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*Case Report*

# Metastatic choroid plexus papilloma presenting as a sellar mass: A case report and literature review

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## ABSTRACT

**Background:** Choroid plexus papillomas (CPPs) are rare neoplasms arising from choroid plexus epithelium representing <1% of all intracranial tumors. Symptoms vary based on location and regional mass effect; however, hydrocephalus is common due to cerebrospinal fluid flow obstruction and/or overproduction. Distant site metastasis or *de novo* formation in extraventricular sites is rare.

**Case Description:** A 57-year-old female with a history of a 4th ventricular CPP status post resection in 2004 and 2018 with subsequent gamma knife therapy in 2019 presented with increased thirst and urination. Since her initial surgery, she has experienced significant gait imbalance, diplopia, dysphagia, and right-sided hemiparesis and hemisensory loss. Magnetic resonance imaging revealed a new 1.5 × 1.8 cm suprasellar lesion. She underwent a left supraorbital craniotomy for tumor resection, with pathology revealing metastatic World Health Organization grade II CPP.

**Conclusion:** Extraventricular manifestation of CPP is rare. *De novo* or metastatic involvement of the sella has seldom been reported. Treatment should target gross total surgical resection. Adjuvant chemotherapy and radiation may be useful in higher-grade lesions.

**Keywords:** Choroid plexus papilloma, Metastatic, Sella, Supraorbital craniotomy

## INTRODUCTION

Choroid plexus papillomas (CPPs) are rare neoplasms arising from choroid plexus epithelium and represent <1% of all intracranial tumors.[17] CPPs may occur in both adults and children with a slight male predilection (1.6:1).[7] The lateral ventricles are the most common location in children, while 4th ventricular tumors are more common in adults.[6] Symptoms vary based on location and regional mass effect; however, hydrocephalus is common due to cerebrospinal fluid (CSF) flow obstruction and/or overproduction. Signs of elevated intracranial pressure, including headache, nausea, vomiting, cranial nerve deficits, gait imbalance, and altered mental status are commonly seen.[12]

According to the World Health Organization (WHO) classification, choroid plexus tumors are classified as papillomas (grade I), atypical tumors (grade II), and carcinomas (CPC; grade III) based on the number of mitotic figures per high-power field on histologic examination. Papillomas are typically soft, friable masses with high vascularity.[15] The preferred treatment is gross total surgical resection and is typically curative in cases of papillomas.[14] Incomplete

resection and increased mitotic activity are predictive of tumor recurrence.[5]

Less commonly, CPP may invade surrounding parenchyma or disseminate throughout the central nervous system (CNS). Cases of leptomeningeal spread and the formation of numerous cystic lesions in both the brain and spine have been reported.[6,12,13]

Here, we present a 57-year-old patient with prior 4th ventricular CPP with a solid distant metastasis to the sella, treated with supraorbital craniotomy for resection.

## CASE PRESENTATION

A 57-year-old female with a history of a 4th ventricular CPP presented with increased thirst and urination. She initially underwent suboccipital craniotomy for tumor resection in 2004 and again in 2018 for tumor recurrence. She received subsequent Gamma Knife radiotherapy in 2019. Pathology from both resections revealed atypical CPP, WHO grade II. Since her initial surgery, she has experienced significant gait imbalance, diplopia, dysphagia, and right-sided hemiparesis and hemisensory loss. Surveillance magnetic resonance imaging (MRI) was done, showing an enhancing 1.5 × 1.8 cm suprasellar lesion, as well as a stable residual 5 × 5 mm lesion in the left posterolateral margin of the 4th ventricle [Figure 1]. Preoperative visual field testing showed decreased left eye visual acuity without field cuts. She has patched the left eye since her initial surgery to improve diplopia. The endocrine evaluation revealed a prolactin level of 82.2 ng/mL attributed to the stalk effect without additional overt hormonal deficiency. Given the growth of the suprasellar lesion and the need for tissue diagnosis, she presented in April 2023 for left-sided craniotomy for tumor resection.

The patient was treated with a left supraorbital craniotomy for tumor resection. Given her baseline functional status, we felt that an eyebrow incision provided the least invasive approach to access the tumor. A craniotomy was performed on the left side due to her existing right-sided deficits. We decided against an endoscopic transnasal transsphenoidal approach, given the patient's obesity and associated increased risk for CSF leak. We additionally felt an endonasal approach and potential complications may exacerbate the patient's baseline dysphagia. During resection, the firm lesion was densely adherent to the hypothalamic surface. Meticulous dissection with microinstruments allowed a tissue plane to be developed, and tumor resection continued toward the sellar floor. Preservation of functioning pituitary tissue was attempted; however, the normal-appearing pituitary gland was not visualized throughout tumor dissection and debulking. A small tumor was residual and densely adherent to the optic apparatus and was left behind to allow vision preservation. Closure was completed in standard fashion using a pericranial graft for watertight dural repair.

