Spinal drop metastasis from a benign fourth ventricular choroid plexus papilloma in a pediatric patient: case report

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Choroid plexus papillomas (CPPs) are typically benign tumors that can occur in any age group but are more commonly found in pediatric patients. Although these tumors are benign, there are several reports in adult patients of distant metastases present either at the time of diagnosis or occurring months to years after initial resection. Here, the authors report the case of a 14-year-old boy who presented with symptoms of elevated intracranial pressure due to obstructive hydrocephalus that was caused by a large fourth ventricular mass. Preoperative imaging included a full MRI of the spine, which revealed an intradural lesion that encased the distal sacral nerve roots at the tip of the thecal sac and was concerning for a drop metastasis. The patient underwent gross-total resection of both the fourth ventricular and sacral tumors with histology of both lesions consistent with benign CPP (WHO Grade I). In addition, the authors review prior reports of both pediatric and adult patients in whom benign CPPs have metastasized with either benign or atypical pathology found at a distant site. Taking into account this unusual case and reports in the literature, patients with even benign CPPs may warrant initial and routine follow-up imaging of the total neural axis in search of the rare, but possible, occurrence of drop metastasis.

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KEY WORDS choroid plexus papilloma; metastasis; disseminated; pediatric; oncology

CHOROID plexus papillomas (CPPs) are typically extraaxial benign tumors that originate from the neuroepithelial lining of the choroid plexus. These tumors are quite rare, accounting for about 0.4%–0.6% of all intracranial tumors.^{21,37} CPPs can be found in any age group but are more commonly found in pediatric patients, with a median age at diagnosis of 3.5 years and an incidence of 3.9% in children younger than 12 years.^{21,37} As the origin of CPP is from the choroid plexus, it can be found anywhere the choroid plexus is located. While an infratentorial location is typical in adults (e.g., fourth ventricle), these tumors are more frequently found in a supratentorial location (e.g., lateral ventricles) in children.^{15,37} Benign CPPs (WHO Grade I) resemble normal choroid plexus with papillary projections containing cuboidal-to-columnar epithelium lining fibrovascular bundles. While occasional mitoses are present, in general these tumors do not demonstrate cytological atypia. Atypical CPPs (WHO Grade II) demonstrate increased mitotic activity with at least 2 mitoses per 10 hpf. Other features may include hypercellularity, cytological atypia, and necrosis.²⁹ Given that

CPPs are generally benign and locally slow growing, these tumors are typically cured through gross-total resection alone.^{28,30}

Although rare, there have been several reported cases of benign CPPs forming distant metastases in adults.^{1–6, 8–10,12–14,16–19,22–27,31–36,38,39} In contrast, there are even fewer reported cases that demonstrate similar findings in the pediatric population. In this report, we present the rare case of a pediatric patient who was found to have a benign fourth ventricular CPP and a sacral drop metastasis at initial presentation. In addition, we review the literature comprehensively and summarize prior reports in both pediatric and adult patients in whom benign CPPs have metastasized to the distant sites.

Case Report

History and Examination

A 14-year-old boy presented to the emergency department with new complaints of pressure-like headaches and a 2-month history of morning vomiting. On examination,

ABBREVIATIONS CPC = choroid plexus carcinoma; CPP = choroid plexus papilloma.

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FIG. 1. Preoperative MR images. **A and B:** Sagittal (A) and axial (B) postcontrast T1-weighted images showing a 4 × 3-cm heterogeneously enhancing extraaxial fourth ventricular mass. This mass was associated with significant obstructive hydrocephalus. **C and D:** Sagittal T1- (C) and T2- (D) weighted images demonstrating a sacral lesion at the bottom of the thecal sac (arrows).

the patient demonstrated mild bilateral upper-extremity dysmetria but otherwise had normal findings on formal neurological testing. Brain MRI demonstrated a 4 × 3-cm heterogeneously enhancing extraaxial fourth ventricular mass causing local mass effect and obstructive hydrocephalus (Fig. 1A and B). Given initial concern for a malignancy, MRI of the entire spine was also performed and revealed a lesion intertwined within the sacral nerve roots (Fig. 1C and D). The patient was asymptomatic from this sacral lesion.

