SVOA Neurology

ISSN: 2753-9180

ScienceVolks Open Access

Case Report

Pilocytic Astrocytoma Mimicking Neurocysticercosis: Atypical Presentation

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***Received:** December 30, 2021 **Published:** January 11, 2022*

Abstract

Pilocytic astrocytomas (PAs) are neoplasms frequently diagnosed during childhood and adolescence, and the most common location is infratentorial, as a solitary lesion. Here we present an unusual case of multifocal cystic PA in a 20 year-old female patient. The patient was referred to our hospital with a presumptive diagnosis of neurocysticercosis. After an exhaustive study of the magnetic resonance imaging (MRI) that revealed multiple cystic lesions involving the cerebellum, brain stem and upper spinal cord, spectroscopic analysis and the negative results of the cerebrospinal fluid (LCR) analysis, we decided to perform a stereotactic biopsy and finally arrived to the diagnosis of PA.

***Keywords:** pilocytic astrocytoma - neurocysticercosis - multifocality- atypical clinical presentation.*

Introduction

Among primary Central Nervous System (CNS) tumors, PAs are frequently observed during childhood and adolescence [1]. In an epidemiological study in the US, PAs represented 1.3% of all CNS tumors, the median age of presentation was 12 years old, with a slightly male to female preponderance and an incidence of 0,8/100.000; 75% of PAs were diagnosed before the age of 20 [2, 3].

The most frequent location is infratentorial, with the cerebellum being the most prevalent location, as well as, along midline structures of the neuroaxis, such as the third ventricle, optic chiasm and optic nerves, basal ganglia and spinal cord [2]. These are low-cellularity, slow-growing lesions that are considered World Health Organization (WHO) grade 1 tumors. Features like multifocality, malignant transformation and appearance during adulthood are rare [1, 4]. Metastasis and malignant transformation are unusual, and the recognition of its radiological, pathological and molecular features are of paramount importance for proper diagnosis, treatment and prognosis [5]. Possible differential diagnosis to bear in mind when studying these lesions include metastatic disease, other gliomas, parasitic infections and vascular lesions. Regarding differentiation with parasitic disease, here we present a case of PA previously diagnosed as neurocysticercosis.

Case Presentation

A 20 year-old, right-handed female patient was referred to our hospital because of a fever, altered mental status and presumptive ventriculoperitoneal shunt (VPS) dysfunction, whereby she was admitted to our Neurosurgical department. She had a history of hypothyroidism, type II diabetes mellitus, depression, hydrocephalus treated with a VPS and a 6-year history of presumptive neurocysticercosis treated with albendazole; no evidence of serological studies were found. Two years before presentation, she was treated with a VPS for non-communicating hydrocephalus produced by one of the presumptive neurocysticercosis lesions located in the aqueduct, which was reviewed a year after deployment, with no complications.

Positive findings on physical examination included bilateral mild mydriasis, bilateral Collier's sign, divergent strabismus manifested by upper vertical gaze, vertical gaze palsy and convergence-retraction nystagmus. Eye fundus showed chronic bilateral papilledema.

During hospitalization she presented with an acute decrease in level of consciousness, computed tomography of the brain was performed, showing signs of acute hydrocephalus and multiple infratentorial cystic lesions, thus radiological suspicion of VPS dysfunction was made (Fig 1).

A new revision of the VPS was planned and performed. Hydrocephalus was successfully resolved. A cerebrospinal fluid (CSF) analysis was made, including cysticercosis serology. Results yielded no infectious agents at that time. Due to the unusual location of the cystic lesions, a brain magnetic resonance imaging (MRI) was ordered.

Cerebral Magnetic Resonance Imaging (MRI) showed multiple cystic lesions involving the cerebellum, brain stem and upper spinal cord, the major one located in the cerebellar culmen (Fig. 2). These lesions were hypointense on T1-weighted images. T2-weighted images presented hyperintensity on the inside. Diffusion-weighted images showed restriction to diffusion. Finally, the lesions presented mild enhancement in the gadolinium series (Fig. 3). Spectroscopic analysis showed increased choline and lactate, and decreased N-acetyl aspartate (Fig. 4). The findings were consistent with neoproliferative disease.