Similarly to her prior surgeries, pathology revealed WHO II atypical CPP, suggesting metastasis. She developed panhypopituitarism treated with a regimen of desmopressin, levothyroxine, and hydrocortisone. She had a prolonged hospital course complicated by worsened dysphagia and altered mental status attributed to hypoxia secondary to atelectasis. Her respiratory status gradually improved with positive pressure ventilation through BiPAP, and her mental status returned to her preoperative baseline. Her worsened that dysphagia, however, did persist and ultimately required gastrostomy tube placement. Postoperative imaging demonstrated a small residual lesion within the sella [Figure 2] without evidence of surrounding

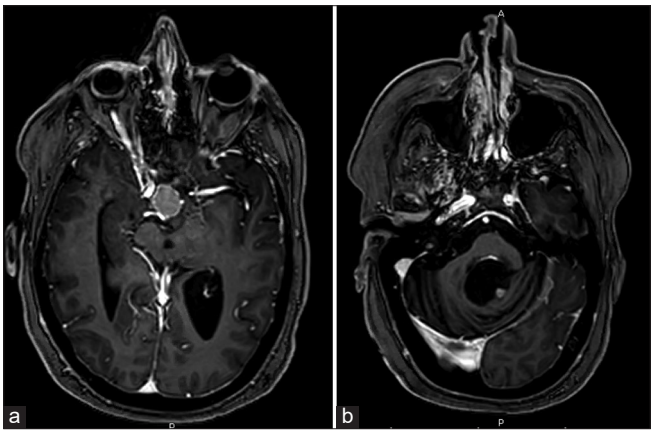


**Figure 1:** (a) Preoperative axial T1-weighted magnetic resonance imaging (MRI) brain with contrast showing a 1.5 × 1.8 cm sellar mass. (b) Preoperative axial T1-weighted MRI brain with contrast showing a small 5 × 5 mm enhancing nodule in the left posterolateral margin of the 4th ventricle, representing residual choroid plexus papilloma.

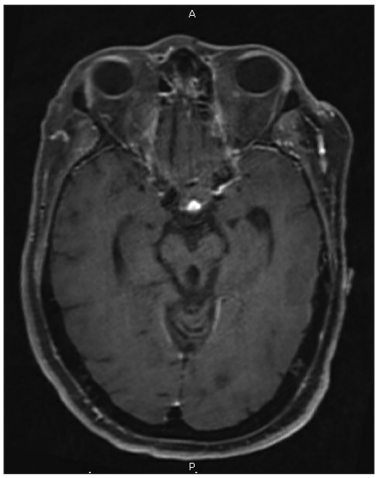

**Figure 2:** Postoperative T1-weighted magnetic resonance imaging brain with contrast showing expected post-surgical changes and decreased tumor volume, with an 8 × 6 mm area of tumor residual.

areas of CNS insult or ischemia. She was discharged home in June 2023.

## DISCUSSION

Although rare, there have been several documented cases of CPP with distant lesions throughout the neuroaxis. Metastatic lesions may present as solid-enhancing lesions, "drop metastases" in the spine, or leptomeningeal seeding.[1,3] Ortega-Martínez *et al*. reviewed 15 publications describing 16 patients with disseminated CPP.[13] All but one patient had primary lesions in the posterior fossa. Eleven patients had leptomeningeal spread, with variable involvement of supratentorial, infratentorial, and spinal sites. Five patients specifically had leptomeningeal involvement in the suprasellar region. Three patients developed solid parenchymal lesions, and three had new intraventricular lesions in the 3rd or 4th ventricle.

Numerous widespread cystic lesions have also been described. Johnson *et al*. reviewed a series of eight such patients.[6] Six patients had primary solitary lesions (five of which were in the 4th ventricle), two of which had multiple solid lesions. Cystic lesions were widely distributed throughout subarachnoid spaces, including the spine. Pathology in seven of the lesions was WHO grade I, with one patient having WHO II atypical CPP.

Given the reported cases of disseminated CPP, Mazur-Hart *et al*. proposed a new classification system describing the spectrum of disease.[12] Pathologic grades are subdivided based on the presence of solitary or multiple lesions. The WHO grade I multiple lesions are further described as disseminated disease, multifocal disease, Zuckerguss seeding of the leptomeninges, or ependymal disease.

