Case Report

Choroid plexus carcinoma of the third ventricle: A rare tumor presenting at an unusual site

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ABSTRACT

We present an extremely rare case of choroid plexus carcinoma (CPC) of the third ventricle of the brain. An adult female presented with symptoms of raised intracranial pressure. On magnetic resonance imaging brain, a solid cystic mass was seen in the third ventricle along with obstructive hydrocephalus. Ventriculoperitoneal shunt was done initially to relieve hydrocephalus followed by complete tumor excision. Pathologically, it was confirmed as CPC. She was given adjuvant radiation to a dose of 58 Gy in 29 fractions by volumetric modulated arc therapy. After 1 year of follow-up, the patient is disease-free.

Key words: Choroid plexus carcinoma, Radiation, Surgery, Third ventricle

Choroid plexus carcinomas (CPCs) are rare neoplasms arising from the choroid plexus of the brain or spinal cord which produces cerebrospinal fluid. They are classified as World Health Organization (WHO) grade 3 tumors and 80% are seen in children under the age of 5 years with extremely low incidence in the adult population [1]. Most commonly, CPCs present with increased intracranial pressure leading to headache, vomiting, difficulty in walking, and seizures. Gross total resection followed by adjuvant chemotherapy and radiotherapy is the current treatment, although this remains to be standardized [2]. CPC has a poorer prognosis and the extent of surgery is the most important factor in treatment outcome. CPCs presenting in the third ventricle in adult age group is an extremely rare finding. In the literature, only four such cases are reported to our knowledge [3].

Henceforth, we report an exceptionally rare case of CPC presenting at a rare site in the third ventricle of the brain.

CASE REPORT

A 48-year-old female presented with headache and vertigo leading to a fall and loss of consciousness. There was no past history of any comorbidities or any neurological illness.

On examination, her blood pressure was 100/70 mmHg, pulse rate was 82/min, and Glasgow coma score was E1V1M5.

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On investigation, complete blood counts, liver function, and renal function tests detected no abnormality. Contrast-enhanced magnetic resonance imaging (MRI) of the brain revealed a well-defined solid cystic mass measuring 2.2 × 2.5 × 2.6 cm in relation to the pineal gland (posterior third ventricle) causing marked compression on the aqueduct resulting in dilatation of the third and lateral ventricles, leading to obstructive hydrocephalus. The lesion was hypointense on T1 and heterogeneously hyperintense on T2 and FLAIR images. A whole-body positron emission tomography was done which showed no evidence of disease elsewhere.

Hence, a diagnosis of a primary brain tumor was made and the patient was taken up for surgery. Initially, a ventriculoperitoneal shunt was done to relieve hydrocephalus followed by complete tumor excision by supra-cerebellar approach through suboccipital craniectomy in a sitting position. Intraoperatively, the brain was tense; tumor was soft and moderately vascular. The tumor appeared to arise from the choroid plexus of the third ventricle. The post-operative period was uneventful.

Histopathology showed a high-grade tumor composed of cells arranged in a papillary pattern with increased cellularity and necrosis. On immunohistochemistry, the tumor cells expressed pan-cytokeratin, patchy synaptophysin, and ki-67 index was 10%, whereas, GFAP, PAX-8, TTF-1, and CK-7 were negative. Therefore, a diagnosis of high-grade CPC of the third ventricle was confirmed. Post-operative MRI showed no residual tumor and cerebrospinal fluid (CSF) cytology was negative for malignant

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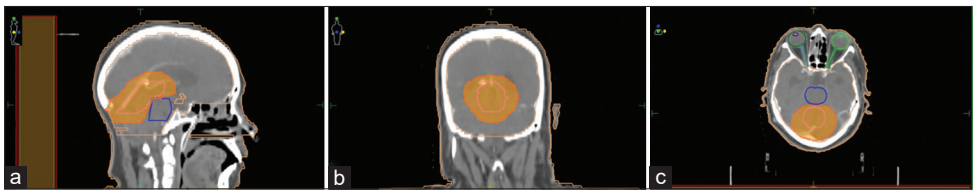

