



An Atypical Choroid Plexus Papilloma of IV Ventricle with Dissemination to a Sacral Tarlov Cyst: A Case Report

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Abstract

Choroid Plexus Papillomas (CPP) is uncommon tumors of the CNS. Atypical CPPs represent an intermediate-grade subtype (Grade II) introduced in the update 2007 of the WHO classification. Distal spread of atypical CPPs through the subarachnoid space is very rare. We present a 53 year-old female with history of macroscopic surgical excision of an atypical CPP in the IV ventricle, with multiple local relapses and who ten years after diagnoses, presented tumor metastasis in a Tarlov cyst in the sacrum.

Atypical CPPs are uncommon tumors, and to the best of our knowledge, no cases linking atypical CPP tumor dissemination to a Tarlov cyst have been reported in English or Spanish language medical literature.

Keywords: Atypical choroid plexus papilloma; Tarlov cysts; Metastasis; Neurography MRI

Abbreviations

TCs: Tarlov Cysts; CPP: Choroid Plexus Papilloma; NMRI: Neurography Resonance Magnetic Imaging

Introduction

Choroid plexus tumors are rare, making up only about 0.2% of all CNS tumors, and most commonly arise in young children in the lateral ventricles. A very uncommon tumor in adults, the most common site is the IV ventricle, less often in the third ventricle or the cerebellopontine angle [1]. Based on histology, choroid plexus tumors have been sub-classified into Choroid Plexus Papilloma (CPP, World Health Organization [WHO] grade I), atypical Choroid Plexus Papilloma (aCPP, WHO grade II), and Choroid Plexus Carcinoma (CPC, WHO grade III), with aCPP showing increased mitotic activity and CPC displaying common histological hallmarks of malignancy [2]. CPPs are typically treated by surgical resection alone, with excellent outcomes. In contrast, treatment outcomes for CPC remain poor, especially when gross total resection is not feasible [3]. While adjuvant chemotherapy and radiotherapy appear to improve survival in CPC, and use of radiotherapy is limited, especially in infants and very young children because of treatment-related toxicities. Whether CPPs behave, (and therefore should be treated) in similar fashion to CPPs or CPCs remains controversial, with some data suggesting response may be age-dependent [4]. Subarachnoid spread of atypical CPPs is uncommon [5].

We report a case of atypical choroid plexus papilloma of the fourth ventricle in an adult patient, in whom, ten years after surgical resection; spinal tumor metastasis to a Tarlov cyst of the sacrum was diagnosed.

Case Presentation

Fifty-three year-old female, who consulted at our institution in 2005 because of recent onset headache associated with vertigo. Physical examination revealed right gaze torsional nystagmus, left appendicular ataxia, tandem gait with instability and positive Romberg test. MR showed the presence of a tumor lesion in the IV ventricle of intermediate signal strength on T2-weighted images,

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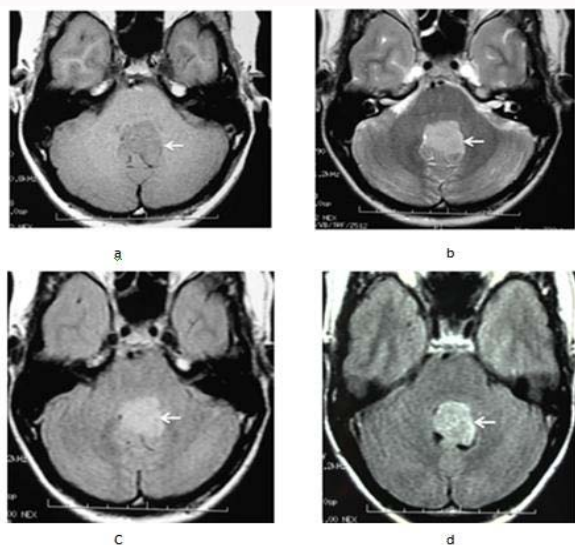
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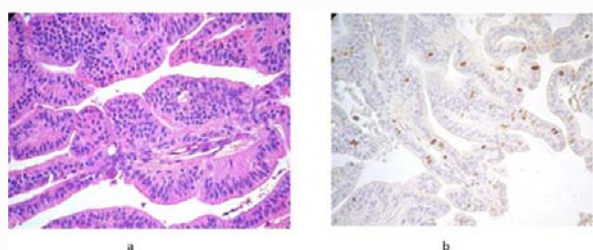
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Figures 1: (a) Brain MR imaging showed tumor lesion in the IV ventricle of intermediate signal strength on T1, (b) T2-weighted images, (c) and FLAIR, (d) with intense enhancement after gadolinium injection.



