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Short Illustrated Review

Diffuse leptomeningeal seeding from benign choroid plexus papilloma

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Summary

Choroid plexus papillomas (CPP) are rare intracranial tumours with a favourable long-term outcome after surgical excision. Although they are histologically benign, local recurrences may occasionally occur, but leptomeningeal dissemination is exceptional. We report an unusual example of a fourth ventricle choroid plexus papilloma with diffuse leptomeningeal seeding. Neither the initial tumour nor the recurrence showed malignant histological features. Treatment with systemic and intrathecal chemotherapy was ineffective in this patient. We review the literature concerning leptomeningeal dissemintation of benign choroid plexus papillomas.

Keywords: Choroid plexus papilloma; choroid plexus tumours; disseminated choroid plexus papilloma; leptomeningeal seeding.

Introduction

Choroid plexus tumours represent 0.4–0.6% of intracranial tumours at all ages [1, 26]. The World Health Organization (WHO) classification of tumours distinguishes two types, choroid plexus papilloma (benign) and the choroid plexus carcinoma (malignant). Papillomas represent 60–80% of all choroid plexus tumours. Characteristically, they are neoplasms composed of papillae formed by a single layer of cuboidal or columnar cells, overlying a layer of vascularised connective tissue, with low mitotic activity [1, 26]. Complete resection should result in a cure and recurrence of the tumour is rare. Although nodular single metastases have been occasionally reported, the occurrence of a diffuse leptomeningeal seeding from a benign CPP is exceptional. We have conducted a thorough review of the literature pertaining to this topic and present a case exemplifying the rare occurrence of diffuse leptomeningeal seeding of a benign choroid plexus papilloma.

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Literature review

The data for this review were gathered using the medline database (PubMed, http://www.ncbi.nlm.nih.gov/entrez/query.fcgi). A combination of the words "*choroid plexus papilloma*", "*metastasis*", "*metastases*", "*disseminated*" and "*seeding*" were used. Fifteen articles were initially selected and, after reading them, 14 new articles pertaining to the subject were identified and added to the selection. In a second stage, these 29 articles were carefully scrutinised and out of them 15 articles, reporting 16 patients, were finally selected. The criteria for selection were: 1) resection of a benign CPP that produced distant seeding, 2) reported after advent of computed tomography, and 3) English, French or Spanish literature.

Analysis of literature review

Our literature review included 15 articles reporting 16 patients with CPP associated with dissemination pub-

Table 1. *Reports in the post-CT period of choroid plexus papillomas that produced distant seeding*

Ref.	Age/ sex	Primary site	Type of seeding	Interval from first surgery	Treatment (primary tumour)	Treatment (2nd neoplasm)	Outcome and follow-up	Histology (2nd neoplasm)	Immunohistochemistry
Guidetti and Spallone [13]	3/F	third ventricle	intraventricular (IV ventricle)	3 years	GTR	none	N/A	none	N/A
Leys *et al.* [20]	40/M	IV ventricle	intraventricular (lateral ventricle) + LR	9 years	GTR	surgery (subtotal for LR, total for lateral ventricle mass)	18 months, progression	CPP	N/A
Girardot *et al.* [12]	30/F	Cisterna magna	intraventricular (IV ventriculo) + LR	5 years	N/A	surgery	N/A	CPP	N/A
Domingues *et al.* [6]	35/F	Foramen magnum	leptomeningeal (spinal)	concomitant	STR	none	N/A	N/A	N/A
Enomoto *et al.* [9]	46/F	CPA	parenchymal (temporal lobe contralateral)	concomitant	STR	surgery (total resection)	20 days, stable disease?	CPP (focal parenchymal infiltration)	N/A
Niikawa *et al.* [27]	38/M	IV ventricle	leptomeningeal (spinal, CPA, suprasellar)	6 years	GTR	surgery (subtotal resection) + RT	8 months, stable disease	CPCarcinoma	N/A
Shakespeare *et al.* [34]	27/F	IV ventricle	leptomeningeal (cranial and spinal) + parenchymal (cerebellum) + LR	3 years	STR	biopsy + QT and RT	13 months, stable disease	CPP	1st neoplasm: CTK+ ; GFAP± ; S100± ; VIM−
Leblanc *et al.* [17]	19/F	IV ventricle	leptomeningeal (supratentorial, infratentorial and spinal). CYSTIC appearance	concomitant	GTR	biopsy	3 years, radiological progression	CPP	1st and 2nd neoplasms: TTr+; CTK+
Talacchi *et al.* [36]	38/M	CPA	leptomeningeal (suprasellar) + LR	5 years	GTR + RT	radiosurgery	2 years, death	N/A	N/A
Irsutti *et al.* [15]	48/F	IV ventricle	leptomeningeal (suprasellar) + LR	8 years	GTR	surgery (total resection in both)	6 months, disease free	CPP (focal parenchymal infiltration)	1st and 2nd neoplasms: S100+; CEA−; CTK+; TTr+; Ki67 <4%
Valencak *et al.* [37]	33/F	IV ventricle	intraventricular (third ventricle) + leptomeningeal (spinal) + LR	2 years (LR), 3 years (third ventricle) and 4 years (spinal)	STR	surgery (local recurrence) + RT + radiosurgery + QT	42 months, stable disease after QT	CPP (increased mitotic activity and pleomorphism)	1st neoplasm: Ki67 4,1%, 2nd neoplasm: Ki67 17,2%
Jagielski *et al.* [16]	50/M	IV ventricle + third ventricle	parenchymal (temporal lobe and cerebellum) + leptomeningeal (infratentorial and spinal) + LR	4 years	STR + RT	subtotal surgery	1 year, death	CPP	1st and 2nd neoplasms: CTK+; GFAP−; VIM−
McEvoy *et al.* [25]	51/M	IV ventricle	leptomeningeal (infratentorial, suprasellar and spinal)	5 years	GTR	biopsy	N/A	CPP	1st and 2nd neoplasms: CTK+; TTr+; S100+; GFAP±; Ki67 "low"

