**Management of Choroid Plexus Carcinoma in a Young Adult: A Case Report and Review of Treatment Strategies**

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**Abstract:**

Choroid plexus carcinoma (CPC) is an extremely rare disease in adults, with an incidence ranging from 0.76% to 4%. It is more commonly observed in children, accounting for 2-5% of all pediatric brain tumors. There are no established guidelines for the optimal management of CPC in adults. This case report details a 20-year-old male diagnosed with CPC, who presented with positive CSF cytology and an isolated spinal deposit. His treatment included surgery followed by adjuvant radiation and concurrent chemotherapy.

**Keywords:** Choroid plexus carcinoma, CSI (cranio-spinal irradiation) GTR (Gross total resection).

**Introduction:**

Choroid Plexus carcinoma is considered an extremely rare primary neoplasm of the brain. It arises from the differentiated epithelial tissue of choroid plexus of ventricles and 90% of times, from the lateral and fourth ventricles[1]. The occurrence of choroid plexus neoplasm is 0.3 to 0.6% of all intracranial tumors. These are found more frequently in childhood as compared to adults. In children, they are found in the lateral ventricles, while in adults, they are in the fourth ventricle[2].

Choroid plexus tumors can be grossly divided into 2 categories: choroid plexus papilloma (CPP) and Choroid plexus carcinoma (CPC). CPP are considered benign lesions (WHO Grade 1) with good prognosis. CPC are typically malignant (WHO Grade III) they have an aggressive nature and reported 5 year overall survival is around 40% [3, 4].

Choroid plexus carcinomas are similar to adenocarcinomas in terms of morphology. They must be differentiated from metastatic carcinomas, which are more common[5]. Determining the epidemiology of choroid plexus carcinoma is extremely difficult. However, an analysis of the National Cancer database showed that the mean of patients diagnosed with this disease is 2.34 years[6]. The literature shows a low survival rate in children suffering from Choroid Plexus Carcinoma, with a median ranging from 9 months to 58 months after total resection[7].

The histopathological diagnosis in primary choroid plexus carcinoma is more complex than in choroid plexus papilloma [8]. The microscopic picture depicts clear evidence of malignancy, including frequent mitoses (>5/10 HPF), nuclear polymorphism, and necrosis [9]. On immunohistochemistry, these tumors show positivity for S100, GFAP, and cytokeratin. Of these, cytokeratin positivity is essential as it excludes meningioma and ependymoma [10]. As per previous literature, around 25% of these cases exhibit tumor dissemination into the cerebrospinal fluid (CSI) or even distant metastases, indicating its aggressive nature [9]. After surgery, the treatment of choroid plexus carcinoma involves adjuvant chemotherapy, such as the use of carboplatin. These have been proven to decrease metastasis and increase overall survival in patients suffering from such tumors[11].

According to the literature, Choroid Plexus carcinoma, being sporadic, is primarily observed in children. Yet, we present a case of further uniqueness as our patient was a young adult starting his 20s. Additionally, our patient survived despite the disease having a poor prognosis.

**CASE REPORT:**

**Case History/ Examination**

A 20-year-old male student of bachelor’s in computers was in his usual state of health until around four months back when he presented with complaints of headache and vomiting which were non-projectile in nature. There were 1-2 episodes of vomiting per day. The headache was temporarily relieved by vomiting. He was investigated at a private hospital, and MRI Brain with contrast **(Figure1-3)** showed a right lateral ventricular mass centered on the choroid plexus representing choroid papilloma measuring 3.3\*3.4\*3.2cm; it was iso-intense on T1 and hyper-intense on T2 and FLAIR sequence. He was operated on 25/01/2024 and underwent maximal safe resection: Post-surgery, both the headache and vomiting resolved. The histopathology of the specimen came out to be choroid plexus carcinoma **(Figure 4)**. CKAE1/AE3: +, p53: +, EMA: -. Post-operative MRI showed an abnormal signal intensity tracking from the site of the scar up to the posterior horn of the lateral ventricle with an internal area of hemorrhage more in favor of post-procedural changes; however, few patchy areas of enhancement were also seen that may represent residual disease. He was then referred to our hospital for further management. Upon first presentation at the outpatient department, the young boy was alert and active, well-oriented in time, place, and person. His GCS was 15/15, and he had no focal neurological deficit. His vision and hearing were normal, and power in all four limbs was 5/5.