Solid lesions involving the sellar region appear to be exceptionally rare. We identified eight publications describing eight patients with sellar CPP, including our own [Table 1]. All previously reported patients had *de novo* formation of the WHO grade I CPP without evidence of metastatic spread from elsewhere. To the best of our knowledge, our case is the first to document metastatic spread from a distant lesion to the sella. Six of the eight patients reported were female. Ages ranged from 31 to 57. One patient received adjuvant radiotherapy postoperatively. MRI findings appear nonspecific, with variable degrees of contrast enhancement on T1-weighted images. One case demonstrated a large cystic component resembling craniopharyngioma.

While the mainstay of treatment involves gross total resection, the role of adjuvant therapies is less well-established. Chemotherapy, particularly in cases of CPC, has been reported to increase overall survival.[18] Although data regarding specific dosing regimens is largely lacking, etoposide, vincristine, cisplatin, cyclophosphamide, carboplatin, or ifosfamide are most commonly utilized. In a randomized trial, Wolff *et al*. noted an improved 5- and 10-year progression-free survival in patients receiving carboplatin versus cyclophosphamide in combination with etoposide and

**Table 1:** Summary of reported CPP cases in the sellar region.

| Author, year | Age/sex | Pathology | Radiographic appearance (MRI) | Surgical approach | Adjuvant therapy | *De novo* | Metastatic |
|---|---|---|---|---|---|---|---|
| Kimura *et al*. (1992)[9] | 34/F | WHO I | • Homogenous T1 enhancement<br>• T2 isointense | Pterional, STR | - | + | - |
| Ma *et al*. (2008)[11] | 49/F | WHO I | • Heterogenous T1 enhancement<br>• Mixed T2 hyper-/hypointensity | Endoscopic TNTS, GTR | - | + | - |
| Sameshima *et al*. (2010)[16] | 51/F | WHO I | • Heterogenous T1 enhancement<br>• Heterogenous T2 iso/hyperintensity | Endoscopic TNTS, STR | - | + | - |
| Bian *et al*. (2011)[2] | 31/F | WHO I | • Heterogenous T1 enhancement<br>• Mixed T2 hyper-/hypointensity | Endoscopic TNTS, GTR | - | + | - |
| Keskin *et al*. (2016)[8] | 50/M | WHO I | • T1 hypointense with enhancing nodule<br>• T2 hyperintense, cystic appearance | Pterional, GTR | - | + | - |
| Gong *et al*. (2017)[4] | 43/F | WHO I, pigmented | • Homogenous T1 enhancement<br>• T2 hypointense | Endoscopic TNTS, STR; subsequent subfrontal | RT | + | - |
| Kuo *et al*. (2018)[10] | 43/M | WHO I | • Heterogenous T1 enhancement<br>• Mixed T2 hyper-/hypointensity | Endoscopic TNTS, GTR | - | + | - |
| Our case | 57/F | WHO II | • Homogenous T1 enhancement | Supraorbital, STR | - | - | + |

CPP: Choroid plexus papillomas, MRI: Magnetic resonance imaging, GTR: Gross total resection, RT: Radiation therapy, STR: Subtotal resection, TNTS: Transnasal transsphenoidal, WHO: World Health Organization

vincristine for higher-grade lesions.[19] Regimens consisted of six cycles of chemotherapy every 28 days using etoposide 100 mg/m$^2$ on days 1–5, vincristine 1.5 mg/m$^2$ on day 1, and either carboplatin 350 mg/m$^2$ or cyclophosphamide 1 mg/m$^2$ on days 1–2. Fractionated radiation therapy may also be used in select cases.

The findings in our case highlight the importance of recognizing potential metastasis in CPP, including new lesions that may arise in extraventricular locations. Sellar lesions may mimic typical adenomas or craniopharyngioma; however, in patients with a history of known CPP, metastatic disease should remain in the differential. Multiple lesions may arise regardless of pathologic grade. Surveillance imaging remains essential, and full neuroaxis imaging should be considered to identify potential distant spread.

## CONCLUSION

Extraventricular manifestation of CPP is rare. *De novo* or metastatic involvement of the sella has seldom been reported. Treatment should involve gross total surgical resection to prevent recurrence. Adjuvant chemotherapy and radiation may be appropriate in higher-grade lesions.

### Ethical approval

The Institutional Review Board approval is not required.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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### Conflicts of interest

There are no conflicts of interest.

### Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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