Operative Course and Pathological Findings

The patient was subsequently brought to the operating

FIG. 2. Photomicrographs. **A:** Histological examination of the fourth ventricular mass demonstrating classic findings of CPP, including columnar-appearing epithelium surrounding fibrovascular projections. There were fewer than 2 mitoses per 10 hpf on section analysis, and this lesion was classified as a benign CPP (WHO Grade I). **B:** Similar findings were observed within histological sections of the sacral mass with a diagnosis of benign CPP (WHO Grade I). H & E, original magnification ×200. Figure is available in color online only.

FIG. 3. Postoperative sagittal MR images showing complete resection of both the fourth ventricular (A) and sacral (B) tumor components.

room for a suboccipital craniotomy for tumor resection. Intraoperatively, it was noted that the tumor was moderately tough in consistency and slightly adherent to the floor of the fourth ventricle. A week after the craniotomy, the patient was brought back to the operating room for a sacral laminotomy and resection of the intradural sacral mass. During this procedure, intraoperative ultrasonography was used to confirm the location of the lesion prior to opening the dura mater. A purple-reddish tumor was found at the bottom of the thecal sac intertwined with the sacral nerve roots. Using sharp microdissection and nerve stimulation, the tumor was separated from the nerve roots; postresection neuromonitoring demonstrated no signal changes. Histological examination of both tumors revealed benign CPP (WHO Grade I), with no concerns for a malignant phenotype (Fig. 2).

Postoperative Course

Postoperative MRI of the brain and spine revealed gross-total resection of both lesions (Fig. 3). The patient's dysmetria improved, and he was weaned off his external ventricular drain and did not require permanent CSF diversion. The patient was eventually discharged home in good condition. Follow-up MRI of both the brain and total spine at 3, 6, and 9 months continued to demonstrate no recurrence or new lesions. Given the metastatic presentation of the lesion and young age of the patient, we continue to perform MRI of the brain and total spine every 3 months with a plan to lengthen the imaging interval if scanning findings remain negative.

Discussion

Choroid plexus tumors are typically categorized histologically according to the WHO grading system: benign CPP (WHO Grade I), atypical CPP (WHO Grade II), and choroid plexus carcinoma (CPC; WHO Grade III). While metastases occur more frequently with CPCs, metastases in benign lesions are thought to be rare. Yet, in adult patients, there are several examples demonstrating that benign CPP can form distant metastases.^{1–6,8–10,12–14,16–19,22–27,31–36,38,39} A review of the literature demonstrated 35 cases in adults (age range 19–74 years, mean 40.3 years; 14 males, 19 females, and 2 patients in whom sex and age were not reported) (Table 1). These included cases in which the metastatic