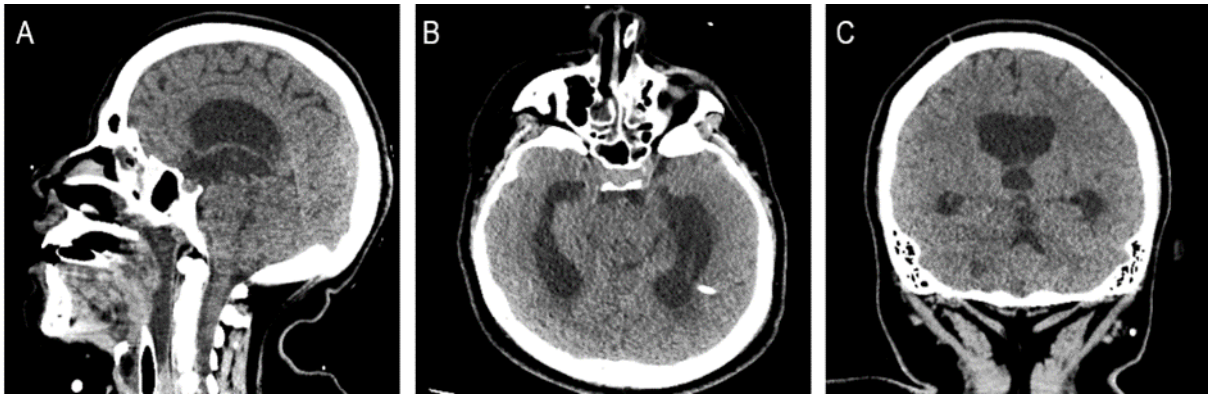

Figure 1. *Brain Computed Tomography showing signs of acute hydrocephalus. Note in A and C, there are hypodense infratentorial lesions suggesting vesicular lesions.*

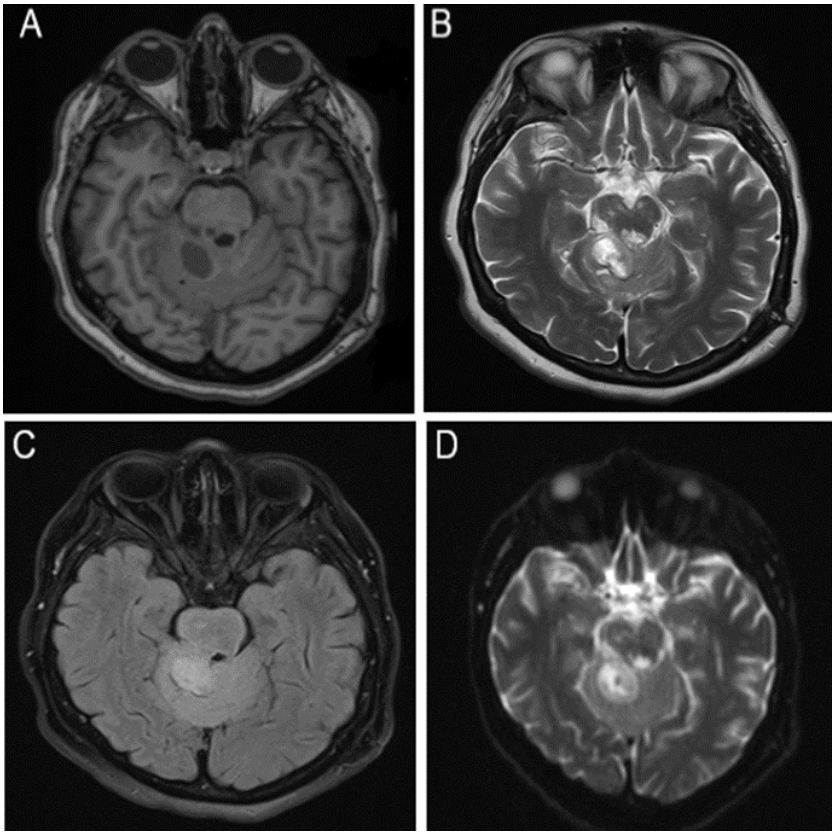

Figure 2. *MRI axial sequences showing the most prominent cystic lesion located at the culmen. A. T1-weighted sequence showing a hypointense cystic lesion. B. T2-weighted imaging sequence showing hypointense cystic lesion with hyperintense content. C. FLAIR sequence showing isointense-walled cystic lesion with hyperintense content. D. Diffusion-weighted image showing restriction to diffusion. Note the presence of multiple lesions with similar characteristics.*

