Unusual presentation of more common disease/injury

Atypical transformation in sacral drop metastasis from posterior fossa choroid plexus papilloma

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Summary

Choroid plexus papillomas (CPP) are rare tumours and spinal metastases of CPP are even less common. We report a 50-year-old woman with spinal drop metastases at Th9 and S1–2 6 years after total resection of a posterior fossa CPP. The metastasis at S1–2 was resected and histological examination showed transformation to an atypical CPP. Atypical transformation in a metastasis years after resection of a benign posterior fossa CPP has been described once. We would like to advocate craniospinal MRI at the time of initial diagnosis as well as periodic follow-up after total and subtotal resection of a posterior fossa CPP in adults at least once in 1 or 2 years, depending on the histological grading of the primary CPP. In our case report, this could have resulted in earlier diagnosis of the locoregional recurrence or of the spinal drop metastasis.

BACKGROUND

Choroid plexus tumours (CPTs) arise from the neuro-epithelial lining of the choroid plexus mostly in the lateral ventricles in children whereas in adults they occur more commonly in the fourth ventricle of the posterior fossa. CPTs are rare tumours and represent 0.4–1% of all adult and 4% of all paediatric brain tumours.¹ CPT comprise benign choroid plexus papillomas (CPP, WHO grade I), atypical CPP (WHO grade II) and choroid plexus carcinoma (CPC, WHO grade III). Histological definitions of atypical CPP are not quite clear. In contrast to benign tumours, they display an increase in mitotic activity, nuclear pleomorphism and cellularity.² CPP have a good prognosis if total resection is achieved. However, local recurrence, malignant transformation and spinal metastasis can occur. Spinal metastasis from benign CPPs has been reported infrequently and usually occurs within a few years of diagnosis of the primary lesion. Atypical transformation in spinal metastases is even more rare.² We present a case of atypical transformation in a sacral drop metastasis from a benign posterior fossa choroid plexus papilloma 6 years after surgical resection.

CASE PRESENTATION

A 50-year-old woman presented in 2004 with a 2-month history of dizziness, headache, nausea and vomiting. On neurological examination no abnormalities were seen, besides slight bilateral papilloedema. MRI showed a large inhomogeneous contrast-enhancing tumour with a diameter of 3 cm occupying the fourth ventricle, resulting in a supratentorial hydrocephalus (figure 1A). A posterior fossa craniotomy with total resection of the tumour was performed in May 2004. Cranial postoperative MRI showed no residual tumour in the fourth ventricle. The histological diagnosis was a

choroid plexus papilloma without atypism (CPP WHO grade 1). Because of the intraoperative impression of ependymoma by the neurosurgeon, a postoperative spinal MRI was made, which showed no spinal metastasis. Cranial MRI and clinical follow-up were performed once every 2 years. The patient was last seen in January 2008 without any symptoms and the cranial MRI showed no residual tumour. The follow-up period was extended to 3 years, based on clinical judgement. In October 2010, she presented with intense lower back pain and right-sided radicular pain of the fifth lumbar nerve root. Neurological examination showed sensory loss in the right fifth lumbar dermatome. Spinal MRI revealed a round homogeneous contrast-enhancing intradural mass of 2 cm in length occupying the spinal canal at the level of the S1 and S2 vertebrae (figure 1B). Her cranial MRI at that time showed a relapsing tumour on the right side of the fourth ventricle with cystic degeneration and contrast enhancement. A sacral S1–3 laminectomy was performed with intradural resection of the tumour. Minor capsule residues were left on the sacral roots. The histological diagnosis was a metastasis of a CPP with atypism (WHO grade II, figure 1C,D). Postoperative spinal MRI showed no residue at the sacral level, but a new small metastatic tumour at Th9 level was seen. There was no leptomeningeal enhancement. The patient received radiotherapy with 36 Gy to the craniospinal axis followed by boosts of 50, 4 Gy to the relapsing tumour in the fourth ventricle as well as the Th9 and S1–2 regions.

OUTCOME AND FOLLOW-UP

The radicular pain disappeared when there were no further neurological complaints. In January 2012, she was in a good condition and had no neurological

Figure 1 Axial T1 MRI-cerebrum with gadolinium at initial presentation in 2004 showing an inhomogeneous contrast-enhancing tumour occupying the fourth ventricle (A). Six years after the primary posterior fossa tumour surgery sagittal spinal T1 MRI revealed an intradural tumour of 2 cm with homogeneous contrast-enhancement with gadolinium at S1–2 level (B). Photomicrograph of the spinal tumour specimen showing nuclear pleomorphism in atypical choroid plexus papillomas (CPP). H&E-stained section, original magnification $\times 630$ (C) and increased MIB labelling in atypical CPP, MIB-1-stained section, original magnification $\times 400$ (D).

complaints. Follow-up MRI of the craniospinal axis showed no change of the relapsing tumour in the fourth ventricle as well as the metastasis at Th9. Spinal MRI showed no relapsing tumour at the sacral level. The next follow-up MRI of the craniospinal axis has been arranged in 6 months.

