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M. Irsutti
M. Thorn-Kany
P. Arrué
J. Richaud
J. C. Sol
M. B. Delisle
C. Manelfe

Suprasellar seeding of a benign choroid plexus papilloma of the fourth ventricle with local recurrence

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C. Manelfe (✉) · M. Irsutti ·
M. Thorn-Kany · P. Arrué
Service de Neuro-Radiologie Diagnostique et Interventionnelle, Centre Hospitalier Universitaire Purpan, Place du Dr Baylac, 31059 Toulouse Cedex, France,
Tel.: (33) 561 77 23 39
Fax: (33) 561 77 76 51

J. Richaud · J. C. Sol
Service de Neurochirurgie,
Centre Hospitalier Universitaire Rangueil, Toulouse, France

M. B. Delisle
Laboratoire d'Anatomie et de Cytologie Pathologiques, Centre Hospitalier Universitaire Rangueil, Toulouse, France

Abstract A suprasellar location of a benign choroid plexus papilloma is reported. Local recurrence within the fourth ventricle was also present, 8 years after apparently complete removal. Imaging and histological findings were similar to those of the initial lesion. At surgery, the suprasellar lesion had no connection with the ventricular system. Seeding of choroid plexus papillomas is discussed, and the pertinent literature reviewed.

Key words Papilloma, choroid plexus · Tumour seeding

Introduction

Choroid plexus papillomas (CPP) are benign neoplasms which represent less than 5 % of intracranial tumours in children and are even less common in adults. They mainly arise from the choroid plexus of the ventricular system. Symptoms induced by hydrocephalus are the most frequent mode of presentation, and surgical excision is usually curative. Local recurrence and distant seeding along the cerebrospinal fluid (CSF) pathways have been observed, but, to our knowledge, an isolated suprasellar recurrence of a benign CPP has not been documented. We describe clinical, imaging, surgical and pathological features of such a case in an adult.

Case report

A woman was referred at the age of 48 years because of acutely increased intracranial pressure. CT and MRI revealed hydrocephalus related to a contrast-enhancing tumour of the fourth ventricle (Fig. 1). Total surgical resection and ventriculoperitoneal shunting were performed. Histologically, a regular single layer of cells overlying a thin fibrovascular stroma was observed, with no mitotic activity or atypia; some calcification was present. The lesion stained positively for S-100 protein and negatively for carcinoembryonic antigen (CEA); it was also positive for cytokeratin and transthyretin (prealbumin). Proliferative activity as measured by MIB-1 labeling index was less than 4 %. The diagnosis was a benign CPP. The sellar and suprasellar regions were normal. Mild ataxia was the only residual symptom. MRI after 1 and 3 years showed no recurrence.

Eight years after surgery, the patient complained of diplopia, visual loss and exacerbation of the cerebellar signs. CT revealed a dense tumour of the posterior cranial fossa, with a small calcified

