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An unexpected sudden death due to a choroid plexus papilloma: an autopsy case report

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Abstract

Background: Choroid plexus papillomas (CPPs) are histopathologically benign and rare central nervous system tumors. These tumors remain more frequent in children than adults. It is infrequent for these tumors to cause a sudden unexpected death. We aim in this case to discuss the unusual and fatal presentation of choroid plexus papilloma and the mechanism of death.

Case presentation: we report the case of a 61-year-old man with no medical history, diagnosed at autopsy with a previously unknown CPP. Initial complaints were chronic headache occurring in the last month and acute chest pain for two days. The forensic autopsy including the histopathologic examination showed a tumoral mass of the choroid plexus in the fourth ventricle diagnosed as a psammomatous CPP. The cause of death in this case was attributed to a massive cerebral edema caused by the tumoral mass.

Conclusions: Through this case report, we stress the importance of an early and a vigorous investigation of every headache and an early detection of this tumor and we highlight as well the role of the post mortem examination to detect such a fatal complication.

Keywords: Choroid plexus, Papilloma, Sudden death, Autopsy, Case report

Background

Choroid plexus papillomas (CPPs) are benign neoplasms representing less than 1% of intracranial tumors (Sodhi et al. 2013). The annual incidence is reported to be approximately 0.3 per 1.000.000 population inhabitants (Kayaselçuk et al. 2002). CPPs in adults occur mostly in the fourth ventricle; however, lateral ventricles are a common location in children (Singh et al. 1998). Typically, signs of intracranial hypertension are the first findings in these cases.

Herein, we report the case of a 61-year-old man with a history of chronic headaches who died suddenly. The unexpected diagnosis of psammomatous CPP, in this case, caused a sudden death and was made during the post mortem examination.

Case presentation

A 61-year-old man with no medical history had been suffering from an intermittent headache for 1 month and chest pain for 2 days before coming to the hospital. On the admission, his neurological state deteriorated and he died before undergoing any explorations. Due to the absence of an evident death cause, a medico-legal autopsy was requested by the prosecutor's office.

The height of the corpse was 175 cm. The external examination showed no evidence of traumatic injuries. In the autopsy, there was cervicofacial cyanosis. The heart weighed 446 g. There were multiple plaques of

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atheromatous in the three coronary arteries and more specifically in the anterior interventricular artery where a stenosis of 20% of the arterial lumen was noticed. The myocardial sections showed a transmural scar in the posterior wall of the septum measuring 7.5 cm^2 and starting from the apex to the atrio-ventricular level. There was no identified macroscopic focal hemorrhage in the scar. Right and left lungs weighed 588 g and 510 g, respectively, and were congestive and edematous. The examination of abdominal organs did not show any specific lesion.

The brain weighed 1550 g and was very edematous. Dilation of the third and the fourth cerebral ventricles was noticed. A mass of the choroid plexus was found in the fourth ventricle measuring 0.8 × 1 cm (Fig. 1). This mass was taken for histopathologic analysis as well as samples from all the organs. A blood sample was taken for toxicological analysis.

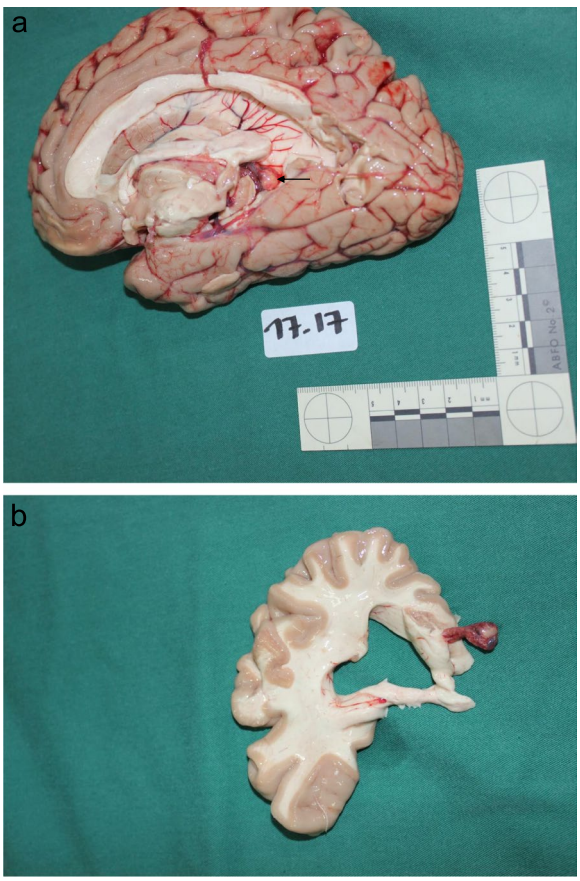

Fig. 1 Photographs illustrating fourth ventricle mass attached to the choroid plexus. **a** A front view of the cerebral hemisphere with the choroid plexus mass (arrow). **b** Photograph illustrating a closer look at the tumoral mass attached of the choroid plexus attached to the fourth ventricle