Figure 1: Isodose coverage of planned target volume of 95% of prescribed dose in all three sections (a) sagittal view; (b) coronal view; and (c) axial view

cells. The case was discussed in the institutional tumor board and planned for adjuvant radiation in view of a high-grade tumor. She was given radiation to a dose of 40 Gy in 20 fractions followed by a boost of 18 Gy in 9 fractions by volumetric modulated arc therapy technique prescribed at 95% isodose (Fig. 1). She tolerated radiation well with no complications. After 1-year of follow-up, she is neurologically stable with no signs of recurrence on the MRI brain.

DISCUSSION

The WHO in 2016 has classified choroid plexus tumors into three grades based on their characteristic [1]. Grade 1 choroid plexus tumors are benign and are also called choroid plexus papillomas. Grade 2 tumors are atypical choroid plexus papillomas and have a higher chance of recurrence after surgery. Grade 3 tumors are choroid plexus and they are malignant. CPCs are seen mainly in children under the age of 5 years with extremely low incidence in the adult population [4]. Our patient presented in the middle-age group.

These tumors show a slight male preponderance, whereas, more female patients are seen with tumors located in the fourth ventricle. Genetic association with Li-Fraumeni syndrome and Aicardi syndrome is seen in a few patients [4]. Location-wise, they can present in third, fourth, or lateral ventricles; however, the most common location is the lateral ventricle in the pediatric and the fourth ventricle in adult patients. CPCs presenting in the third ventricle in adult age group is an extremely rare finding.

Contrast-enhanced MRI of the brain is the main diagnostic modality to diagnose CPCs. They typically appear in the ventricles and have irregular borders with a "cauliflower-like appearance with areas of cyst and necrosis along with perilesional edema. Calcification may be seen in 20–25% of cases [5]. CPCs may also have CSF seeding so imaging of the entire neural axis is recommended either before surgery or after surgery. The differential diagnosis of these tumors is pineoblastoma, germinoma, ependymoma, medulloblastoma, and primitive neuroectodermal tumor. Pathologically, these tumors have increased cellularity, nuclear pleomorphism, high mitotic rate, and more importantly distorted papillary structure as defined by the WHO recently [1]. The presence of brain parenchymal invasion is the hallmark to differentiate carcinoma from papilloma. Immunophenotypically, CPCs are cytokeratin and p53 positive whereas S100 and epithelial membrane antigen are negative [6].

Surgical resection is the mainstay of treatment and the extent of surgery determines the prognosis. A median survival of 58 months versus 36 months has been reported with gross total excision as opposed to subtotal resection [2]. The other prognostic factors are age, location, size of tumor, and CSF spread. Patients with younger age and larger tumor size have poor prognoses. After surgery, treatment options are adjuvant radiation with or without chemotherapy depending on the age and neurological condition of the patient. Radiation is associated with significantly better survival in many series as nowadays newer techniques deliver targeted radiation with minimal toxicity [7]. However, in the majority of cases, radiation cannot be given due to the young age of the patient. Alternatively, chemotherapy is given in such patients and studies have shown that chemotherapy also prolongs survival [8]. Since the incidence of CPCs is very low, the experience with chemotherapy is also limited, and chemotherapeutic agents used in these cases are still not well defined.

The prognosis of CPCs is poor with a 5-year survival rate of 40–50% as they are rapidly growing tumors and treatment options are limited. Adults have a better prognosis with a 5-year survival of up to 70% as they can tolerate radiation and chemotherapy better [9]. Clinical trials with immunotherapy and targeted therapy are in progress for better treatment options [10].

CONCLUSION

CPC arising from the third ventricle in an older age group is extremely rare tumors. Adequate surgery followed by radiation and/or chemotherapy is the treatment of choice. Unfortunately, the incidence of CPC is too low to standardize adjuvant radiation or chemotherapy protocols for such patients.

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