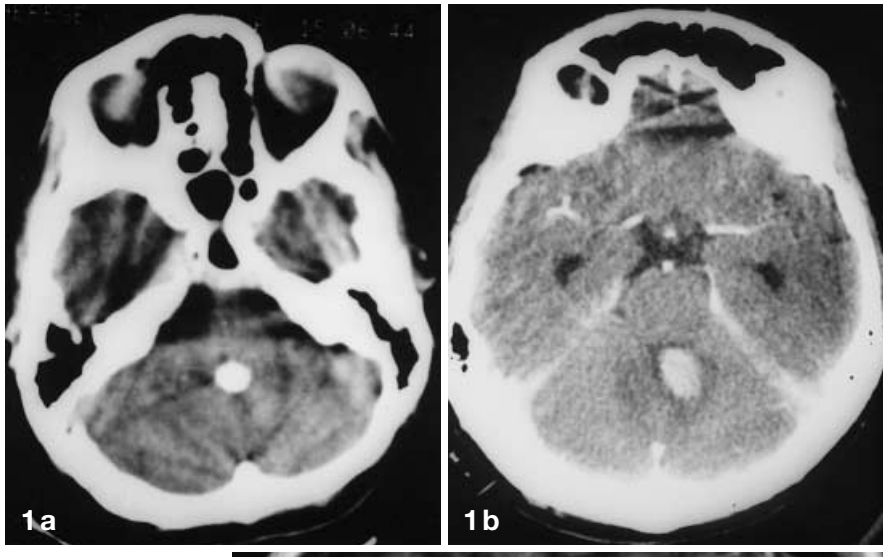
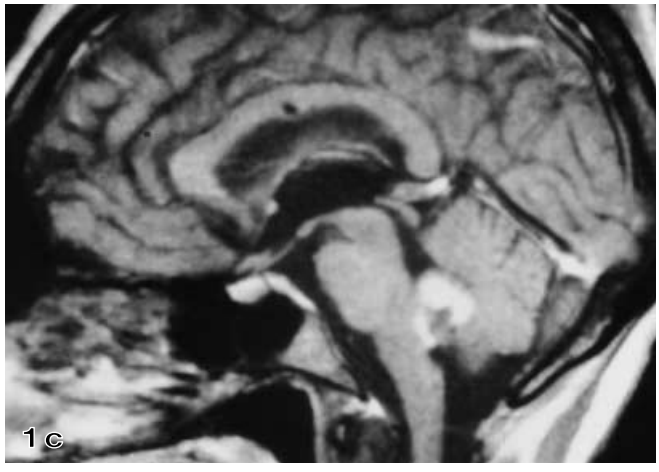


Fig. 1a–c CT and MRI at initial presentation. **a** precontrast CT showing a calcified mass in the fourth ventricle. **b** Contrast-enhanced image shows homogeneous enhancement. No abnormality is observed in the suprasellar region. **c** Sagittal contrast-enhanced T1-weighted image demonstrating a well-delineated, enhancing tumour of the fourth ventricle. Sellar and suprasellar areas are normal



nodule (Fig. 2a). A round, well-circumscribed, partially calcified 1 cm diameter lesion was seen in the suprasellar region (Fig. 2b). MRI was highly suggestive of recurrence within the fourth ventricle, with a soft-tissue contrast-enhancing nodule and a cyst. The midline suprasellar lesion was adjacent to the floor of the third ventricle and the pituitary stalk, and caused mild anterior displacement of the optic chiasm. Its signal was similar to that of the solid component of the fourth ventricle tumour on all sequences: isointense to the brain parenchyma on T1- (Fig. 3a), with moderately increased signal intensity on T2-weighted images (Fig. 3b). It showed marked, homogeneous contrast enhancement (Fig. 3c, d). Review of the previous imaging confirmed the normality of the suprasellar region at the time of first admission, which made the diagnosis of craniopharyngioma unlikely. On the basis of the history and radiological findings, a cisternal recurrence of the papilloma was considered most likely.

The patient first underwent new surgical procedure on the posterior fossa. The tumour was adherent to the floor of the fourth ventricle, but was totally removed. The lesion showed some tumour foci within brain tissue and some areas of pseudostratification, but these changes were not considered definite features of malignancy. Immunohistochemical findings were similar to those of the initial tumour. Postoperative permanent palsies of the left sixth and seventh nerves were observed, with improved but persisting ataxia.

Examination 4 months later revealed significant loss of visual acuity, but no visual field defect. A left pterional craniotomy was performed. The tumour was soft, pale, located above the diaphragma sellae, slightly adherent to the floor of the third ventricle and the pituitary stalk. There was no intraventricular component. Gross total resection was achieved. Histopathological and immunohistochemical features were identical to those of both previous lesions (Fig. 4a, b). MRI 6 months later, including images of the spine, showed no recurrence or distant seeding.