**Management**

Since only a few cases of choroid plexus carcinoma were reported in the literature, a further management plan was decided after a detailed discussion in the multi-disciplinary tumor board. Since the tumor originated in the ventricles, CSF cytology was advised that it was positive for malignant cells. MRI of the Spine with contrast to see for any drop metastases revealed a well-formed enhancing intradural lesion seen inseparable from the right S2 nerve root indenting the spinal thecal sac measuring approximately 6.9\*6.8mm concerning for metastatic deposit **(Figure 5 & 6)**. Bilateral S1 roots appeared slightly bulky, but no lesion could be seen. Because of these findings (presence of drop metastases and positive CSF cytology), he was treated with radiotherapy to both cranio-spinal axes, followed by a boost to the post-operative disease site and spinal deposit. CT simulation was done in the supine position, and the patient was immobilized using a Thermoplastic mask and Vac lock. CT slices of 1mm thickness were obtained. A dose of 30 Gy in 15 fractions was planned for cranio-spinal irradiation, followed by an additional dose of 30 Gy in 15 fractions to the post-operative disease site **(Figure 7)**. The volumes were contoured using pre-operative gross tumor volume (GTV) with 1.5cm margin for clinical target volume and a further 5mm margin to account for setup error was given for planning treatment volume (PTV). He also received 150mg weekly Carboplatin during this second course of 15 fractions concurrently with radiation, given 1 hour before radiotherapy. His hematological and biochemical profile was done at baseline and weekly before chemotherapy was normal. He was then planned on an SBRT boost to the spinal deposit and after contouring the gross tumor volume (GTV), a margin of 5mm was given for CTV and an additional 3mm for PTV. The doses to all organs at risk, particularly the genitals were kept well below the tolerance limit.

**Result**

During the entire course of radiotherapy, the patient did not report any major issues except mild headache, nausea, and hair loss at the site of radiotherapy. The patient recovered and was well-oriented at the end of treatment. No major abnormalities or clinical signs were observed after the treatment. Thus, following concurrent radiotherapy, the patient is asymptomatic and is advised to follow up with MRI Brain and Spine. Post-Operative MRIs showed recovery of patient. **(Figures 8-10)**



Figure 1: Pre-operative MRI of the patient showing a lesion involving the right lateral ventricle. (Axial images)



Figure 2: Pre-operative MRI of the patient showing a lesion involving the right lateral ventricle

(Sagittal images)



Figure 3: Pre-operative MRI of the patient showing a lesion involving the right lateral ventricle.

(Coronal images)

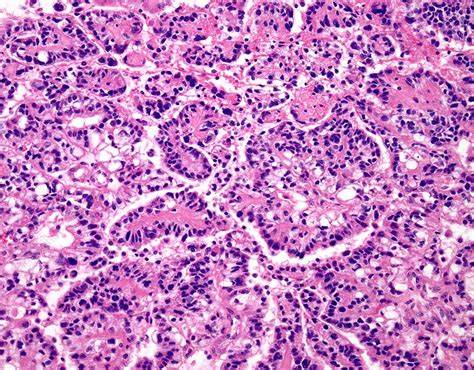


Figure 4: Histopathology of choroid plexus carcinoma showing increased mitosis and blurring of papillary architecture.