Figures 2: (a) Haematoxylin and eosin stain $\times 40$: APP composed of a single layer of tumor cells with mild pleomorphism and increase in cellularity, (b) Ki67 showing increase of proliferation index.

with intense enhancement after gadolinium injection (Figure 1).

In December of 2005, the patient was subjected to surgical resection in the Concorde position through a medullary (telovelotonsilar) approach. Complete resection was possible and no post-operative sequelae were observed. Pathology report indicated presence of choroid plexus papilloma, with 2% Ki 67 expression (Figure 2).

Post-op control MRI of the brain and spine showed no residual macroscopic tumor.

In the year 2009 however, routine follow up MR imaging showed tumor recurrence in the left lateral recess of the IV ventricle, of similar morphology to the original tumor (Figure 3). This was resected at another institution and the pathology report confirmed atypical choroid plexus papilloma recurrence with 8% Ki-67 proliferation fraction.

In 2010, a new local recurrence was diagnosed and treated with Gamma Knife radiosurgery; the procedure was repeated in 2013, due to a new local recurrence.

In March 2015, MRI showed increased size of the IV ventricle nodular lesion, as well as presence of other lesions of similar behavior along the walls of the IV ventricle, suggesting meningeal spread. MRI of the spine showed multiple perineural Tarlov cysts, in the lumbar and sacral regions, with the largest on the left side at S1 level. The cyst had induced bone remodeling, and showed lateral wall thickening

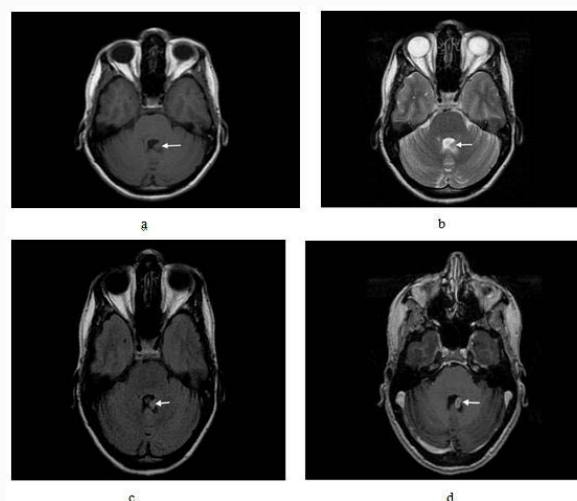


Figure 3: (a) Brain MR imaging showed tumor recurrence in the left lateral recess of the IV ventricle, of similar morphology to the original tumor with intermediate signal strength on T1, (b) T2-weighted images (c) and FLAIR (d) with intense enhancement after gadolinium injection (arrows).

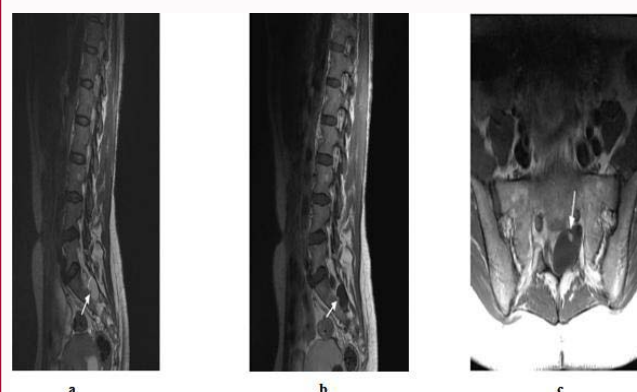


Figure 4: (a) MR images of lumbosacral spine showed a sacral 1 Tarlov cyst with small nodular lesion with intermediate signal in T2- weighted images, (b) and intense enhancement after gadolinium injection (sagittal plane in b) (c) coronal plane in c (arrows).

with contrast enhancement, which was interpreted as presence of spinal dissemination (Figure 4). Patient received chemotherapy with Vincristine (VCR), Cyclophosphamide (CMF) Andetoposide (VP 16) for four cycles.