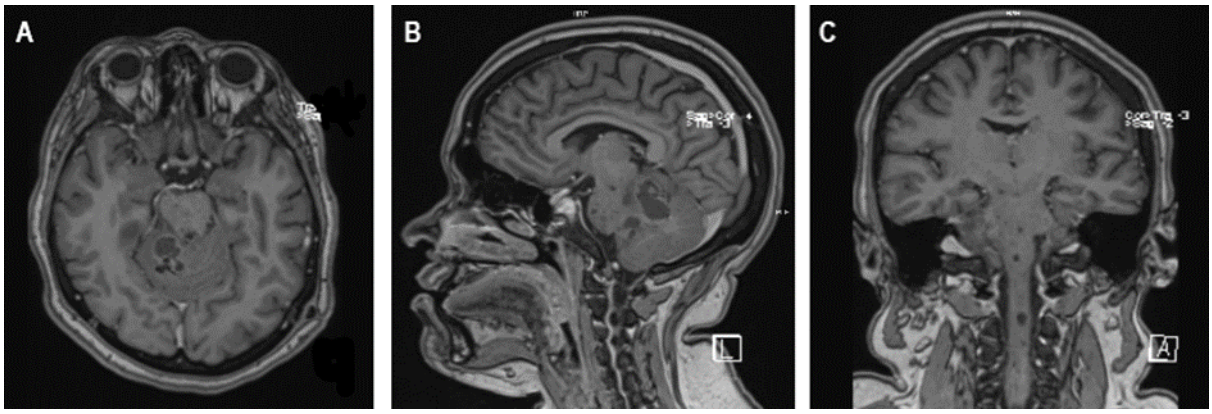

Figure 3. *Contrast enhanced T1-weighted images, showing enhancement of cerebellar lesion (A, B) and presence of multiple lesions involving the brainstem and cervical spinal cord (C).*

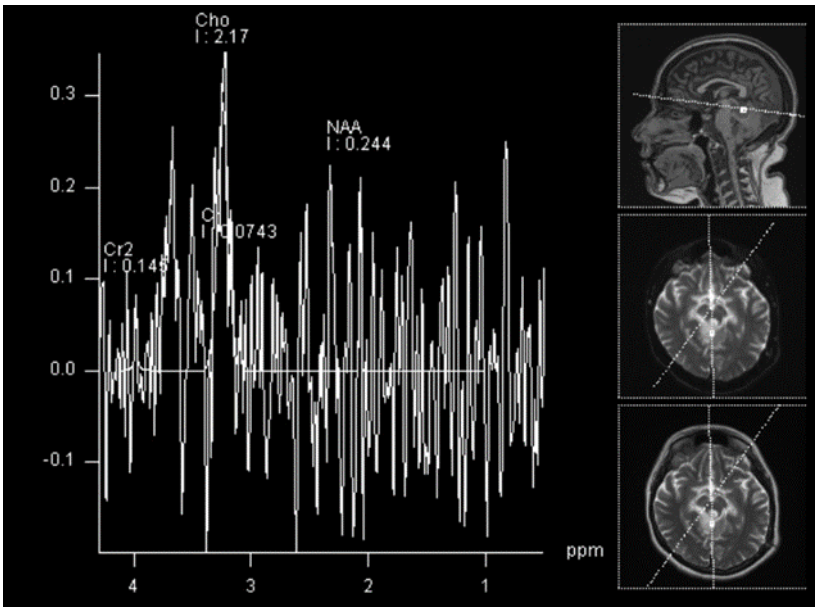

Figure 4. *Spectroscopic analysis of cerebellar lesion.*

Imaging findings, disease progression and unresponsiveness to antiparasitic treatment opened the venue for differential confirmation, and thus, a stereotactic biopsy of the greater cerebellar lesion was performed.