McCall *et al.* [24]	30/F	IV ventricle	leptomeningeal (supratentorial, infratentorial and spinal) + LR. CYSTIC appearance	8 years	surgery	biopsy + QT	17 months, stable disease	CPP	N/A
McCall *et al.* [24]	22/F	IV ventricle	leptomeningeal (suprasellar and spinal) +LR	2.5 years (LR) and 5 years (leptomeningeal dissemination)	GTR	surgery (local recurrence) + RT (spinal) + radiosurgery (suprasellar) + QT	4 years, progression	CPP	N/A
Yu *et al.* [41]	30/M	posterior fossa	leptomeningeal (spinal)	19 years	GTR	subtotal surgery	2 months, stable disease?	atypical CPP (pleomorphism and increased cellularity)	2nd neoplasm: CTK+; S100+; GFAP−; thyroglobulin−; thyroid transcription factor-1−; CEA−; BerEP4−; Ki67 10%
Ortega-Martínez *et al.* (present report)	20/F	IV ventricle	leptomeningeal (infratentorial and spinal) + LR	6 years	GTR	biopsy + QT	2 months, death	CPP (focal parenchymal infiltration)	1st and 2nd neoplasms: S100+; ki67 <2%, GFAP+

CPA Cerebellopontine angle; *GTR* gross total resection; *STR* subtotal resection; *F* female; *LR* local recurrence; *M* male; *RT* radiotherapy; *QT* chemotherapy; *N/A* not available or not obtained; *CTK* citokeratin; *GFAP* glial fibrillary acid protein; *TTr* transthyretin; *VIM* vimentina; ± focal reactivity.

lished [6, 9, 12, 13, 15–17, 20, 24, 25, 27, 34, 36, 37, 41] in the post-CT period (Table 1). Before this period, in 1960, Matson and Crofton [23] published a review of 67 tumours of the choroid plexus of which 56 were CPPs and 11 choroid plexus carcinomas. They encountered 6 instances of dissemination of the CPPs, one of them with malignant progression. The majority of patients operated on for these tumours in the pre-CT period showed severe deficits and seeding, when it occurred, was demonstrated only when post-mortem examination was undertaken. In 1970 Rovitt *et al.* [32] reviewed 234 previous CPPs and included 11 patients of their own. They noted that in 3.7% the tumours were multiple, presenting simultaneously in more than one site, but no data about dissemination were given.

In 1977 and 1981 Wolfson and Brown [40] and Masuzawa *et al.* [22], respectively, reported two new examples of disseminated CPPs in which ultrastructural studies were performed in both primary and secondary neoplasms. They found that ultrastructure was quite similar in the primary and the secondary tumour. The main difference was found in the capillary endothelium in that the secondary neoplasm was vascularised by vessels typical of the site of implantation and not by vessels from the choroid plexus. Light microscopy was more useful to detect any malignant change, such as the increased number of mitoses and nuclear pleomorphism that were found in the patient reported by Masuzawa *et al.* [22].

In the examples of disseminated CPPs confirmed by CT or MRI that we have collected (including the one we are reporting), the mean age at presentation of the primary tumour was 32.9 years (range between 3 and 51 years), and the interval until tumoral dissemination ranged between 0 and 19 years with a median of 5 years. In 3 patients [6, 9, 17] the dissemination was diagnosed at the same time as the primary tumour. There was a predilection for females (64.7%). All primary tumours but one [13] was located in the posterior fossa.

In 2 reports [12, 24], no data about the extent of resection was available. Gross total resection of the primary tumour was achieved in 9 patients [12, 15, 17, 20, 24, 25, 27, 36, 41] besides the patient we are reporting, and no additional treatment was applied except in the patient reported by Talacchi *et al.* [36], who received radiotherapy after the surgery. Subtotal resection of the primary tumour, achieved in 5 patients [6, 9, 16, 34, 37], was followed by additional therapy (radiotherapy) only in the patient reported by Jagielsky *et al.* [16].

Distant seeding can be subdivided into three different types, (1) diffuse leptomeningeal seeding (including drop metastasis), (2) intraparenchymal and (3) intraventricular lesions. Diffuse leptomeningeal seeding is the most frequently encountered and although generally affects the spinal cord and the infratentorial compartment, suprasellar leptomeningeal seeding has been reported in 5 patients [15, 24, 25, 27, 36]. A cystic appearance of the leptomeningeal dissemination has occurred in 2 patients [17, 24], leading to a preoperative diagnosis of cysticercosis. Intraparenchymal or intraventricular masses are more uncommon, and have been noted in 3 and 4 patients, respectively [9, 12, 13, 16, 20, 34, 37]. Local recurrence together with distant seeding has been noted in 9 patients as well as in our patient [12, 15, 16, 20, 24, 34, 36, 37].

