Paratrigonal choroid plexus papilloma presenting with satellite multiple supra- and infratentorial hemorrhages. Neuroanatomical basis and pathological hypothesis

G. Maimone a,⁎, M. Ganaub, N. Nicassio b, S. Paterniti a

a Department of Neurosurgery, Policlinic University of Messina, 98126 Messina, Italy
b Department of Neurosurgery, Hospital “Ospedali Riuniti - Cattinara”, 34149 Trieste, Italy

Abstract

Introduction: Choroid plexus papillomas (CPP) are rare tumors arising from the neuroepithelium of the choroid plexus.

Presentation of Case: We report a case of a patient operated for a paratrigonal hemorrhagic WHO class I CPP presenting with multiple satellite supra- and infratentorial hemorrhages. Clinical presentation was characterized by sudden hemiparesis, speech impairment and consciousness deterioration; neuroradiological imaging showed a huge contrast-enhanced solid hemorrhagic left paratrigonal lesion along with others multifocal right occipital and vermis hemorrhages. The patient underwent urgent intervention for excision of the paratrigonal lesion, whose histological analysis led to the diagnosis of CPP. A few days later due to failure of conservative treatment of the satellite hemorrhages the patient underwent a second-time surgery for their evacuation; interestingly histological examination of the tissue probe did not reveal any neoplastic features confirming their sole hemorrhagic nature. Patient’s conditions slowly improved despite severe neurological deficits, without any further tumor recurrence.

Discussion: A thorough revision of the literature is provided including previous reported cases of spontaneous bleeding CPPs and other underlying causes that could lead to multifocal hemorrhages.

Conclusion: Due to the rarity of these events, this case remains still open to speculative hypotheses drawn to explain the neuroanatomical and pathogenetic basis behind this case report.

1. Introduction

Choroid plexus papillomas (CPP) represent rare, vascular, benign tumors accounting for 0.4–0.6% of all intracranial tumors. The malign variants consisting in atypical choroid plexus papilloma (WHO grade II) and choroid plexus carcinoma (WHO grade III) are much less common (Table 1). Generally CPP, which represent more than 85% of intraventricular tumors, arise from the lateral ventricles in pediatric age and from the fourth ventricle in the adulthood; unusual locations like the cerebellopontine angle, the pituitary fossa or even paraventricular ones are also reported in the literature. Due to their highly vascularized structure, hemorrhagic findings representing a neurosurgical emergency despite rare are possible.

We report a case of a patient operated for a paratrigonal hemorrhagic CPP WHO class I presenting with multiple satellite supra- and infratentorial hemorrhages.

2. Presentation of case

A 55-year-old white male presented to our institution with a sudden neurological impairment consisting in headache, right hemiparesis, speech impairment and subsequent consciousness deterioration. A CT-scan revealed multiple hyperdense supra- and infratentorial bilateral lesions located in the left paratrigonal, right occipital and vermis regions (Fig. 1a and b). Due to the complex radiological presentation, the patient promptly underwent further investigation with contrast-enhanced MRI study, which better defined the huge solid hemorrhagic left paratrigonal lesion widely extended in the temporal lobe, and characterized by patchy signal and intense enhancement after Gadolinium administration. (Fig. 2a and b) Interestingly, the other supra- and infratentorial bilateral hemorrhages did not show any pathological enhancement after contrast administration.

The patient underwent urgent intervention for excision of the paratrigonal lesion by left temporal craniotomy and intrasylvian approach. The focal lesion was detected few centimeters from the temporal cortex, within a hemorrhagic cystic wall with a solid paraventricular component. The macroscopic analysis revealed a translucent and apparently vascular tissue that was completely evacuated as demonstrated by the post-surgical CT scan.
Table 1
WHO classification of choroid plexus papilloma.

| Classification                  | ICD-O   | WHO grade |
|---------------------------------|---------|-----------|
| 1.5 Choroid plexus tumors       | 9390/0  | I         |
| 1.5.1 Choroid plexus papilloma  | 9390/1  | II        |
| 1.5.3 Choroid plexus carcinoma  | 9390/3  | III       |

(Fig. 3a). Histological specimens showed a mass lesion formed by vascular convolutions and plexiform shapes with rounded calcification. Nuclear atypias were not demonstrated, and immunohistochemical analysis showed positivity for EMA and S-100 protein leading to the diagnosis of WHO class I CPP.

A few days later due to failure of conservative treatment of the satellite hemorrhages the patient underwent second-time surgery for their evacuation. The intervention was performed with a right-side suboccipital approach, than after transverse sinus identification and ligature a lower craniectomy was extended to complete the cerebellar hemisphere exposition and infratentorial hemorrhage evacuation. Post-surgical CT scans successfully confirmed the evacuation of both cerebellar and occipital hemorrhages (Fig. 3b and c). Interestingly the histological examination of the tissue probe did not reveal any neoplastic features confirming their sole hemorrhagic nature. Patient’s conditions slowly improved despite severe neurological deficits, such as right hemiparesis (3/5 according to the Muscle strength grading scale of Royal Medical Research Council of the Great Britain), Broca’s aphasia, supero-external quadrantopisia and mild dysmetria still presents at a 2-year follow up. To date, no further tumor recurrence was found on contrast-enhanced serial MRI.