Biopsy Findings

Microscopy: sample of cerebellar cortex that depict a focal neoplastic proliferation of astroglia, moderate cellularity. The cells are elongated, irregularly circular, small to mid-sized with central ovoid and hyperchromatic nuclei, with scant acidophilic cytoplasm that contain fine prolongations. There are scattered Rosenthal fibers. No mitosis or necrotic changes are seen. There is moderate vascularization.

Immunohistochemistry: GFAP positive in neoplastic cells; SYN negative in neoplastic cells; Ki 67 positive in 5% of neoplastic cells.

Conclusions: Morphological findings described and immunomarker profile are compatible with a pilocytic astrocytoma.

After confirmation of Pilocytic Astrocytoma diagnosis, the patient was referred to the Oncology Department of our Hospital.

Discussion

Neoplastic lesions could present in various ways and could even emulate non-neoplastic conditions. PAs are tumors that usually present in childhood as solitary, cystic lesions with a brightly enhancing mural nodule located in the cerebellum or optic chiasm [6, 7]. In comparison with other types of gliomas, they rarely manifest as multiple foci [8]. It is believed that multifocality could be the result of metastatic spread within the CNS, a product of the interaction between tumor cells and extracellular matrix as a consequence of cell adherence with laminins, glycosaminoglycans and integrins, resulting in activation of mechanoreceptors and/or the activation of intracellular pathways involved in cell growth [9].

This spread could be facilitated by the contiguous location of the lesions to the ventricular system, favoring the intrathecal dissemination [10, 11]. Moreover, it is thought that the presence of obstruction to CSF flow, as in the case of our patient, produces an increase in local pressures allowing malignant cells to access the Virchow-Robin perivascular spaces [12]. Despite the delay in treatment because of a previous provisional diagnosis of neurocysticercosis, we did not know the conditions of the first presentation, neither we had complementary studies of that time in order to compare them with the presented images. PAs presenting as multifocal lesions are rare, furthermore clinical and imaging characteristics make this diagnosis challenging. There are a few cases reported in the literature. Unlike our case, Nakano et al. presented a case series of six patients with solitary lesions that had different sizes and heterogeneous contrast enhancement [13]. Similarly, Docampo et al. presented a series of thirty-two patients, none of them with multifocal lesions [1]. On the other hand, and similar to our case, Seranatna et al. reported a single case of a pediatric patient diagnosed with PA with multiple lesions, who had a previous provisional diagnosis of neurocysticercosis, and arrived at definitive diagnosis with biopsy and histopathological studies [14].

Differential diagnosis with neurocysticercosis is not straightforward. If presentation of PA is a cyst-solid, solitary lesion, the distinction would be much more evident. Cystic lesions of cysticercoid origin present in various ways according to the stage of the infection [15]. In the reported case, imaging findings show similar characteristics to that of the vesicular stage, except for the absence of an evident scolex and contrast enhancement on the gadolinium series on T1-weighted images [16]. Although colloidal vesicular lesions usually enhance contrast, they also show a hyperintense aspect of the cystic contents on T1-weighted images, characteristics inconsistent with the findings present in our case [17]. The latter incongruencies made us think about a neoplastic origin of the lesions.

Conclusion

In the present study we have depicted the rare case of multiple and purely cystic PA mimicking neurocysticercosis lesions. We have also denoted the history of a single case of similar presentation in the consulted literature. We agree with the opinion of our colleagues that a complete surveillance of the neuraxis should be performed in the case of multifocal lesions, in addition to the high index of suspicion of neoproliferative disease in the scenario of multiple lesions unresponsive to medical treatment, clinical pearls that may account for a decrease in time-to-diagnosis and initiation of an appropriate treatment. Of great relevance is to note the role of neuroimaging in the differential, including advanced spectroscopic images, and of histopathology in the definitive diagnosis of multifocal lesions in the CNS. Moreover, prognosis declines when the multifocal nature of disease is encountered. Finally, we consider the importance of oncologic treatment in this context where surgical treatment is limited to biopsy acquisition.

Conflict of Interest

The authors declare no conflict of interest.

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Citation: Turco CJ, Carbel GVP, Brasca BRA, Maccio A, Cuevas FG, Weigandt DM. "Pilocytic Astrocytoma Mimicking Neurocysticercosis: Atypical Presentation". SVOA Neurology 3:1 (2022) Pages 06-10.