TABLE 1. Cases of metastatic CPP in adult patients

Authors & Year	Age (yrs)*	Primary Pathology	Primary Location	Metastasis Pathology	Metastasis Location	Metastasis Treatment	Time to Metastatic Lesion	Outcome
Wilkins & Rutledge, 1961	56, M	CPP	4th ventricle	Not available	Multiple nodules over cerebellum	None	5 yrs	Death 7 yrs after 1st op
Leys et al., 1986	40, M	CPP	4th ventricle	CPP	Local recurrence & lat ventricle	Not available	9 yrs	Not available
Girardot et al., 1990	30, F	CPP	Cisterna magna	Not available	Local recurrence & 4th ventricle	Not available	5 yrs	Not available
Enomoto et al., 1991	46, F	CPP	Lt CPA	CPP	Rt temporal region	Staged resection of lt CPA & rt temporal lesions	Present on initial imaging	Mild deafness immediately postop
Domingues et al., 1991	35, F	CPP	Lt lat foramen magnum	Not available	Multiple lesions at L3–4	Lumbar lesions not treated	Present on initial imaging	Not stated
Niikawa et al., 1993	38, M	CPP	4th ventricle	CPP (description notes atypical cells)	Lt CPA & suprasellar region; lumbar spinal lesions found later	Staged resection of suprasellar & lt CPA lesion; whole-brain RT & RT to lumbar spinal lesions	6 yrs	Mild limb ataxia after last op w/ stable disease 8 mos after
Shakespeare et al., 1997	27, F	Benign CPP	4th ventricle	Benign CPP	2 local recurrences w/ additional metastases at lt trigeminal ganglia, along rt acoustic nerve, lt cerebellum, & along spinal axis	Re-resection of 4th ventricular tumor; vincristine, carboplatin, & cyclophosphamide but new nodule found after; followed by craniospinal RT	2 yrs for local recurrence, 3 yrs until diffuse metastases	Patient remained symptom free 13 mos after craniospinal RT w/ disease regression in lumbosacral area & stable disease elsewhere
Leblanc et al., 1998	19, F	Benign CPP	4th ventricle	Leptomeningeal choroid plexus cysts	Innumerable cranial & spinal leptomeningeal lesions	4th ventricular tumor removed w/ removal of a few leptomeningeal cysts in passing	Present on initial imaging	Diplopia postop; MRI 3 yrs postop showed enlargement of residual cysts but patient remained asymptomatic
Talacchi et al., 1999	38, M	CPP	Rt CPA	Not available	Local recurrence & suprasellar metastasis	SRS to suprasellar & recurrent CPA lesions	5 yrs	Died 7 yrs after 1st presentation
Valencak et al., 2000	33, F	CPP	4th ventricle	Not available	Local recurrence, 3rd ventricle & spinal axis	Resection of local recurrence followed by RT; progression requiring SRS to local recurrence; SRS to 3rd ventricular lesion; additional RT to local recurrence; clinically worsened so started CCNU	Residual tumor after resection that progressed 8 mos after 1st op; 43 mos for 3rd ventricular lesion; 51 mos for spinal axis lesions	Gait disturbance but otherwise doing well 42 mos after initiation of CCNU

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lesions demonstrated progression to atypical CPPs^{26,33,38} or retained benign features similar to those of the original primary lesion.^{1,2,4,9,16–18,22,23,25,27,31,33,39} Furthermore, these metastatic lesions were found within the ventricular sys-

tem, posterior fossa (including the cerebellopontine angle), or anterior or middle cranial fossa; diffusely involving the leptomeninges; or along the spinal axis.

Among the cases in adults, the majority of patients

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TABLE 1. Cases of metastatic CPP in adult patients