Discussion

Papillomas arise from the epithelium of the choroid plexus. The main site is the lateral ventricle in children and the fourth in adults. More unusual sites are the third ventricle and cerebellopontine angle, including primary extraventricular forms and extension from the lateral recess of the fourth ventricle [1]. The histopathological structure of papillomas classically mimics that of normal choroid plexus, as in our case. The immunohistochemical features were characteristic of a benign lesion. Cytokeratin and transthyretin immunopositivity, although nonspecific [2], was highly suggestive of a CPP. The malignant form, choroid plexus carcinoma, is very uncommon and shows evidence of cellular stratification, mitoses, cytonuclear atypia, rupture of the basal membrane and invasion of adjacent structures. However, parenchymal infiltration is reported in some cases of benign CPP [3, 4].

Choroid plexus papillomas and carcinomas are not distinguishable by imaging. In our case, the CPP of the fourth ventricle showed features similar to those in the literature.

In situ recurrence of a benign papilloma is unusual. It is more common in the malignant form, as par-

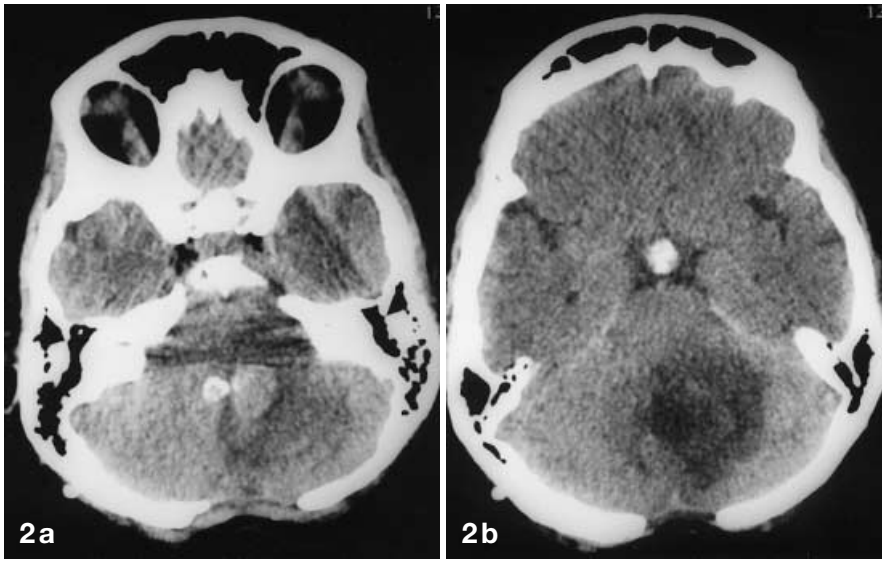


Fig. 2 a, b Precontrast CT scan 8 years later. **a** Local recurrence in the posterior cranial fossa. A low density in the vermis and left cerebellar hemisphere is consistent with oedema. **b** Well-circumscribed, partially calcified 1 cm diameter rounded lesion in the suprasellar region. There is no hydrocephalus, due to the shunt