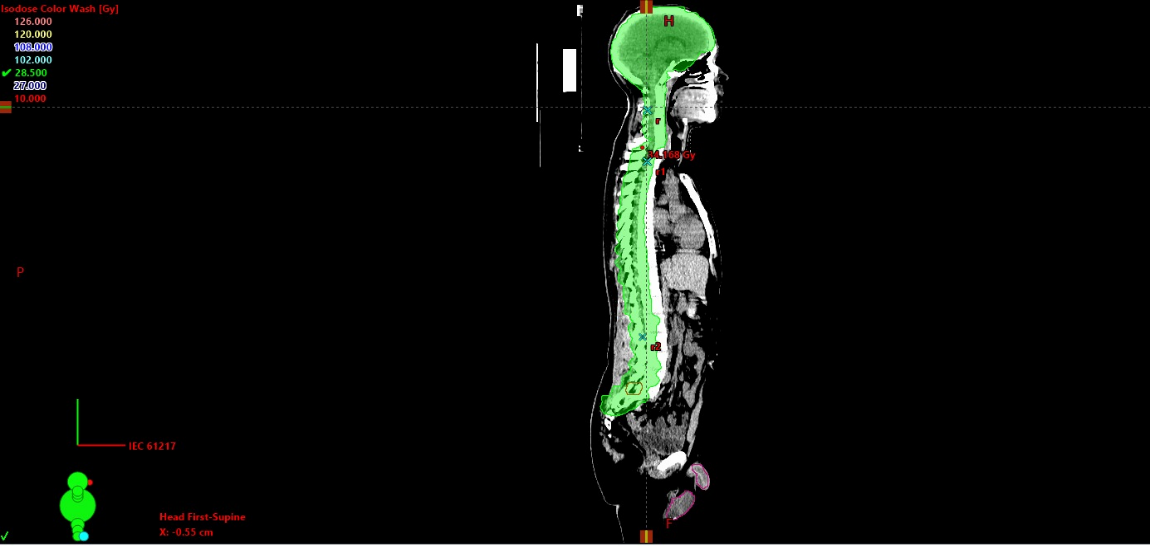
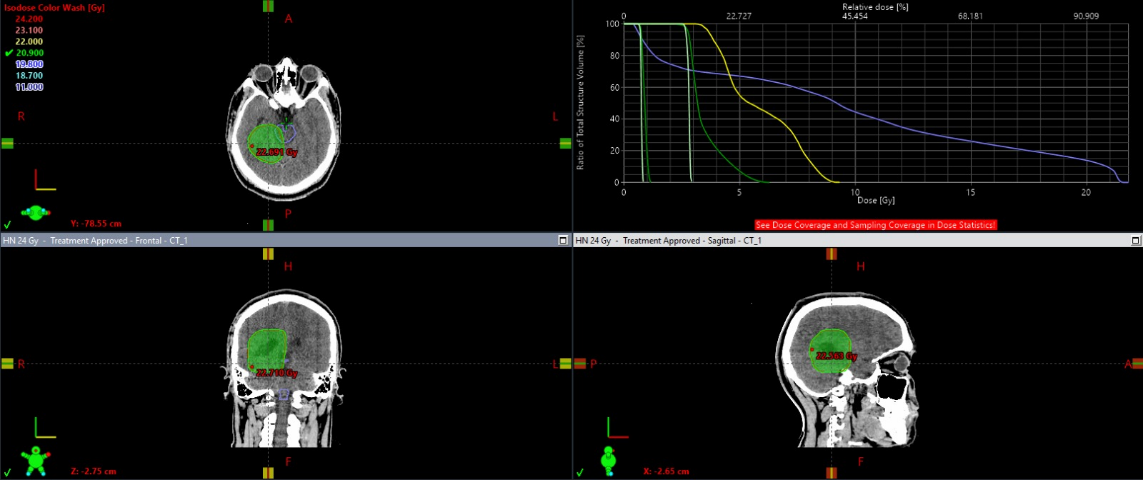
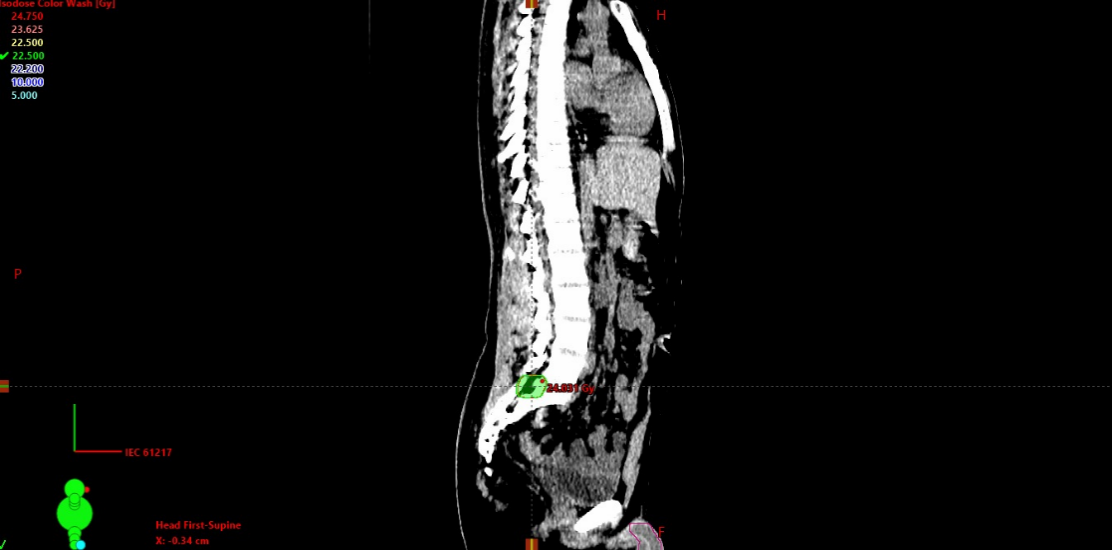
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**Figure 5: Planning CT image of the patient fused with MRI showing pre-operative volumes.**