Authors & Year	Age (yrs)*	Primary Pathology	Primary Location	Metastasis Pathology	Metastasis Location	Metastasis Treatment	Time to Metastatic Lesion	Outcome
Irsutti et al., 2000	48, F	Benign CPP (no atypia)	4th ventricle	Benign CPP (no atypia)	Local recurrence & suprasellar region	Resection of local recurrence; later resection of suprasellar lesion	8 yrs	Lt CN VI & VII palsies & ataxia; MRI 6 mos after last resection showed no recurrence or distant seeding
Jagielski et al., 2001	50, M	CPP (WHO Grade I)	Either 3rd or 4th ventricle	CPP (WHO Grade I) for rt temporal lesion	Parasellar cisterns; later lt temporal lesion, rt CPA, lt cerebellar, & C1–2 lesions	Resection of 4th ventricular lesion w/ postop RT; resection of rt temporal lesion	Parasellar lesion on initial imaging; lt temporal lesion, rt CPA, lt cerebellar, & C1–2 lesions found 4 yrs after 1st presentation	Died 5 yrs after diagnosis
Heese et al., 2002	38, M	CPP	4th ventricle	Not available	Leptomeninges around brainstem, interpeduncular fossa, chiasmatic cistern, & conus medullaris	After resection of 4th ventricular tumor, spontaneous regression of all prior leptomeningeal areas of enhancement; resection of local recurrence	Present on initial imaging; local recurrence w/ cyst 8 mos after 1st op	Discharged from 2nd surgery w/ mild gait disturbance & diplopia
McEvoy et al., 2002	51, M	CPP (no evidence of atypia)	4th ventricle	CPP (no evidence of atypia)	Diffuse craniospinal metastases including foramen magnum, suprasellar, CPA, & leptomeningeal spread from cervical to lumbosacral region	Resection of sacral lesion	5 yrs	Not stated
Yu et al., 2006	30, M	CPP	4th ventricle	Atypical CPP	Spinal lesions btwn conus medullaris & terminal filum, C3–4 & T7	Staged resection of all 3 spinal lesions	19 yrs	Pain resolved but experienced urinary retention, mild lower-extremity weakness, & saddle anesthesia 2 mos after op
McCall et al., 2006	38, F	CPP	Posterior fossa	CPP (cystic lesions)	Multiple cystic lesions involving frontal lobe, middle cranial fossa, cerebellum, brainstem, & cervical cord; small mass in 4th ventricle	TMZ	8 yrs	Stable disease at ~17 mos after initiation of TMZ

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TABLE 1. Cases of metastatic CPP in adult patients

Authors & Year	Age (yrs)*	Primary Pathology	Primary Location	Metastasis Pathology	Metastasis Location	Metastasis Treatment	Time to Metastatic Lesion	Outcome
	22, F	CPP	4th ventricle/lt cerebellar hemisphere	Not available	Local recurrence, suprasellar region, spinal drop metastasis	Focal RT to local recurrence; focal RT to thoracic to sacral spine; SRS to suprasellar region; TMZ given after worsening clinical symptoms; intrathecal MTX after leptomeningeal spread but stopped due to bacterial meningitis; switched to cisplatin, cyclophosphamide, & etoposide	2.5 yrs until local recurrence; 5 yrs: metastases to suprasellar region & spinal column; 9 yrs: diffuse leptomeningeal spread & enlargement of spinal drop metastases	Continued to be followed in clinic after multiagent chemo w/ most recent KPS score 60%
Jinhu et al., 2007	32, F	Benign CPP	4th ventricle	Benign CPP (thoracic lesion)	Leptomeningeal infiltration at cervicomedullary junction, T8–9 lesion	Resection of thoracic lesion	1 mo	Spontaneous regression of leptomeningeal enhancement encasing brainstem 2 mos after resection
Ahn & Cho, 2007	36, F	Benign CPP (no atypism noted)	4th ventricle	Benign CPP (no atypism noted) for both lesions	Local recurrence of intracranial tumor, rt cerebellar lesion, several lesions btwn L-3 & S-1 w/ largest at S-1	Staged resection of cerebellar local recurrence & sacral lesion; adjuvant CCNU after	6 yrs	Not stated
Ortega-Martínez et al., 2007	20, F	CPP	4th ventricle	CPP	Lesions in cerebellar tonsils & obex, both cavernous sinuses, & spinal region w/ lesions by cervical cord down to sacral region	Intrathecal Ara-C w/ systemic cisplatin, bleomycin, & vincristine	6 yrs	Died ~6 yrs after 1st presentation (2 mos after biopsy of cervical lesion)
Kaptanoglu et al., 2007	51, F	CPP	4th ventricle	CPP	L4–5 lesion	Lumbar lesion resected	7 yrs	At 1 yr, neurologically intact & pain free
Akil et al., 2008	74, M	CPP (WHO Grade I)	4th ventricle	Not available	Multiple thoracic & lumbar spine lesions	Only 4th ventricular lesion resected	Present on initial imaging	Patient died 2 wks postop due to aspiration pneumonia
Palmer et al., 2010	74, M	Benign CPP	4th ventricle	Atypical CPP	Local recurrence; T-5 intradural extramedullary lesion	Resection & RT for local recurrence; resection & RT for T-5 lesion	13 yrs (local recurrence), 19 yrs (T-5 lesion)	Remained paraplegic after treatment
Al-Abdullah et al., 2011	35, M	CPP	4th ventricle	CPP (WHO Grade I; subfrontal mass)	Both CPAs, subfrontal region, multiple middle & posterior cranial fossa lesions	Resection of subfrontal mass	16 yrs	Not stated