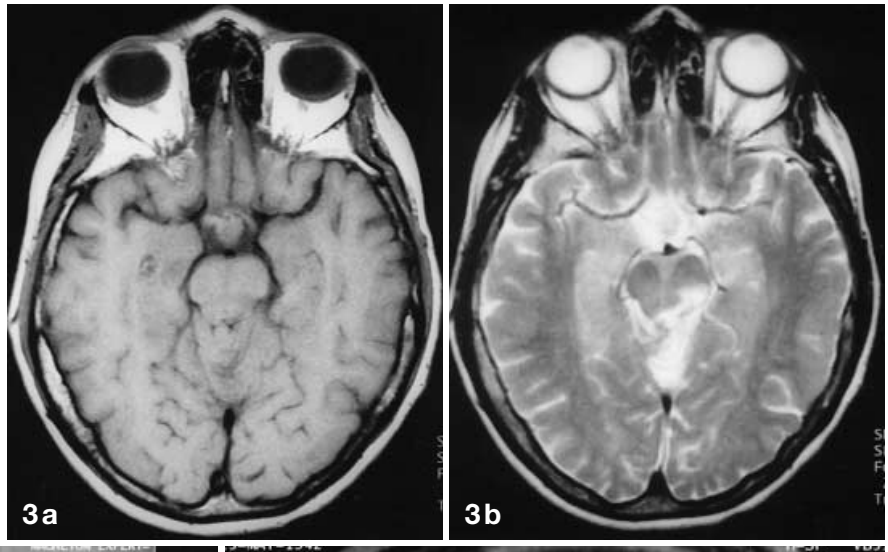
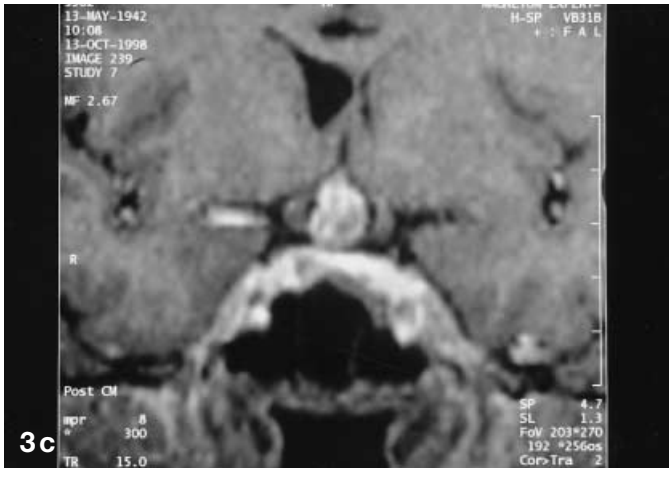
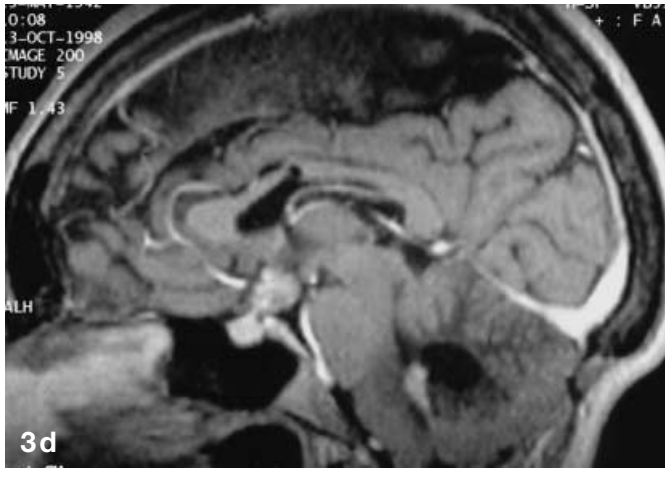


Fig. 3 **a** Axial T1-weighted image: a nodule isointense to the brain, is seen in the suprasellar region. **b** Axial T2-weighted image showing the lesion to give moderately increased signal. Postoperative changes in the posterior midbrain and vermis. **c, d** Coronal and sagittal contrast-enhanced T1-weighted images. The tumour, demonstrating marked, homogeneous enhancement, is adjacent to the floor of the third ventricle, and seems to be independent of the pituitary gland. It is adjacent to the pituitary stalk and posterior to the optic chiasm. The tumour in the fourth ventricle is composed of an anterior soft-tissue nodule and a posterior non-enhancing cyst