DISCUSSION

CPP (WHO grade I) are considered to be benign and have a good prognosis if total surgical resection has been achieved. The malignant form, CPC (WHO grade III) are characterised by anaplasia, increased mitotic activity, nuclear pleomorphism and necrosis. Atypical choroid plexus papillomas (atypical CPP, WHO grade II) represent the transformation from benign to malignant. They only have few histological features of malignancy, especially increased mitotic activity. The histological grade is of prognostic significance because the 10-year survival rates following surgical resection are 77% for CPP and 35% for CPC.^{2 3} The recurrence rate of benign CPP is low, whereas in atypical CPP recurrence is more common and occurs earlier.^{2 4} In a large series of CPP and atypical CPP, tumour recurrence occurred in 6% of the patients with CPP and in 29% of the patients with atypical CPP with a median time to recurrence of 105 months and 22 months, respectively.⁴

Metastasis from a benign CPP is rare. Such metastases are particularly associated with primary tumours in the posterior fossa and occur within a few years after the

primary diagnosis.^{2 3 5} From 1980 until 2010, 22 case reports described metastasis from CPP.^{1–3 5–8} Over 75% of these cases showed spinal seeding with locoregional tumour-recurrence, similar to our case.³ The exact mechanism of seeding could have resulted from direct spread into the subarachnoid space or ependyma of the ventricles anytime before or after surgery or through tumour-spill during the primary operation.^{3 5} Atypical transformation in spinal drop metastases of CPP is even more exceptional. In the literature only one case has been reported with transformation of a spinal metastasis to an atypical CPP 19 years after surgical resection of an intracranial benign CPP.³ More recent, a case has been reported of a benign CPP in which local recurrence and atypical transformation occurred at 13 years, followed by late occurrence of an atypical spinal drop metastasis 19 years after the primary tumour was diagnosed.²

In primary CPP a good prognosis can be achieved by total resection. The extent of tumour resection is a significant prognostic factor. Subtotal resection can confer long-term control, but is also attended with a higher relapsing risk in 5 years.¹ It is recommended to routinely perform a postoperative MRI to confirm the extent of tumour removal.^{1 3} In the case of a residual tumour, repeat surgery is the best treatment option in order to facilitate total resection. There is no role for chemotherapy or radiotherapy in the treatment of benign CPP.^{1–3}

In all patients with a benign CPP, after total as well as subtotal resection, follow-up MRI of the entire neuraxis is

advised in search for locoregional recurrence and spinal and/or leptomeningeal metastasis. With frequent craniospinal follow-up MRI, recurrences and metastatic disease can be detected as early as possible and treatment can be administered.^{1 2 4}

With metastatic CPP the treatment of choice is total surgical resection, when possible.¹ After subtotal excision of a solitary metastasis either a wait-and-scan policy or adjuvant radiation therapy is generally recommended in the literature.^{1–3 5} In one case report of diffuse metastatic atypical CPP, radiation therapy of 35 Gy to the craniospinal axis was administered with boosts of 15 Gy to the spinal drop metastases. The patient's overall survival was 41 months after initial presentation, but he died due to aspiration pneumonia.¹

This case report represents the rare course of a posterior fossa CPP that was totally resected, as was seen on the post-operative cranial MRI. Locoregional recurrence and spinal drop metastasis occurred 6 years after the first presentation. The sacral drop metastasis that was surgically removed showed transformation to an atypical CPP. Only one comparable case has been reported in the literature.³ In our patient cranial MRI follow-up was done once every 2 years. In January 2008, it was extended to 3 years based on clinical judgment. Her locoregional recurrence and solitary sacral metastasis were diagnosed in-between follow-up. With more frequent MRI follow-up the sacral metastasis might have been smaller and less attached to multiple sacral nerve roots, resulting in more radical excision, possibly influencing her overall survival. We would therefore like to advocate craniospinal MRI at the time of initial diagnosis as well as periodic follow-up MRI after total as well as subtotal resection of posterior fossa CPP in adults. We recommend follow-up MRI at least once every 2 years in typical CPP and once a year in atypical CPP, based on the medium time to recurrence in the literature.⁴

Learning points

- ▶ Choroid plexus papillomas are considered to be benign lesions, but histology is not always predictive of their behaviour. Drop metastases may appear years after diagnosis and treatment of a primary choroid plexus papillomas (CPP) in the posterior fossa in typical as well as atypical CPP.
- ▶ On first diagnosis of a choroid plexus papilloma, craniospinal MRI should be made to confirm the extent of resection as well as to exclude spinal and leptomeningeal seeding.
- ▶ Drop metastases present with neurological symptoms varying from radicular symptoms to spinal cord symptomatology.
- ▶ Craniospinal MRI follow-up seems reasonable in subtotally as well as totally resected choroid plexus papilloma, at least once every 2 years in typical CPP and once a year in atypical CPP. This recommendation is based on the recurrence rates and time to recurrence depending on the WHO grade of the plexus papilloma.
- ▶ To date, no evidence-based treatment is available, but surgical resection seems to be the treatment of choice, repeatedly if necessary. Adjuvant radiotherapy is generally recommended for incompletely resected metastases.

Competing interests None.

Patient consent Obtained.

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