Histopathological analysis confirmed the diagnosis of CPP in all patients and this was also the diagnosis in most of the secondary tumours examined. Only in the patient reported by Niikawa *et al.* [27] was malignant transformation of the secondary tumour recorded. In the remaining examples the same benign histology was seen, although some features of ''malignancy'', namely pleomorphism, brain infiltration, increased celullarity or mitoses, have been noted. Immunohistochemical study data were available in 7 cases [15–17, 25, 34, 37, 41] in addition to the present report.

Management of the secondary tumour was not standardised and a wide range of modalities of treatment have been used including gross total resection, subtotal resection, biopsy or no surgical intervention, alone or in combination with radiotherapy, radiosurgery and several chemotherapeutic schemes. Two patients died in the follow-up period besides the patient we are reporting [16, 36], at 2 months, 1 year and 2 years, respectively. Among the remaining fourteen patients, one of them is free of disease [15], six patients have stable disease [9, 24, 27, 34, 37, 41], and three patients are in clinical or radiological progression [17, 20, 24]. In four patients, no data about outcome and follow-up are available [6, 12, 13, 25].

Case report

This 20-year-old woman was operated on August 1997, to completely remove a tumour of the fourth ventricle (Fig. 1). Histopathological examination revealed a well differentiated CPP with absence of anaplasia, necrosis, parenchymal invasion or other features of malignancy (Fig. 2A). The MIB-1 index was less than 1%. The tumour cells were diffusely positive for S100 protein stain (Fig. 2B).

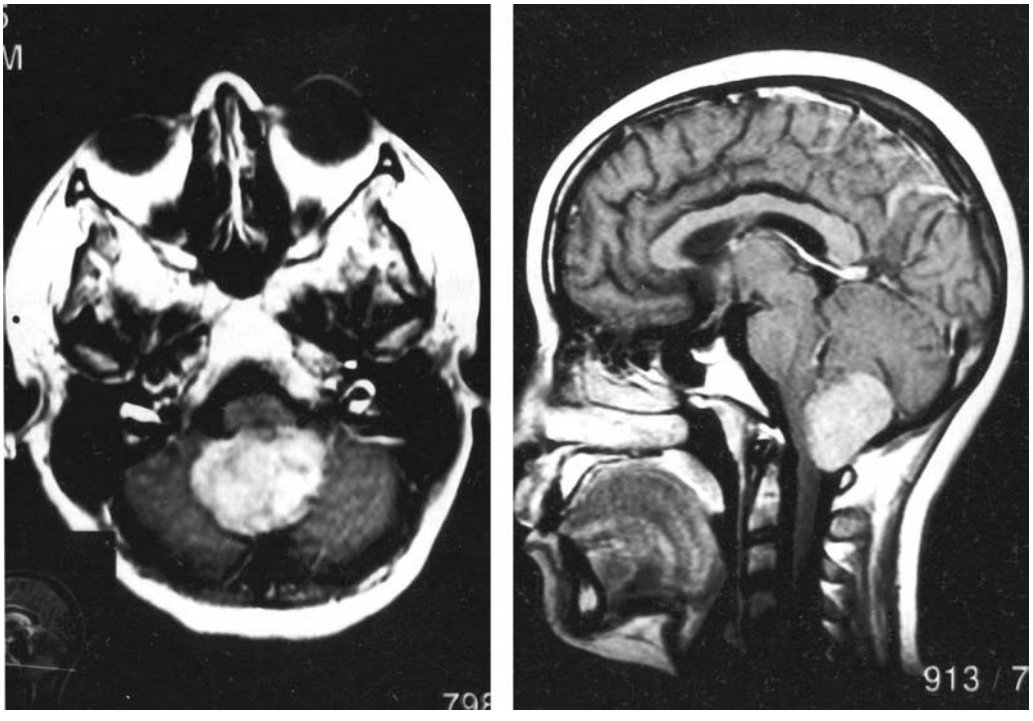

Fig. 1. Gadolinium enhanced T1-weighted MRI showing an enhancing mass in the inferior region of the fourth ventricle and foramen magnum

