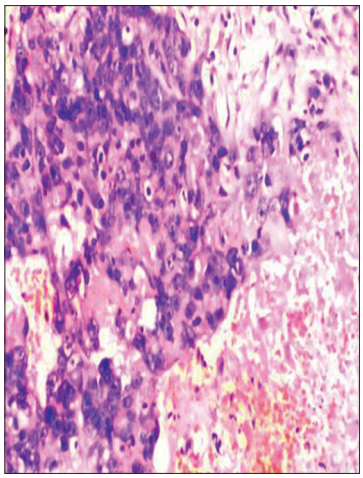

Figure 5: Neuropathological examination showing peripherally arranged tumor cell in sheets and syncytium. These are large cells having moderate eosinophilic to clear cytoplasm with dense hyperchromatic nuclei, increased nucleocytoplasmic ratio, and extensive areas of necrosis and hemorrhage

Neuropathological [Figure 5] examination showed a tumor composed of peripherally arranged tumor cell in sheets and syncytium. These were large cells having moderate eosinophilic to clear cytoplasm with dense hyperchromatic nuclei and increased nucleocytoplasmic ratio and prominent macronucleoli. The tumor showed mitotic activity of about 1-2/high power field, and extensive areas of necrosis and hemorrhage suggestive of primary choriocarcinoma of the left parietal lobe.

In view of the diagnosis of choriocarcinoma, the patient was investigated extensively for the detection of the primary lesion. β-Human chorionic gonadotropin (β-HCG) was 20,000 IU. Gynecology opinion was taken Patient was pregnant once and had delivered full-term baby vaginally 2 years back without complications. Past and present menstrual history was normal. Transvaginal ultrasound was normal. X-ray chest was normal. CT scan abdomen and pelvis did not reveal any abnormality.

The patient has been referred to another specialized institute for adjunctive treatment. She has been following up regularly with evidence of widespread metastasis over a period of 6 months.

Discussion

Trophoblastic diseases comprise four pathological entities: 1) molar pregnancy, 2) invasive mole, 3) placental site trophoblastic tumors, and 4) choriocarcinoma.[3] Choriocarcinoma is the least common but the most malignant of these.

Choriocarcinoma arises from the mononuclear cytotrophoblasts and multinuclear syncytiotrophoblasts and can arise from any type of gestation, however, the most common being hydatiform mole followed by normal pregnancy, spontaneous abortion, or ectopic pregnancy.[4]

The cases of metastatic choriocarcinoma without known primary are reported to arise from the placenta of the normal prior pregnancy which was missed due to its small size.[5] Likewise, in our case, there was history of normal pregnancy 2 years back without complications.

According to Suresh *et al.*,[5] choriocarcinoma constitutes 4% of all brain metastatic tumors; however, none of their patients were diagnosed to have primary gestational choriocarcinoma. Choriocarcinoma spreads by the vascular route and the incidence of brain metastasis varies from 3 to 28%.[5] Metastatic brain choriocarcinoma without pulmonary metastasis has been reported in the literature,[6,7] as in our case where there was no evidence of either primary lesion or pulmonary metastasis.

These tumors have the highest incidence in Asia, Africa, and South America. The most common age of presentation is women less than 35 years,[8] like in our case where the patient was a 22-year-old female.

The common presentation on imaging is that of a hemorrhagic lesion with significant perilesional edema and mass effect which is often mistaken for primary intracerebral hemorrhage due to local pathology instead of metastasis, particularly in the absence of primary lesion. Therefore, diagnosis is established only after pathological examination of blood clots.[5]

Our case is different in that the imaging findings were suggestive of a thick-walled ring-enhancing lesion which is more suggestive of either glioma or tuberculoma particularly in the absence of primary lesion and this type of presentation has not yet been reported in the literature. In such cases, diagnosis can be established only by histopathologic examination of the lesion as in cases of hemorrhagic lesion as described above.

β-HCG level measurement by radioimmunoassay of the blood and CSF is required to confirm the diagnosis. Serum: CSF ratio of less than 60:1 is a sensitive indicator of CNS metastasis.[6]

Surgical excision of the tumor is the treatment of choice for primary brain metastasis presenting with mass effect. Adjunctive treatment in the form of chemotherapy and irradiation has been shown to improve survival.[9] Intrathecal prophylaxis with methotrexate in high-risk patients has been shown to improve survival in patients with brain metastasis.[6]

Our patient has been referred to another specialized institute for adjunctive treatment. However, even with adjunctive treatment, she developed widespread metastasis over a period of 6 months.

Conclusion

Primary brain choriocarcinoma should be considered in the differential diagnosis of a ring-enhancing lesion, especially in young females of child-bearing age group. Histopathologic examination of the lesion is required for the definitive diagnosis of the lesion. CSF and serum β-HCG levels help in supporting the diagnosis and monitoring the treatment. Complete surgical excision with adjunctive treatment is required to achieve prolonged remissions.

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Conflicts of interest

There are no conflicts of interest.

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