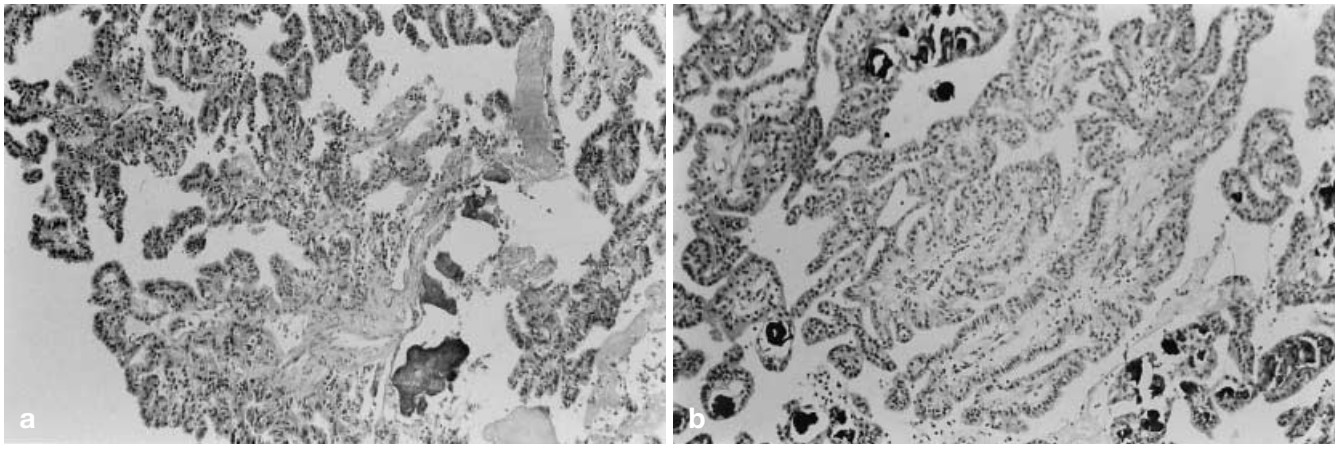


Fig. 4a,b Photomicrographs of the initial papilloma of the fourth ventricle (**a**) and the suprasellar lesion (**b**) showing a regular, single layer of cells overlying thin fibrovascular stroma. No evidence of mitotic activity or atypia. Some calcification is present (original magnification × 160)

enchymal infiltration often precludes entire resection. In the series of Guidetti and Spallone [5] including 27 patients, only two developed local recurrence, of whom one had a malignant form. Leys et al. [6] described two recurrences of the same benign fourth ventricle papilloma, 5 and 9 years after removal. Case 6

Table 1 Reported cases of distant seeding of benign CPP

Primary tumour Age (years) at admission	Site	Local recurrence	Secondary tumour(s)		Histology	Treatment	Follow-up	Reference
			Interval	Site				
Single location								
3	Third ventricle	No	6 months	Fourth ventricle	None	None	3 years	[5]
40	Fourth ventricle	Twice	9 years	Lateral ventricle	Benign	Surgery	18 months	[6]
46	Cerebellopontine angle		Concomitant	Basal cisterns and temporal lobe	Benign and infiltrating	Surgery	20 days	[4]
48	Fourth ventricle	Yes	8 years	Suprasellar	Benign	Surgery		This report
Multiple locations								
11	Fourth ventricle		5 years	Spinal subarachnoid space	Benign	Surgery (repeated)	3 years (death)	[10]
19	Fourth ventricle	None	Concomitant	Cranial and spinal subarachnoid space, leptomeninges	Benign	None	3 years	[13]
27	Fourth ventricle	Yes	3 years	Cranial and spinal subarachnoid space, cerebellum	Benign	Chemo- and radiotherapy	13 months	[12]
35	Foramen magnum		Concomitant	Lumbar subarachnoid space	None			[11]
38	Fourth ventricle	None	6 years	Suprasellar, cerebello pontine angle	Malignant	Subtotal resection, radiotherapy	8 months	[8]
				Lumbar subarachnoid space	None	Radiotherapy		
38	Fourth ventricle, cerebellopontine angle	Yes	5 years	Suprasellar	None	Radiosurgery	2 years (death)	[7]

of Talacchi et al. [7] showed both local recurrence in the cerebellopontine angle and a probable suprasellar metastasis [7]. In all these cases, initial surgery was reported as radical.