At the histopathologic examination, the choroid plexus mass was formed by papillae covered by simple cuboidal epithelium cells within connective vascular tissue (Fig. 2). Many psammoma bodies were identified in the tumor with few areas of osseous metaplasia (Fig. 3). The histological examination of the rest of the brain tissue showed foci of cerebral edema and did not show any signs of malignancy. Microscopic examination of lung samples was normal. There was a myocardial scar on the septum and no signs of acute myocardial infarction. Pathologic examination of the other samples didn't show abnormalities. Toxicological analyses were negative. The cause of death was attributed to cerebral edema due to choroid plexus papilloma.

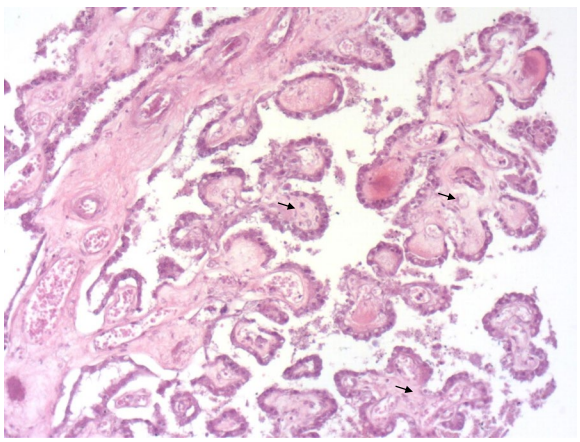

Fig. 2 Microscopic finding showing many papillar structures (arrows) lined by regular cuboidal cells (HE ×200)

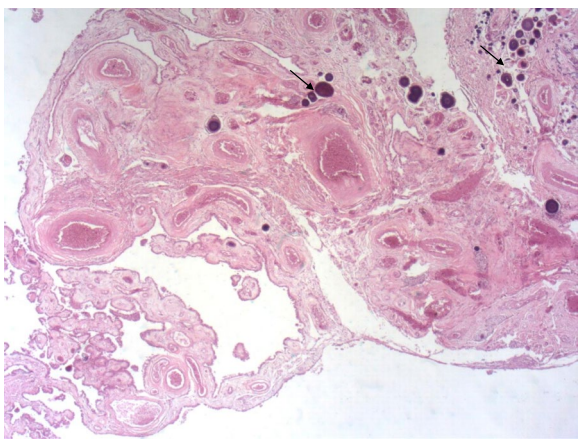

Fig. 3 Psammomatous calcifications (arrows) surrounding choroid plexus papilloma (HE ×100)

Discussion

Choroid plexus papilloma is a rare high vascular benign neoplasm arising from the epithelium of the choroid plexus (Karim et al. 2006). This tumor accounts for 0.4 to 1% of all intracranial tumors in adults (Kayaselçuk et al. 2002; Horská et al. 2001). CPPs in children make up 2% to 4% of brain tumors and 5 to 15% occurring among patients aged less than 1 year old (Raimondi and Gutierrez 1975; Mishra et al. 2014). CPPs are mostly found in the fourth ventricle in adults whereas lateral ventricles are the most common localization for children (Singh et al. 2006; Horská et al. 2001; Raimondi and Gutierrez 1975). CPPs appear as cauliflower-like masses attached to the wall of the ventricle (Pencalet et al. 1998). Microscopically, CPPs show a layer of cuboidal epithelial cells arranged in a papillary configuration with finger-like projections (Lee et al. 2009). Signs of malignancy in CPPs include nuclear pleomorphism, raised cellular density, cell necrosis, brisk mitoses, and invasion of surrounding brain tissue (Jeibmann et al. 2006). Recently, an additional entity with intermediate characteristics "atypical choroid plexus papilloma" was introduced. This entity was distinguished from the CPP by raised mitotic activity and increased nuclear-cytoplasmic ratio (Lee et al. 2009). Differential diagnosis of CPP should be made also from bilateral villous hypertrophy. This entity has often been erroneously referred to as CPP. In bilateral villous hypertrophy, diffuse enlargement of the choroid plexus in the lateral ventricles is associated with hypersecretory function all with normal histologic structure (Erman et al. 2003)

The evolution of symptoms in CPPs differs among adults and children. Headaches and visual symptoms are usually the leading symptoms in children while in adults, a more rapid clinical presentation with a neurological deficit and seizures is noticed in most of the cases (Karim et al. 2006).