3. Discussion

CPP are rare, primary, slow-growing brain tumors arising from the neuroepithelium of the choroid plexus. They commonly arise...
from the ventricular regions where choroid plexus is normally located. In children they are prevalently found in the lateral ventricles whereas in the adulthood they are mostly described in the forth ventricle.1 Atypical localizations have been described in literature: cerebellolopontine angle localizations account for 9% of all CPPs,8: direct extension of tumor through the foramen of Luschka, from the fourth ventricle and cerebrospinal fluid dissemination could explain this neuroanatomical distribution. Other rare extraventricular localizations have also been described in the pituitary fossa,2,5 suprasellar region,4 third ventricle6 and pineal region.7

Due to the highly vascular structure of CPPs, bleeding is a common complication of their surgical excision9; nevertheless hemorrhages have also been described in post-traumatic brain injury patients,10 or even spontaneously.2,11

The first report of hemorrhagic CPP was described in 1955 by Ernsting12: the Author reported a case of a spontaneous subarachnoid hemorrhage caused by a CPP located in the temporal horn of left lateral ventricle. Previously findings of hemorrhagic intraventricular CPPs were only demonstrated in necropsy (Graves and Fless in 1934 and Friedman and Solomon in 1936), but never described in living patients.

Most of the cases of bleeding CPPs reported in the literature describe SAH as the main hemorrhagic event, and in all of them surgery was firstly performed to decompress the surrounding brain parenchima by evacuating the hemorrhagic lesion. Interestingly, Matsushima et al.11 reported also a primary intraventricular hemorrhage due to a papilloma and a venous angioma of the choroid plexus. Furthermore, most of the histologically confirmed CPPs described in ectopic areas seem to be more susceptible to bleedings, suggesting a stronger tendency in bleeding in such locations.

Generally speaking, no radiological investigation is specific to raise the suspicion of ectopic CPP: CT scans show a heterogeneous density in 75% of cases and hypodensity in 25%, calcifications are seen in a variable rate ranging between 25% and 10%.13 MRI scans may reveal isointense lesions on T1-weighted image and heterogeneous hyperintense lesions on T2-weighted image with marked enhancement following administration of Gd-DTPA.14 Regarding blood supply there are not data demonstrating pathological vascularization except for some slight segmental dilatations of the posterior circulation1,14; for instance on vertebral angiography a tumor blush is a common finding.8 To this regard, Furuya et al.3 reported a well-developed choroid branches of PICA and AICA in his cases, whereas in a similar case Zhang8 found only a blood supply from AICA. Due to the severe clinical presentation and evidence of multifocal bleedings in the case herein reported preoperative angiographic data are missing, nevertheless postoperative cerebral angiography did not show any pathological finding.

Immu-no-histochemistry shows no specific markers, but positivity for epithelial membrane antigen (EMA), S-100 protein, transthyretin and in lower rate Glial Fibrillary Acidic Protein, suggesting the epithelial nature of the nature. In our case tumor specimens from the paratrigonal CPP resulted positive for EMA and S-100 protein; interestingly, histological examination of the other supra- and infratentorial hemorrhages did not show any neoplastic features.

To the best of our knowledge this is the first report of a patient with ectopic hemorrhagic CPP presenting with satellite multiple hemorrhages; therefore their occurrence in different cerebral areas is still open to speculative hypothesis.

In our patient the raise in intra-cerebral pressure (ICP), the occurrence of seizures and focal deficit were the first and foremost clinical findings. To this regard it is well known that any pathology determining an increase in ICP could facilitate vessel disruption in different cerebral areas, even far away from the primary lesion.15 A few studies reporting subarachnoid hemorrhage associated with choroid plexus papilloma suggest this hypothesis, and raise the suspect of absorptive hydrocephalus as the primary cause of multiple bleedings. Nevertheless, in our case the MRI study while showing ventricle compression and blood flooding into the perimesencephalic cistern, did not highlight any ventricular enlargement.

4. Conclusion

Due to the rarity of these events, this case remains open to all possible speculative hypothesis. On one hand, we may hypothesize that underlying clinical conditions, leading to a sudden rise in blood pressure and ICP, could have been responsible for the satellite supra- and infratentorial hemorrhages. On the other we should suspect that even the parietal and cerebellar hemorrhages could be the expression of ectopic bleeding papillomas, disregated and made undetectable to the histological examination by the acute event. In fact this mechanism, that is reported in literature,16 seems to be responsible for an atypical intraparenchimal hemorrhage, even though the postoperative histological examination fails to confirm a pathological substrate, raising a strong suspicion of vascular malformations.

Conflicts of interests statement

All authors do not have any conflicts of interest.
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Ethical approval

The patient cited in this article was fully informed and gave his written consent.

Author contributions

Clinical study was done by Maimone Giuseppe. The study was designed by Maimone Giuseppe and Ganau Mario. Clinical data were collected by Maimone Giuseppe and Paterniti Sebastiano. Maimone Giuseppe, Ganau Mario and Paterniti Sebastiano performed the data analysis. Manuscript was written, revised and finally approved by all the authors.

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