CPP are known to seed along the CSF pathways, but this occurs quite rarely (Table 1). In the case of Talacchi et al. [7], a suprasellar metastasis of a cerebellopontine angle papilloma was suspected, but to our knowledge, not documented histologically. Niikawa et al. [8] described a case resembling ours, but the secondary suprasellar lesion proved to be malignant, and was not isolated. Distant seeding usually occurs several months or years postoperatively. Guidetti and Spallone [5] suggested that one use cottonoid packs to isolate the tumour from the rest of the operative field, as they believed this might minimise the risk of seeding. In many cases of disseminated CPP, the spinal subarachnoid space is involved [8–13]. These lesions are not always symptomatic. We suggest that one routinely perform sagittal contrast-enhanced T1-weighted MRI of the spine at follow-up examinations.

Regarding the imaging findings, differential diagnosis for the suprasellar tumour in our case was craniopharyngioma, but this was unlikely, considering the absence of abnormality on recent previous MRI and the lack of cysts. A pseudotumoural form of neurosarcoidosis would usually be associated with diffuse leptomeningeal infiltration. Langerhans-cell histiocytosis was very improbable, given the clinical background. Meningioma was not considered, as there was no radiological evidence of attachment to the dura mater. Primary suprasellar CPP has been reported [14]. In our case, this possibility could not be excluded, but was unlikely, as multifocal papillomas are even more uncommon than distant seeding.

Management of the suprasellar lesion was difficult: according to the imaging findings, the optic chiasm showed minor anterior shift, but there was no evidence of compression of the intracranial optic nerves. Therefore, it was debatable whether it was the tumour that affected visual acuity. However, surgery was chosen, considering that the lesion could grow and become more difficult to resect.

References

1. Jackson A, Panizza BJ, Hughes D, Reid H (1992) Primary choroid plexus papilloma of the cerebellopontine angle: magnetic resonance imaging, computed tomographic and angiographic appearances. Br J Radiol 65: 754–757
2. Scheithauer BW (1992) Comments on "Primary choroid plexus papilloma located in the suprasellar region: Case Report". Neurosurgery 31: 563–566
3. Girardot C, Boukobza M, Lamoureux JP, et al (1990) Choroid plexus papillomas of the posterior fossa in adults: MR imaging and gadolinium enhancement. Report of four cases and review of the literature. J Neuroradiol 17: 303–318
4. 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
5. Guidetti B, Spallone A (1981) The surgical treatment of choroid plexus papillomas. Neurosurg Rev 4: 129–137
6. 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
7. Talacchi A, De Micheli E, Lombardo C, Turazzi S, Bricolo A (1999) Choroid plexus papilloma of the cerebellopontine angle: a twelve patient series. Surg Neurol 51: 621–629
8. Niikawa S, Ito T, Murakawa T, et al (1993) Recurrence of choroid plexus papilloma with malignant transformation. Case report and lectin histochemistry study. Neurol Med Chir 33: 32–35
9. Masuzawa T, Shiwabukuro H, Yoshimizu N, Sato F (1981) Ultrastructure of disseminated choroid plexus papilloma. Acta Neuropathol 54: 321–324
10. Wolfson WL, Brown WJ (1977) Disseminated choroid plexus papilloma. An ultrastructural study. Arch Pathol Lab Med 101: 366–368
11. 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
12. Skakespeare TP, Slancar MM, Mallik AR, Bell DR (1997) CSF dissemination of a benign choroid plexus papilloma (CPP). Aust NZJ Med 27: 597–598
13. Leblanc R, Bekhor S, Melanson D, Carpenter S (1998) Diffuse craniospinal seeding from a benign fourth ventricle choroid plexus papilloma. J Neurosurg 88: 757–760
14. Kimura M, Takayasu M, Suzuki Y, et al (1992) Primary choroid plexus papilloma located in the suprasellar region: case report. Neurosurgery 31: 563–566