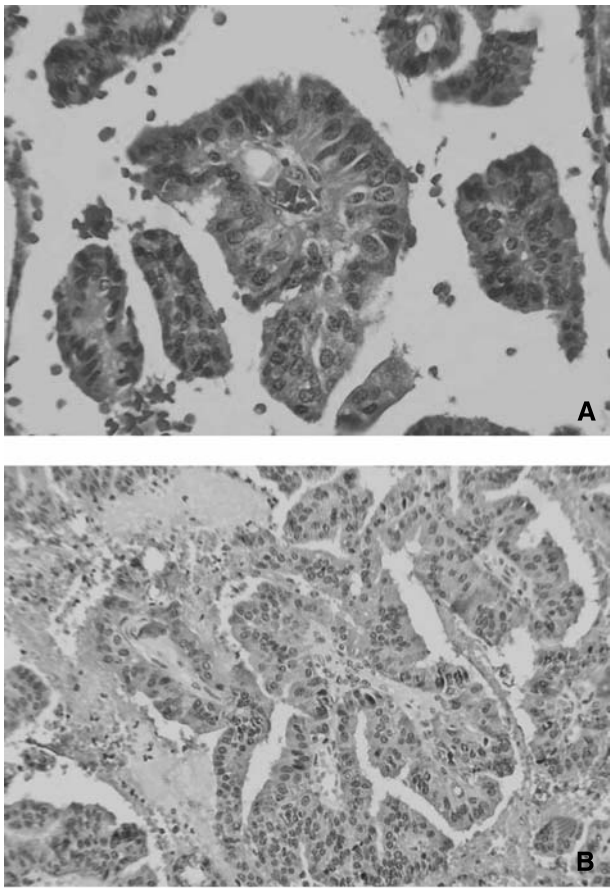

Fig. 2. Histological sections of the original lesion: (A) H/E, 40×: papillary architecture and no apparent cytological atypias, (B) S100 stain, 20×: diffuse reactivity to S100 protein

The patient was asymptomatic for six years and the routine surveillance MRI's did not show any residual tumour. In October 2003, she presented to our service with a progressive palsy in both arms, and paraesthesiae in her left leg, that progressed to a cauda equina syndrome within two months. MRI demonstrated tumour deposits in the cerebellar tonsils and obex, around both cavernous sinuses and along the dorsal surface of the cervical and thoracic spinal cord down to T3, together with cervical syringomyelia and syringobulbia (Fig. 3). Furthermore, the conus medullaris and cauda equina roots showed a diffuse meningeal enhancement and some ''drop'' metastases (Fig. 4). Believing that the tumour had evolved to a malignant form, namely to a choroid plexus carcinoma (CPC), we performed a cervical laminectomy at C7 level, in order to obtain a sample for histological examination. This again showed a well-differentiated papillary neoplasm, without mitoses or anaplasia, and a positive stain to S100 protein (Fig. 5A and B). The MIB-1 index was less than 2%. Both, the primary and the disseminated lesions, showed diffuse reactivity to GFAP (Fig. 6). Focal arachnoidal infiltration could be observed, but was not considered a definitive feature of malignancy, and the final diagnosis was CPP. As the lesion was considered unresectable, the patient was treated with chemotherapy (intrathecal Ara-C, together with systemic Cisplatinum, Bleomycin and VC 16), but this treatment could not halt the progression of the disease and she died two months after the biopsy.

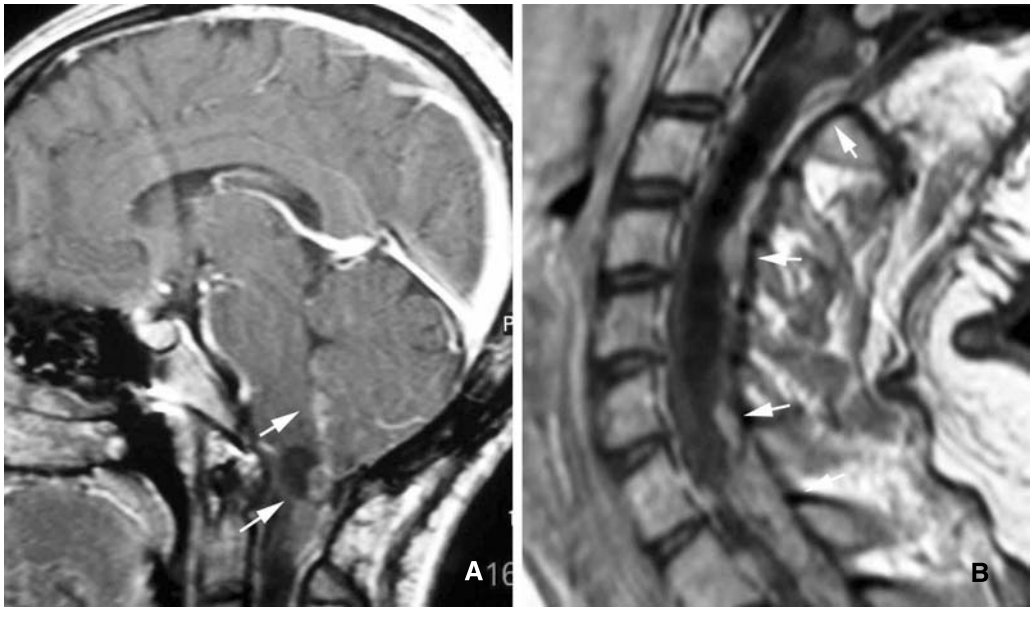

Fig. 3. Gadolinium enhanced T1-weighted MRI showing local recurrence in the cerebellar tonsils and obex (A), and tumour seeding along the dorsal surface of cervical and thoracic spine until T3, together with cervical syringomyelia and syringobulbia (B)