In August 2015, the patient referred pain in the left hamstring. MRI showed progression of the Tarlov cyst lesion, which had evolved to an oblong-shaped mass extending through the sacral foramen, hypointense on T2-weighted imaging, with enhancement post-gadolinium (Figure 5).

Patient did not respond to pain medication and was re-operated in September 2015.

The cyst was accessed through a posterior approach, and a grey/pinkish lesion of intermediate consistency removed. The remaining cavity was plugged with Absorbable Haemostatic Gelatin Sponge (Spongostán™), a Dural graft implant (Duraform®) and fibrin glue (Beriplast®).

Histology of the lesion showed a proliferative neoplasm consisting of cuboidal epithelial cells, with mild pleomorphism and

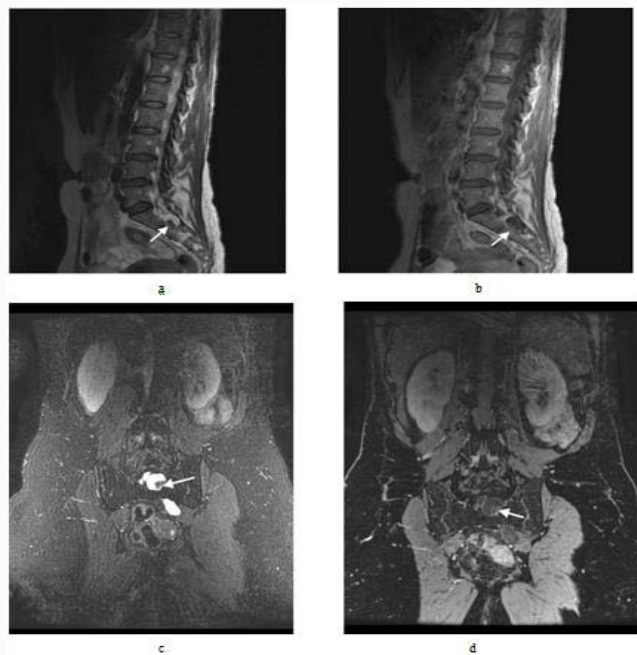


Figure 5: (a,b) MR images of lumbosacral spine, (c,d) MR Neurography of lumbosacral plexus (a and c) showed progression of the Tarlov cyst lesion, which had evolved to an oblong-shaped mass extending through the sacral foramen, with hypointense signal on T2-weighted imaging, and (b,d) enhancement after gadolinium injection (arrows).

acid-staining cytoplasm, lining papillary structures. Amorphous acidophilic material and positive focal GFAP immunohistochemistry, as well as 6% Ki67 confirmed diagnosis of metastatic atypical choroid plexus papilloma (Figure 6).

The patient was discharged on second day after surgery with no neurological sequelae. In November 2015, the patient received craniospinal irradiation (3000 cGy) with a boost of 600 cGy in the upper spine and 1500 cGy in the lower spine.

At most recent follow up visit in November 2016, patient remains asymptomatic with no signs of radiological progression.

Discussion

Choroid Plexus Papillomas (CPP) is rare neoplasms originating in the choroid plexus epithelium, and constitute approximately to 0.2% of all CNS tumors [6]. WHO classification of these tumors will depend on the number of mitoses, degree of nuclear pleomorphism, cell density and presence or absence of necrosis.