**Figure 6: Planning CT image showing treatment volume of metastatic spinal deposit R side.**

**Figure 7: Treatment plan of the patient CSI 30Gy in 15 fractions, Primary tumor boost 30 Gy in 15 fractions using IMRT, and Spinal boost of 22.5Gy in 5 fractions using SRT, respectively.**

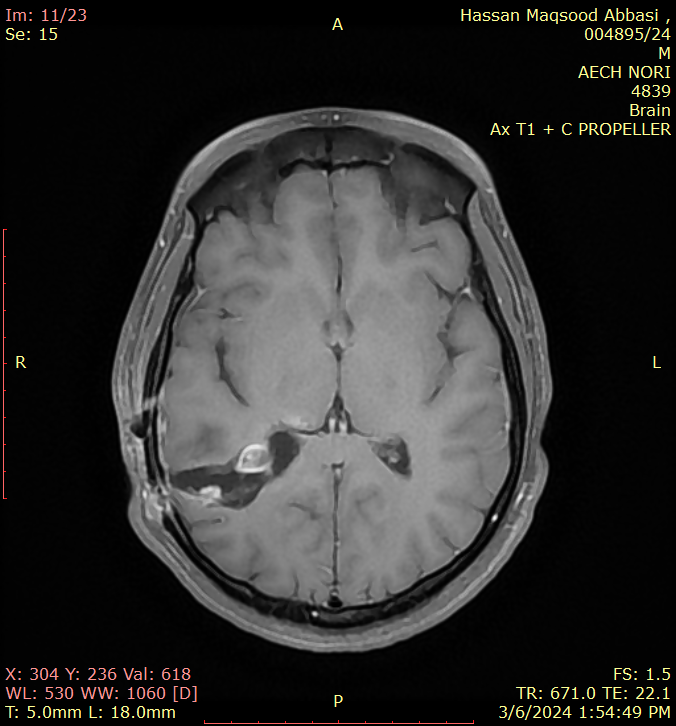


Figure 8: Post-operative MRI of the patient showing post-surgical changes.

(Axial images)