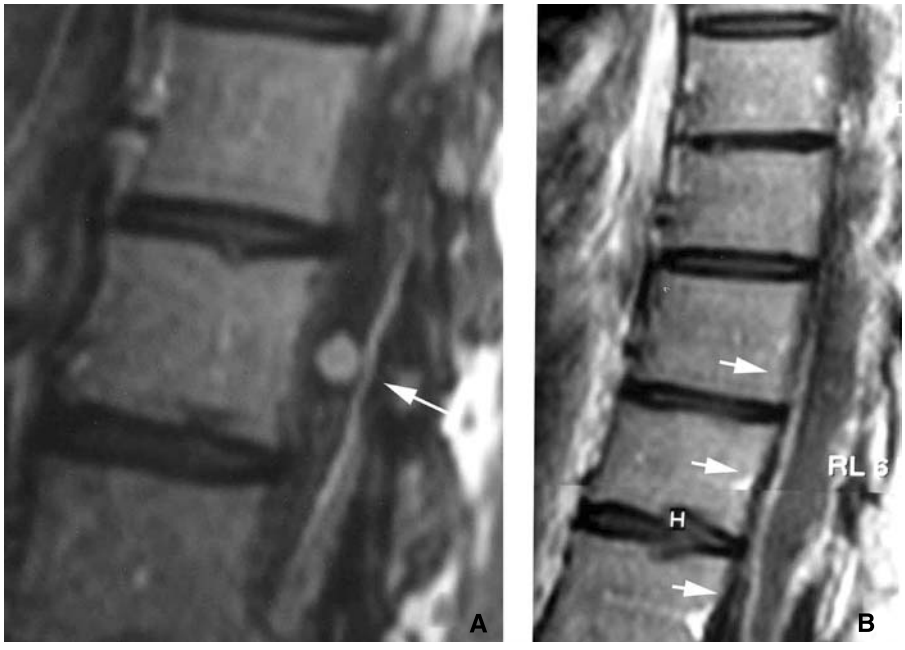

Fig. 4. Gadolinium enhanced T1-weighted MRI showing a "*drop*" metastasis (A), and diffuse leptomeningeal enhancing in the conus medullaris and cauda equina (B)

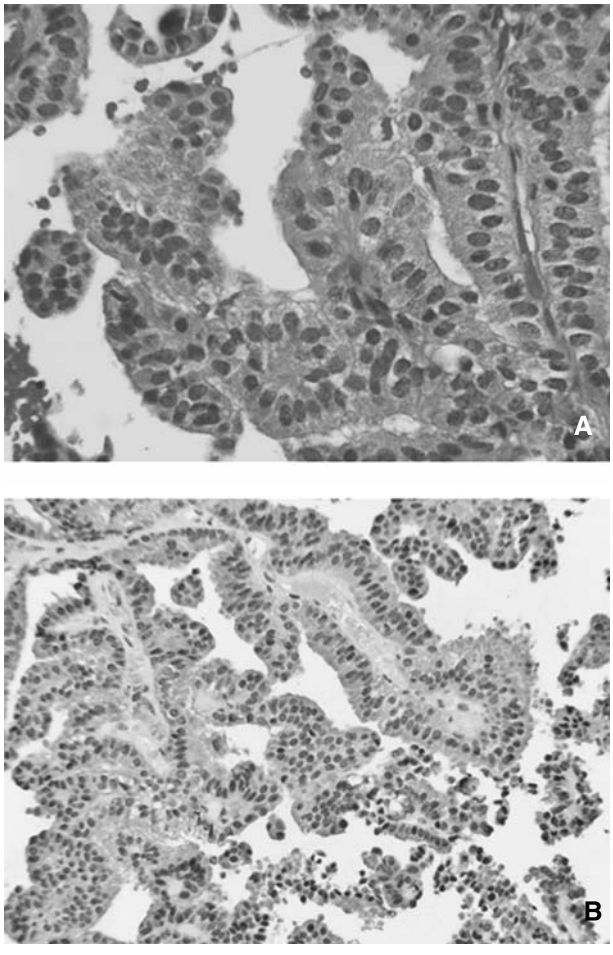

Fig. 5. Histological sections of the disseminated lesion: (A) H/E, 40×: papillary architecture and columnar epithelium, without cytological malignancy, (B) S100 stain, 20×, with diffuse reactivity, similar to the primary neoplasm