Typical CPPs are considered Grade I tumor according to the WHO classification. Atypical or anaplastic choroid plexus tumors showing more pleomorphism, cellularity and mitoses, correspond to grade II, and represent the transition from a low grade to a high grade tumor. Choroid plexus carcinomas are classified as Grade III according to the WHO, and present evident signs of malignancy. The atypical subtype was incorporated in the 2007 version of WHO classification [2].

The patient we present showed metastatic leptomeningeal dissemination in a Tarlov cyst. Distal metastases may occur as a result of the spread of tumor cells through CSF. These have been subdivided into three subgroups: Leptomeningeal, Intraparenchymal and Intraventricular [7].

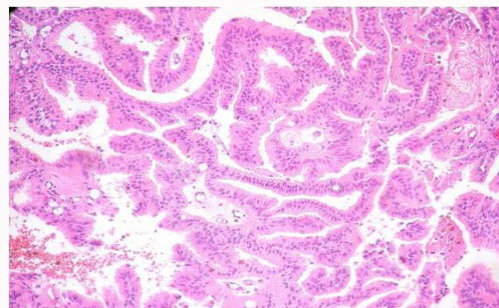


Figure 6: Papillary structure in the Tarlov cyst.

Distal spread to the spine is rare [8]. Distal metastases of CPPs are more common in tumors located in the posterior fossa, [9] and have been associated most often with choroid plexus carcinoma. However, the multicenter CPT-SIOP-2000 study reports 21% of carcinomas and 19% of atypical CPPs present distal spread [10].

After searching the English and Spanish language medical literature, we were unable to find any reports on atypical CPP spread to a Tarlov cyst.

Tarlov or perineural cysts develop as a result of dilated sheaths between endo and perineural spaces of nerve roots [11]. Incidence in the general population is between 4.6% and 9%, with equal gender distribution and higher prevalence among young adults [12]. Peripheral nerve fibers and ganglion cells have been found in the wall of these cysts, which on occasion connect with the subarachnoid space through microscopic channels [13].

In the case presented, leptomeningeal spread in the Tarlov cyst occurred 10 years after primary tumor resection. Time elapsed between initial diagnosis and detection of tumor spread is variable [14]. This may be present at time of diagnosis, develop shortly after surgery (as a result of intraoperative seeding) or less often, after a few years [15,16].

Atypical CPPs, of intermediate malignancy (grade II) in the WHO classification, present 2 or more mitoses in 10 high power fields selected at random. This increased mitotic activity is the basis for the differing prognosis and treatment with respect to the typical form [17].

First line therapy is always surgical excision, both for all variants of CPP [18]. Prognosis in typical CPP cases is excellent, with survival rates reported to be 100%, 5 years after resection. No adjuvant treatment is indicated in these patients [16,19]. Conversely, prognosis for patients with choroid plexus carcinoma is less favorable, with 5-year survival rates ranging between 26% and 50%. Residual tumor tissue after resection remains the most ominous negative prognostic factor. Indication of adjuvant treatment is still under debate, and more results are needed to confirm its advantage. Radiotherapy and chemotherapy may increase survival, but this has yet to be confirmed [20].

The first clinical trial specific for CPPs was begun in 2000 under the guidance of the International Society of Pediatric Oncology (SIOP). The CPT-SIOP-2000 study protocol 10 recommends, both for CPP and CPP patients in whom surgical resection was complete, watch and wait follow up with brain and total spine MR imaging, with contrast (gadolinium). For patients with carcinoma, metastatic disease, or CPP in whom resection was incomplete, the

guideline recommends complementary treatment with 6 cycles of chemotherapy and cranoespal radiotherapy.

Conclusion

We present a case of a 53 year-old female with a history of CPP of the IV ventricle, who developed tumor metastasis in a sacral Tarlov cyst 10 years after surgical excision of the primary tumor and multiple local relapses.

Atypical CPP tumors are rare, and the possibility of leptomeningeal spread, though infrequent, should be considered. We underscore the importance of use of contrast MR imaging during patient follow up, in order to detect small implants such as the one here described, in a Tarlov cyst of the sacrum.

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