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TABLE 1. Cases of metastatic CPP in adult patients

Authors & Year	Age (yrs)*	Primary Pathology	Primary Location	Metastasis Pathology	Metastasis Location	Metastasis Treatment	Time to Metastatic Lesion	Outcome
Peyre et al., 2012	39, F	Benign CPP	4th ventricle	CPP	Prepontine cistern to Meckel's cave	SRS to Meckel's cave lesion; resection of 4th ventricular mass; later resection of prepontine mass due to progression	Present on initial imaging	1 yr after resection showed small residual tumor in Meckel's cave w/ stable diplopia after op
	46, F	Not stated	4th ventricle	Benign CPP (lt CPA lesion, rt fronto-temporal lesion)	Lt CPA, rt cavernous sinus, temporal lesion	Staged resection of lt CPA & rt fronto-temporal lesions; adjuvant RT after 2nd op	Present on initial imaging	5 yrs after initial presentation lesions stable in size, patient neurologically stable
Anderson et al., 2013	58, M	CPP	4th ventricle	Not stated	Lumbosacral spine	EBRT to lumbar spine; after progression, received CCNU, 6-thioguanine, & bevacizumab	5 mos until lesions w/ in lumbosacral region; disease progression 3 yrs after EBRT to spine	Radiographic stability of lesions w/ improved pain control 12 mos following bevacizumab initiation
Zachary et al., 2014	21, F	CPP	Involving 4th ventricle, brainstem, & rt PICA	CPP (WHO Grade I)	Local recurrence in addition to lesion in rt anterior & middle fossa regions & multiple lesions from cervical to sacral spine	Resection of rt temporal lesion & craniospinal RT followed by boost RT to cribriform plate, brainstem, cervical & thoracic spine, & cauda equina	8 yrs	3 yrs after RT, no recurrence or progression w/ patient returning to work
Scholsem et al., 2012	59, F	Multifocal atypical CPP	Rt lat ventricle & 4th ventricle tumors discovered 1st	Low-grade CPP (4th & rt lat ventricular tumor)	4th ventricle, rt lat ventricle, lt lat ventricle	Staged resection of 4th ventricular tumor & rt lat ventricular tumor; patient refused resection of lt lat ventricular tumor	Rt lat & 4th ventricular tumors present on initial scan; lt lat ventricular discovered 6 yrs later	Not stated
Stuivenvolt et al., 2012	50, F	Benign CPP (WHO Grade I)	4th ventricle	Atypical CPP (WHO Grade II)	Local recurrence, S1–2 level intradural extramedullary, T-9 level intradural	Resection of sacral lesion; RT to craniospinal axis w/ boosts to relapsed 4th ventricular tumor, T-9, & S1–2 regions	6 yrs	8 yrs after initial presentation, no neurological complaints; MRI showed no change of recurrent tumor in 4th ventricle or at T-9
Bettegowda et al., 2012	Not stated	CPP	4th ventricle	Not stated	Lumbosacral spine	Not stated	Not stated	Not stated
	Not stated	CPP	4th ventricle	Not stated	Lumbosacral spine	Not stated	Not stated	Not stated

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had a primary lesion within the fourth ventricle, although this was not essential to metastasis formation. In addition, while some metastatic lesions were discovered in concur-

rence with a primary lesion at initial presentation, other groups have reported that evidence of metastasis did not become apparent until years later. Yu et al. reported an