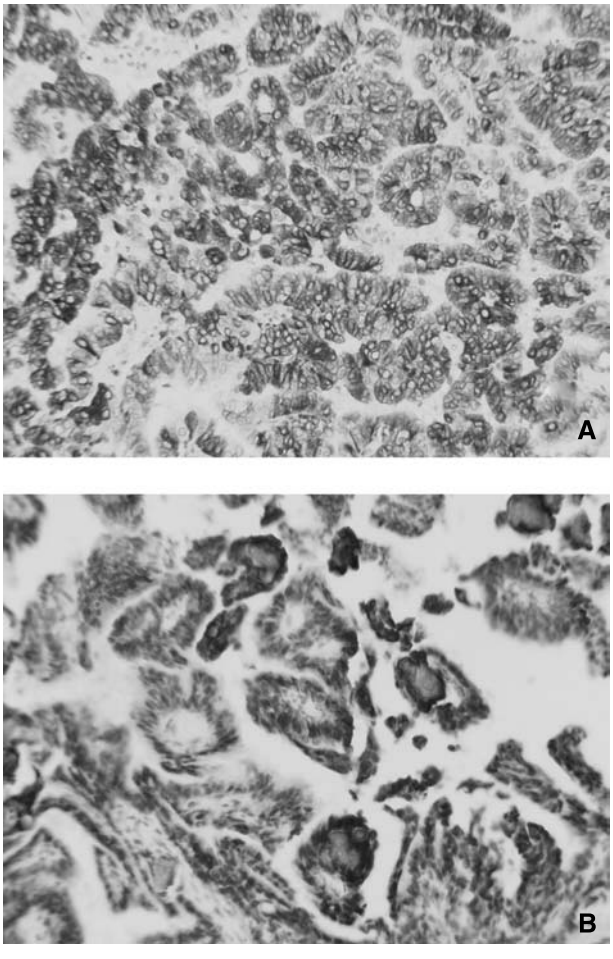

Fig. 6. Histological sections showing positive staining for GFAP in both the original (A) and (B) the disseminated lesion (GFAP stain, 20×)

Discussion

There are several accepted criteria to distinguish CPC from CPP. They were first developed by Lewis in the 1960s, later refined by Russell and Rubinstein in the 1980s, and now recoded by the World Health Organization [18, 19, 33]. These criteria are: 1) obvious invasion of adjacent neural tissue with the infiltrating cells exhibiting a poorly defined pattern of growth, 2) loss of regular papillary architecture, and 3) evidence of cellular malignancy (increased mitotic activity, nuclear atypia and necrosis). In spite of these strict criteria, the difference between choroid plexus tumours may not be clear in some instances. On the one hand, there are tumours with mixed characteristics (benign histoarchitecture, but with areas of anaplasia and mitotic figures), that are difficult to classify and have a more aggressive behaviour than CPPs [30, 29] corresponding to the ''atypical'' CPP; on the other hand, in the experience of other authors, some of these criteria, when present alone, are not associated with a worse outcome. In 1991, Paulus and Jänisch [29] reported a review of 52 patients with tumours of the choroid plexus, and identified several factors associated to local recurrence and/or fatal outcome. These factors, in order of decreasing significance, are: less than 50% of tumour cells heavily positive for S100, presence of mitosis, absence of transthyretin-positive tumour cells, brain invasion by neoplastic cell nests, absence of marked stromal oedema and presence of necrotic areas. Others features, usually associated with malignancy, such as nuclear pleomorphism, blurred papillary structure or increased cellular density, did not correlate with a more aggressive behaviour, and the presence of necrosis or brain invasion occupied a secondary place.

Other reports, designed to identify prognostic factors in choroid plexus tumours, have confirmed the prognostic importance of the S100 protein, and have also shown the prognostic value of the MIB-1 labelling index that was 3.7% for the papilloma group, and 14% for carcinomas [35, 38]. In fact, there are several reports in the literature that show malignant evolution from CPPs to choroid plexus carcinomas, in which the presence of mitoses was the only feature of malignancy in the first specimen [4, 5, 14].

In 2001, Levy *et al.* [18] found that out of 12 CPPs they had operated upon, 4 showed parenchymal infiltration by the tumour. Although these patients were treated only with surgical resection, they did not show a different evolution. They conclude that parenchymal infiltration by itself should not be considered as a criterion of malignancy.

The importance of determining as exhaustively as possible the tumour type and identifying unfavourable prognostic factors is related to the need to determine the best treatment protocol in each individual case. While CPCs need adjuvant therapy with chemotherapy and/or radiotherapy, CPPs should be managed with surgery alone [25, 30, 39]. When poor prognostic indicators can be identified in a CPP that has been totally removed, the patient should have a close follow-up, but if there is any residual tumour, adjuvant therapies should be undertaken.

We report a patient with a CPP in which histological features of poor prognosis were absent but exhibited a malignant behaviour. This has been reported occasionally in the literature, and, curiously, almost all secondary tumours retained a benign histology; only some scarce signs of malignancy can be detected when compared with the primary tumour, namely, slight increase in mitoses or celullarity, focal parenchymal infiltration or nuclear pleomorphism, that occasionally lead to the diagnostic of ''atypical'' CPP. Only in one patient did the tumour evolve to a malignant CPC [27].

Management of these disseminated CPPs is based in the experience obtained from isolated clinical reports. In our patient, the diffuse leptomeningeal seeding prevented surgical removal. In view of several reports that had shown a good response to chemotherapy for CPCs with local recurrence or with subtotal resection [2, 3, 7, 8, 10, 11, 28, 31, 37], and for CPPs with multiple local recurrences [21], we decided to treat our patient with Ara-C intrathecal together with systemic Bleomycin, Cisplatinum and VC-16, but could not induce tumour regression.

Conclusions

It is accepted that when total surgical resection can be performed in CPPs there is no need of adjuvant therapy. Dissemination of this tumour is exceptional and, in those instances, treatment is based on isolated experiences. Chemotherapy, effective in some choroid plexus tumours, could not induce any response in our patient. With this report, we want to demonstrate a rare behaviour of these benign tumours, adding our experience to the literature about leptomeningeal dissemination of low grade neoplasms.