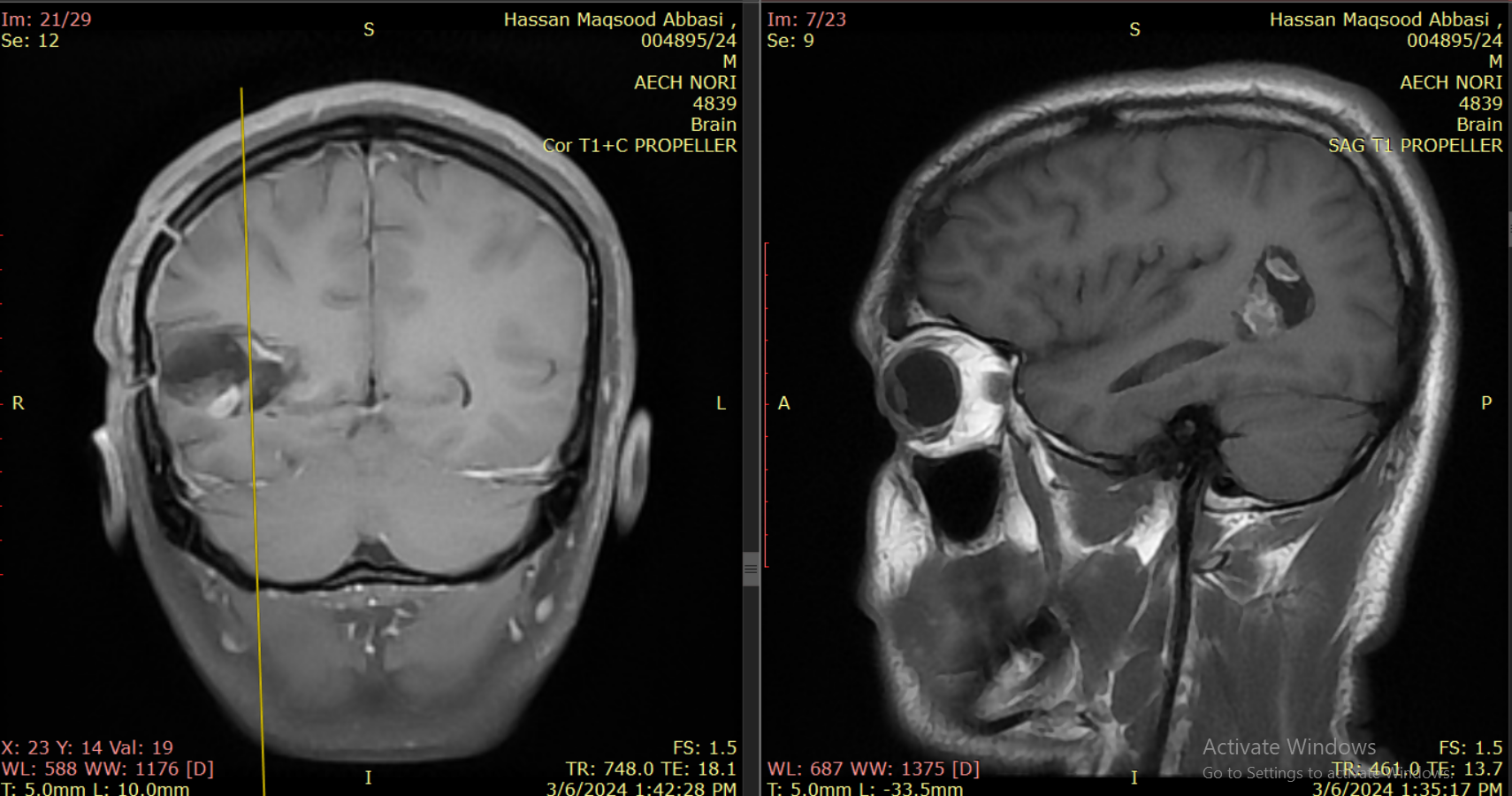
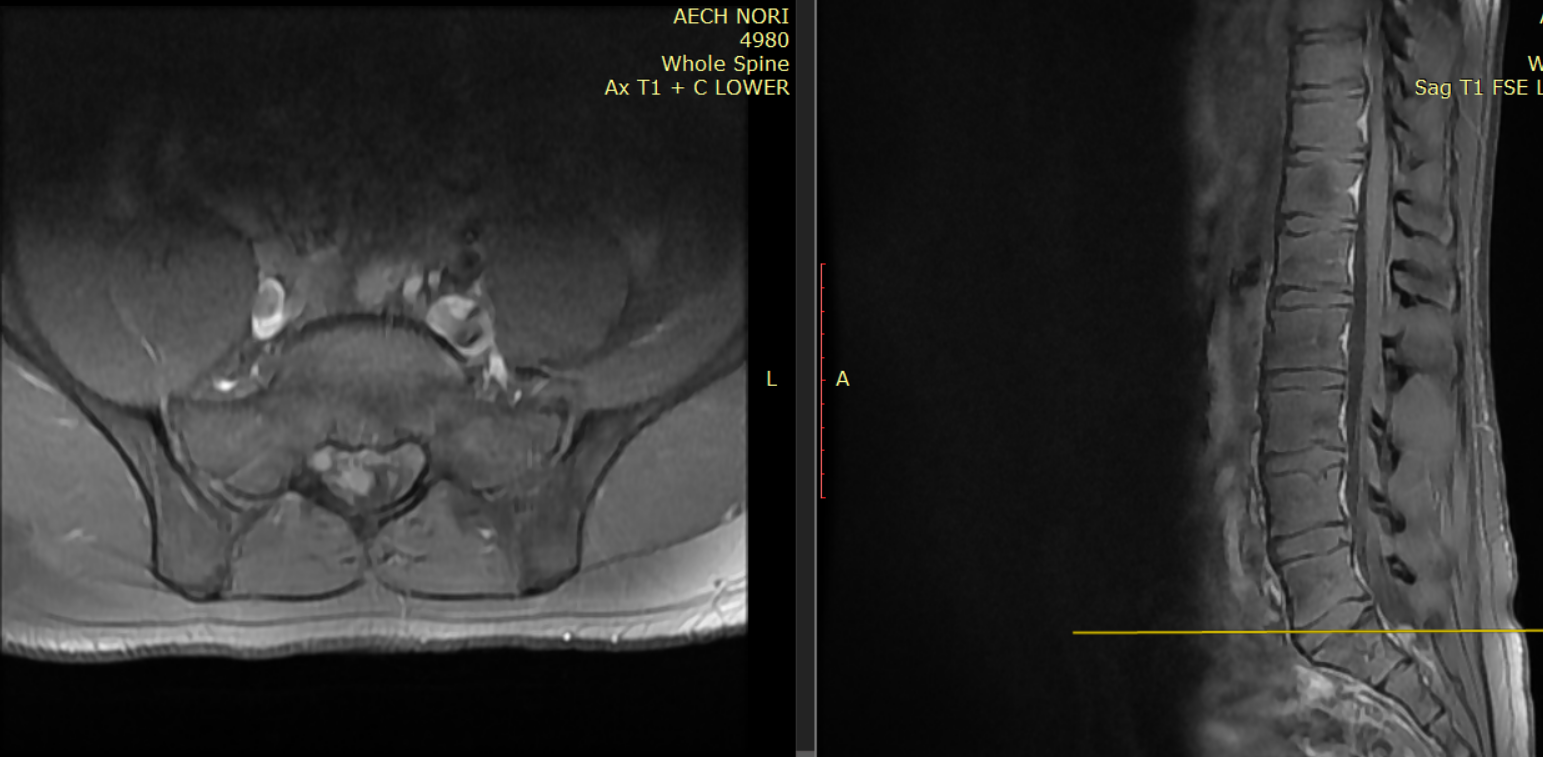