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TABLE 1. Cases of metastatic CPP in adult patients

Authors & Year	Age (yrs)*	Primary Pathology	Primary Location	Metastasis Pathology	Metastasis Location	Metastasis Treatment	Time to Metastatic Lesion	Outcome
Abdulkader et al., 2016	19, F	CPP	Rt lat ventricle (reported to be multifocal on 1st presentation)	CPP	Both lat & 4th ventricles; 3rd ventricular/suprasellar region, rt porus acusticus, Lt CN VI, posterior cerebellar vermis	Resection of 3rd ventricular/suprasellar lesion	19 yrs	Postop developed diabetes insipidus; follow-up MR images revealed slow continuous growth of mass in the rt lat ventricle w/ stable disease elsewhere
	50, M	CPP	4th ventricle	CPP	Multiple posterior fossa lesions & spinal leptomeningeal lesions	Resection of 4th ventricular & large CPA lesions; radiosurgery required starting 12 yrs later; eventually TMZ	Present on initial imaging	TMZ did not lead to tumor regression & patient died
	27, M	CPP	4th ventricle	CPP	3rd ventricle, multiple 4th ventricle lesions, pineal region	Resection & radiosurgery for local recurrence; radiosurgery for distant spread	25 yrs	Died of unknown causes

CCNU = lomustine; chemo = chemotherapy; CPA = cerebellopontine angle; CN = cranial nerve; EBRT = external-beam RT; KPS = Karnofsky Performance Scale; MTX = methotrexate; PICA = posterior inferior cerebellar artery; RT = radiotherapy; SRS = stereotactic radiosurgery; TMZ = temozolomide.

* Age at the first presentation.

extreme example in which a patient developed diffuse spinal lesions 19 years after his initial diagnosis and resection.³⁸ It is unclear whether these differences in time to metastasis are due to seeding at the time of surgery or by innate tumor cell spread within the CSF. Based on the literature and prior reports, it is likely that tumor cells originating within the fourth ventricle have more direct access to the subarachnoid space outside the ventricular pathway through the foramen of Magendie and foramina of Luschka. This may also account for the fewer number of reports of metastatic CPPs in pediatric patients, in whom supratentorial locations are more common.^{15,37}

In pediatric patients, in contrast to adults, there are very few reports of benign metastasis or drop metastasis. A review of the literature demonstrated 3 cases, with 2 males and 1 female ranging from 3 to 16 years of age (mean 8.7 years; Table 2). Doglietto et al. reported on a 16-year-old

boy who, on initial presentation, was found to have diffuse solid lesions in the cisterna magna, both cerebellopontine angles, and lumbosacral space. Surgical removal of the solid and cystic lesions in the cisterna magna confirmed a diagnosis of CPP.⁷ Guidetti and Spallone reported on a 3-year-old girl with a third ventricular CPP that was resected. Unfortunately, 3 years later this patient was found to have a new fourth ventricular mass, which was theorized to be due to tumor seeding rather than an intrinsic lesion originating from the fourth ventricle choroid plexus itself.¹¹ However, the pathological diagnosis and histological characteristics of this new mass were never examined. Masuzawa et al. reported on a 7-year-old patient who was diagnosed with a fourth ventricular CPP and eventually underwent resection of the tumor 9 years after the initial diagnosis. Approximately 4 years after the initial operation, the patient developed symptoms of cervical cord