References

1. Aguzzi A, Brandner S, Paulus W (2000) Choroid plexus tumours. In: Kleihues P, Cavenee WK (eds) World Health Organization classification of tumours. Pathology and genetics: tumours of the nervous system. IARC Press, Lyon, pp 84–86
2. Arico M, Raiteri E, Bossi G, Giordana MT, Corbella F, Locatelli D, Pezzota S (1994) Choroid plexus carcinoma: report of one case

with favourable response to treatment. Med Pediatr Oncol 22: 274–278
3. Berger C, Thiesse P, Lellouch-Tubiana A, Kalifa C, Pierre-Kahn A, Bouffet E (1998) Choroid plexus carcinomas in childhood: clinical features and prognostic factors. Neurosurgery 42: 470–475
4. Burger PC, Scheitahuer B (1993) Tumours of the central nervous system. Atlas of tumour pathology. Armed Forces Institute of Pathology, Washington, DC, pp 394–404
5. Chow E, Jenkins JJ, Burger PC, Reardon DA, Langston JW, Sanford RA, Heideman RL, Kun LE, Merchant TE (1999) Malignant evolution of choroid plexus papillomas. Pediatr Neurosurg 31: 127–130
6. Domingues RC, Taveras JM, Reimer P, Rosen BR (1991) Foramen magnum choroid plexus papilloma with drop metastases to the lumbar spine. Am J Neuroradiol 12: 564–565
7. Duffner PK, Kun LE, Burger PC, Horowitz ME, Sanford RA, Krisher JP, Mulhern RK, James HE, Rekate HL, Friedman HS (1995) Postoperative chemotherapy and delayed radiation in infants and very young children with choroid plexus carcinomas. Pediatr Neurosurg 22: 189–196
8. Ellenbogen RG, Winston KR, Kupsky WJ (1989) Tumors of the choroid plexus in children. Neurosurgery 25: 327–335
9. Enomoto H, Mizuno M, Katsumata T, Doi T (1991) Intracranial metastasis of a choroid plexus papilloma originating in the cerebellopontine angle region: a case report. Surg Neurol 36: 54–58
10. Fiorillo A, Maggi G, Cirillo S, Migliorati R, Buffardi F, Alfieri E, Sabbatino MS, D'Amico A, DelBasso DeCaro ML (2003) Efficacy of sequential chemotherapy including methotrexate and doxorubicin in an infant with partially resected choroid plexus carcinoma. Pediatr Neurosurg 38: 21–26
11. Gianella-Borradori A, Zeltzer PM, Bodey B, Nelson M, Britton H, Marlin A (1992) Choroid plexus tumours in childhood. Response to chemotherapy, and immunophenotypic profile using a panel of monoclonal antibodies. Cancer 69: 809–816
12. Girardot C, Boukobza M, Lamoureux JP, Sichez JP, Capelle L, Zouaoui A, Bencherif B, Metzger J (1990) Choroid plexus papillomas of the posterior fossa in adults: MR imaging and gadolinium enhancement. J Neuroradiol 17: 303–318
13. Guidetti B, Spallone A (1981) The surgical treatment of choroid plexus papillomas. Results of 27 years experience. Neurosurg Rev 4: 129–137
14. Gullotta F, de Melo AS (1979) Das Karzinom des plexus chorioideus. Klinische lichtmikroskopische und elektronenoptische Untersuchungen. Neurochirurgia 22: 1–9
15. Irsutti M, Thorn-Kany M, Arrue P, Richaud J, Sol JC, Delisle MB, Manelfe C (2000) Suprasellar seeding of a benign choroid plexus papilloma of the fourth ventricle with local recurrence. Neuroradiology 42: 657–661
16. Jagielski J, Zabek M, Wierzba-Bobrowicz T, Takomiec B, Chrapusta SJ (2001) Disseminating histologically benign multiple papilloma of the choroid plexus: case report. Folia Neuropathol 39: 209–213
17. Leblanc R, Bekhor S, Melanson D, Carpenter S (1998) Diffuse craniospinal seeding from a benign fourth ventricle choroid plexus papilloma: case report. J Neurosurg 88: 757–760
18. Levy ML, Goldfarb A, Hyder DJ, Gonzales-Gomez I, Nelson M, Gilles FH, McComb JG (2001) Choroid plexus tumours in children: significance of stromal invasion. Neurosurgery 48: 303–309
19. Lewis P (1967) Carcinoma of the choroid plexus. Brain 90: 177–186
20. Leys D, Pasquier F, Lejeune JP, Lesoin F, Petit H, Delandsheer JM (1986) Papillome bénin des plexus choroïdes. Deux récidives locales et essaimage intraventriculaire. Neurochirurgie 32: 258–261
21. Maria BL, Graham ML, Strauss LC, Wharam MD (1985) Response of a recurrent choroid plexus tumour to combination chemotherapy. J Neurooncol 3: 259–262
22. Masuzawa T, Shimabukuro H, Yoshimizu N, Sato F (1981) Ultrastructure of disseminated choroid plexus papilloma. Acta Neuropathol (Berl) 54: 321–324
23. Matson DD, Crofton FDL (1960) Papilloma of the choroid plexus in childhood. J Neurosurg 17: 1002–1027
24. McCall T, Binning M, Blumenthal DT, Jensen RL (2006) Variations of disseminated choroid plexus papilloma: 2 case reports and a review of the literature. Surg Neurol 66: 62–67
25. McEvoy AW, Galloway M, Revesz T, Kitchen ND (2002) Metastatic choroid plexus papilloma: a case report. J Neurooncol 56: 241–246
26. McLendon RE, Enterline DS (1998) Tumors of specialized tissues of central neuroepithelial origin. In Bigner DD, McLendon RE, Bruner JM (eds) Russell & Rubinstein's. Pathology of tumours of the nervous system, Vol. 2, Arnold, London, pp 37–65
27. Niikawa S, Ito T, Murakawa T, Hirayama H, Ando T, Sakai N, Yamada H (1993) Recurrence of choroid plexus papilloma with malignant transformation: case report and lectin histochemistry study. Neurol Med Chir (Tokyo) 33: 32–35
28. Packer RJ, Perilongo G, Johnson D, Sutton LN, Vezina G, Zimmerman RA, Ryan J, Reaman G, Schut L (1992) Choroid plexus carcinoma of childhood. Cancer 69: 580–585
29. Paulus W, Jänisch W (1990) Clinicopathological correlations in epithelial choroid plexus neoplasms: a study of 52 cases. Acta Neuropathol 80: 635–641
30. Pencalet P, Sainte-Rose C, Lellouch-Tubiana A, Kalifa C, Brunelle F, Sgouros S, Meyer P, Cinalli G, Zerah M, Pierre-Kahn A, Renier D (1998) Papillomas and carcinomas of the choroid plexus in children. J Neurosurg 88: 521–528
31. Pierga JY, Kalifa C, Terrier-Lacombe MJ, Habrand JL, Lemerele J (1993) Carcinoma of the choroid plexus: a paediatric experience. Med Pediatr Oncol 21: 480–487
32. Rovit RL, Schechter MM, Chodroff P (1970) Choroid plexus papillomas. Observations on radiographic diagnosis. Am J Roentgenol Radium Ther Nucle Med 110: 608–617
33. Russell DS, Rubinstein LJ (1989) Papillomas and carcinomas of the choroid plexus. Pathology of tumours of the nervous system. Arnold, London, pp 394–404
34. Shakespeare TP, Slancar MM, Mallik AR, Bell DR (1997) CSF dissemination of a benign choroid plexus papilloma. Aust NZ J Med 27: 597–598
35. Shirakawa N, Kannuki S, Matsumoto K (1994) Clinicopathological study on choroid plexus tumors: immunohistochemical features and argyrophilic nucleolar organizer regions values (Abstract). Noshuyo Byori 11: 99–105
36. Talacchi A, De Micheli E, Lombardo C, Turazzi S, Bricolo A (1999) Choroid plexus papillomas of the cerebellopontine angle: a twelve patient series. Surg Neurol 51: 621–629
37. Valencak J, Dietrich W, Raderer M, Dieckmann K, Prayer D, Hainfellner JA, Marosi C (2000) Evidence of therapeutic efficacy of CCNU in recurrent choroid plexus papillomas. J Neurooncol 49: 263–268
38. Varga Z, Vajtai I (1998) Prognostic markers in the histopathological diagnosis of tumors of the choroid plexus (Abstract). Orv Hetil 139: 761–765
39. Wolff JE, Sajedi M, Brant R, Coppes MJ, Egeler RM (2002) Choroid plexus tumours. Br J Cancer 87: 1086–1091
40. Wolfson WL, Brown WJ (1977) Disseminated choroid plexus papilloma. An ultrastructural study. Arch Pathol Lab Med 101: 366–368
41. Yu H, Yao TL, Spooner J, Stumph JR, Hester R, Konrad PE (2006) Delayed occurrence of multiple spinal drop metastases from a posterior fossa Choroid plexus papilloma: case report. J Neurosurg Spine 4: 494–496