Figure 9: Post-operative MRI of the patient showing post-surgical changes.

(Coronal and Sagital images)



**Figure 10: Post-operative MRI Spine of the patient showing post-surgical changes.**

**(Axial and Sagittal images)**

**Discussion:**

It is believed that the presentation of choroid plexus carcinoma is infrequent in adults[2]. Yet, we present a case report of this disease in a young adult. Additionally, it is believed that in adults the choroid plexus carcinoma is commonly observed in the fourth ventricles[2]. However, our patient had this disease in the right lateral ventricle. This is in line with another study by Han et al., who reported a 35-year-old patient with a mass in the right lateral ventricle[12]. Jo et al. also report a mass in the lateral ventricle but on the left side[13]. This shows that the occurrence in lateral ventricles of adults might not be as rare as we initially thought. Or this might be due to regional variation, as these three cases were in Asian countries. However, the patient in Han’s report was unconscious, while ours and Jo’s were oriented and alert at the time of presentation. This might be due to early hospital visits in our and Jo’s cases[12, 13].

Our patient presented with non-projectile vomiting in addition to headache. This was not observed in the earlier cases; however, Zhang et al. reported a similar presentation of a patient with vomiting and nausea. However, its nature of being projectile or non-projectile was not reported[14]. The patient in Han’s case was not conscious, so there might have been an omission or lack of observation of vomiting[12]. Therefore, we believe only further studies on the topic might be able to determine vomiting as a significant or insignificant symptom of CPC in addition to headache.

The literature showed case reports of choroid plexus carcinoma with no spread or leptomeningeal spread. Our patient did not have leptomeningeal spread, but spinal roots were involved [8]. A mass was observed down to the S1 root. The re-irradiation was performed targeting both the cranial and spinal spread.

**Conclusion:**

Choroid plexus carcinoma (CPC) in an adult is a rare scenario, but this must also be considered an important differential while dealing with tumors involving the ventricles. Surgical excision gross total resection (GTR) remains the standard of care. Although there are no clear-cut guidelines regarding adjuvant treatment options, chemotherapy, and radiotherapy, both have been reported to improve outcomes in patients diagnosed with CPC.

**Key Clinical Message**

Thus, we have to point out that despite the scarcity of literature and the rarety of such tumors, their existence cannot be understated. We must not forget to make proper diagnoses via board discussions, as in the case of this patient, so that patient care can be improved and survival can be ensured in future cases.

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Declaration:

**Ethical Approval:**

Written informed consent was obtained from the patient to publish this case report and any accompanying images. A copy of the written consent is available for review by the journal's Editor-in-Chief. It is certified that the case report of a 20-year-old male patient diagnosed with choroid plexus carcinoma of the right lateral ventricle has been approved by the research and training cell of AECH-NORI.

Availability of data:

The results can be openly accessed

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