Herein, we present a case of a 61-year-old man with complaints of intermittent chronic headaches and acute nonspecific chest pain. The patient had lost his consciousness suddenly and arrived dead at the hospital; thus, he did not undergo any imaging techniques. CT scan and especially MRI are the first elements of CPP diagnosis (Hirano et al. 1994). In our case, the choroid plexus mass was first discovered in autopsy and the CPP diagnosis was made during the histopathologic examination. The chest pain, as well as the myocardial fibrosis scar found in the autopsy, raised the speculation about possible ischemic heart disease as the origin of the death. The finding of the choroid plexus mass was unexpected due to the fact that there were no neurological initial complaints and only intermittent chronic headaches were reported by the family one month before death. Nevertheless, we cannot formally eliminate the occurrence of some symptoms not noticed and/or not reported by the family. The episodes of intermittent headaches may have been related to the transient cerebral herniation.

Massive cerebral edema was the cause of death. The fatality of this tumor is explained by the fourth ventricular obstruction in general and particularly in this case. The obstruction of the cerebrospinal fluid outflow may have been the reason for the intermittent chronic headache reported. Ventricular enlargement noticed in the autopsy is seen in most CPPs, which may be due to a combination of factors including over-production of cerebrospinal fluid (CSF) and obstruction of CSF pathways by the mass (Horská et al. 2001; Levy et al. 2001). Fatalities in choroid plexus tumors are seen mostly in malignant tumors, i.e choroid plexus carcinomas. Choroid plexus papillomas are rare and responsible for most of the deaths seen in choroid plexus tumors (Wyatt et al. 2001). Intracranial hypertension and metastases are believed to be the most incriminated mechanisms of deaths in such cases (Wyatt et al. 2001). Central nervous system benign tumors other than CPP can cause fatality (Barranco et al. 2018). Despite the rarity of these cases, Milroy and Smith (1996) and Na et al. (2014) reported several cases where pineal gland cysts, which are benign tumors, caused compression and sudden unexpected deaths. Although in these reports a contributing factor such as an intense effort was responsible for an increase in intracranial pressure and cerebral perfusion and thus death, in our case, we do not know exactly if the deceased had an intense physical effort or any other contributing factor before his death.

The finding of psammomatous elements in CPPs is rare according to previous reports. Case reports of brain tumors that share the psammomatous element were mainly about psammomatous meningiomas (Perry et al. 1997; Kleihues et al. 2002), melanocytic schwannomas (Grayson and Hale 1998), and papillary meningioma (Kleihues et al. 2002). Psamomma bodies (PBs) are changes whose mechanism of formation is still unclear (Hamir et al. 2001). PBs are more frequent in choroid plexus during aging in healthy older people (Hamir et al. 2001). According to previous reports, there was no association between PBs and malignancy in CPPs (Tena-Suck et al. 2006). To our knowledge, the association between PBs with osseous metaplasia and choroid plexus tumors has not been previously well-identified yet (Tena-Suck et al. 2006).

Conclusions

Our case revealed that even a benign CPP can cause an unexpected sudden death when non-diagnosed, thus, only a meticulous post mortem examination and

thorough gross examination, comprehensive for brain specimens are necessary to rule out neurological causes of sudden deaths. A Chronic headache in an adult should always raise the attention of an underlying cerebral disease which, in this case, had a fatal outcome. This case is considered one of the very few cases of fatal psammomatous CPP diagnosed in post mortem examination reported in the literature.

Abbreviations
CPPs: Choroid plexus papillomas; MRI: Magnetic resonance imaging; CSF: Cerebro-spinal fluid; CT: Computerized tomography; PB: Psamomma bodies.

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Not applicable

Authors' contributions
MAZ and SG assisted the autopsy and wrote the first draft of the manuscript. MBD performed the autopsy and commented on the first version of the manuscript. SM and DC performed the histopathological examination. NA contributed on the revision of the first version of the manuscript and to the establishment of the histopathological diagnosis. MM commented on the first version of the manuscript from the point of the view of a pathologist. All authors have read and approved the manuscript.

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Availability of data and materials
Not applicable.

Declarations

Ethics approval and consent to participate
No ethical approval is required in this case.

Consent for publication
An oral consent was taken from the deceased's descendant after informing him of the purpose of the case report (there is no identification of the deceased in the mentioned case and we did not need to take a written consent in this case).

Competing interests
The authors declare that they have no competing interests.

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