TABLE 2. Cases of metastatic CPP in pediatric patients

Authors & Year	Age (yrs)*	Primary Pathology	Primary Location	Metastasis Pathology	Metastasis Location	Treatment of Metastasis	Time to Metastatic Lesion	Outcome
Guidetti & Spallone, 1981	3, F	CPP	3rd ventricle	Not available	4th ventricle	Family refused surgery	3 yrs	Not stated
Masuzawa et al., 1981	7, M	CPP	4th ventricle & CPA region	CPP	C6–7 cervical region	Subtotal resection	13 yrs	Not stated
Doglietto et al., 2005	16, M	CPP	Cisterna magna	CPP (cisterna magna cystic lesions)	Multiple craniospinal lesions	Resection of cisterna magna solid & cystic lesions	Present on initial imaging	Lt hypoacusia 1 yr postop w/o signs of disease progression

* Age at the first presentation.

compression with a mass lesion found at the C6–7 region, with histological findings being consistent with a well-differentiated CPP, albeit with more nuclear pleomorphism and mitotic figures.²⁰ Although not stated, this metastatic lesion may have been classified as an atypical CPP. Here, we describe a pediatric patient who presented with both a benign (WHO Grade I) CPP within the fourth ventricle and a sacral drop metastasis concurrently, prior to any surgical intervention. This finding, in conjunction with the case reported by Doglietto et al., suggests that, although rare, CPPs have an intrinsic ability to metastasize not only in adult patients but also in children.

With this evidence in mind, the workup and long-term management of patients with benign CPPs may need to be reexamined to incorporate the possibility of metastases. For fourth ventricular tumors in a pediatric patient, most practitioners would obtain an MRI study of the entire spine at initial presentation, given the concern for drop metastases from possible medulloblastoma or ependymoma. In patients who may not have undergone spine imaging on initial workup for various reasons, we believe that immediate postoperative imaging of the spine should be performed. Given this case and other reports, it can be argued that MRI of the entire spinal axis on surveillance imaging is also warranted even in patients with benign CPPs (WHO Grade I). This approach has been advocated previously in the context of adult patients, but less so in the pediatric population.^{33,38} As there were some cases in which patients died of spread of their primary tumor, catching a metastatic lesion at initial presentation or earlier on follow-up imaging may prevent such devastating outcomes.

It is unclear what the best treatment strategies are for patients with disseminated disease and if there are prophylactic measures that may be taken to prevent metastasis. Although radiotherapy, stereotactic radiosurgery, and various chemotherapy agents have been used, in some cases with success, there are no standard treatment algorithms for metastatic CPP lesions, and no report has demonstrated the use of adjuvant therapies in pediatric patients. In some reports, metastatic lesions causing significant neurological deficits were resected, but resection in cases of extensive disseminated lesions was limited. With the understanding that each case is unique and challenging in its own manner, our recommendation for the management of benign CPPs with CSF spread and metastasis is generally to remove all lesions if possible, because this approach can be curative. Given the rarity of these cases, however, a prospective clinical trial or even a retrospective comparative study may not be feasible. Moreover, there are no clear guidelines for the length of follow-up needed for these patients, depending strongly on practice habits. As metastases have been found up to 19 years after initial presentation, it may be especially important in the pediatric patient to be attentive to signs and symptoms of potential slow-growing distant lesions. Again, we advocate imaging of the brain and total spine during follow-up to ensure detection of potential new sites of growth.

Conclusions

Here, we report the case of a metastatic benign sacral

CPP discovered at the time of initial presentation in a patient with a benign fourth ventricular CPP. Both lesions were treated successfully with resection alone, and follow-up is currently ongoing. Although metastatic spread of a benign CPP is a rare occurrence in the pediatric population, this case, in conjunction with reports within the literature, illustrates the utility of undertaking imaging of the entire spinal axis at the time of diagnosis. By doing so, metastatic lesions may undergo early intervention, preventing poor outcomes such as disabling neurological deficits or even death.

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Disclosures

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Author Contributions

Conception and design: all authors. Acquisition of data: Lau, Sun, Ostling. Analysis and interpretation of data: all authors. Drafting the article: all authors. Critically revising the article: all authors. Reviewed submitted version of manuscript: Lau, Ostling. Approved the final version of the manuscript on behalf of all authors: Morshed.

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