Comments

The authors present the case of a 20-year old female who underwent gross total resection of a ''benign'' choroid plexus papilloma only to represent 6 years later with diffuse leptomeningeal seeding. Biopsy of the cervical deposit showed well differentiated papillary tumour without histologic evidence of malignant progression. In view of the extensive nature of the recurrence, chemotherapy was offered but unfortunately the patient succumbed 2 months after the biopsy. The literature concerning this curious behaviour was carefully reviewed using the Medline data base.

Although a number of factors were associated with a worse outcome, reduced staining for S100 protein appeared to be consistently so. Interestingly classic features associated with malignant progression e.g. parenchymal infiltration, the presence of necrosis, nuclear pleomorphism and increased cellularity were less important in this regard. A higher labelling index was also associated with a poorer prognosis.

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Ortega-Martinez *et al.* present an illustrated review of a choroid plexus papilloma with unexpected late metastases/seeding leading to poor outcome.

The paper also summarizes similar cases found in the literature and provides an excellent clinico-pathological summary of such cases.

The manuscript highlights the difficulties of drawing a sharp line between choroid plexus papillomas and carcinomas stating that almost all secondary appearances that have been reviewed displayed benign histopathological characteristics.

András Büki
Pecs

This is a well written and interesting case report on a patient with wide dissemination of an apparently well-differentiated choroid plexus tumour. The literature is well reviewed and the problem of distinguishing choroid papillomas from choroid plexus carcinoma and metastatic adenocarcinoma are discussed.

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