

Case Report

Pedunculated intraventricular subependymoma: Review of the literature and illustration of classical presentation through a clinical case

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Abstract

Background: Subependymomas are rare benign, noninvasive tumors, classified by the World Health Organization as low grade neoplasms. International data estimate their frequency between 0.2% and 0.7% of the intracranial tumors, and they usually are an incidental finding in autopsies. Preferably located in the fourth ventricle, these tumors tend to become symptomatic when they cause hydrocephalous by obstructing cerebrospinal fluid circulation.

Case Presentation: We present the case of a morbidly obese, hypertense, and diabetic patient, who presented with symptoms of gait ataxia, sphincter incontinence, and dysarthria in relation to a pedunculated subependymoma in the left lateral ventricle. He underwent a biparietal craniotomy with a microscopic microsurgical approach, through which gross total resection was achieved. No perioperative complications ensued.

Conclusions: Given their benign behavior and their excellent response to surgical treatment, subependymomas should be promptly diagnosed and surgically treated to avoid possible neurological damage when they become symptomatic.

Key Words: Gait ataxia, intraventricular mass, pedunculated, subependymoma

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INTRODUCTION

Subependymomas are rare, benign, noninvasive tumors of ependymal origin. Internationally, their frequency is estimated between 0.2% and 0.7% of intracranial tumors, and they are usually incidental findings in autopsies.^[10] These slow-growing neoplasms occur more frequently in middle-aged men, and they are preferably located in the fourth ventricle.^[15] When they become symptomatic, these tumors can present with symptoms secondary to cerebrospinal fluid (CSF) obstruction and intracranial

hypertension, such as positional headache, nausea, vomiting, papilledema, and alterations in consciousness; or secondary to the compression of adjacent structures, such as sensory or motor disturbances, or seizures.^[6,16] Surgical treatment of these lesions, either by means of partial or total resection, usually alleviates the symptoms, and it can provide good long-term tumor control.^[7] We now present the case of a patient who suffered from symptoms of intracranial hypertension due to a pedunculated subependymoma of the left lateral ventricle.

CASE REPORT

A 51-year-old, morbidly obese male, with past medical history of long-standing hypertension and type 2 diabetes mellitus, consulted because of 3 weeks of gait ataxia, sphincter incontinence, and episodic dysarthria of several minutes duration. Initially, the patient was managed by the Neurology Service, who observed wide-based gait, inability to perform tandem gait, globally reduced reflexes and glove and stocking sensory disturbances. Complementary studies revealed polyneuropathy, likely of diabetic origin, and vertebral insufficiency. The patient was therefore managed as an ischemic syndrome.

Nevertheless, the patient also underwent a computerized tomography (CT) scan, and magnetic resonance imaging (MRI), which showed an isodense lesion [Figure 1]. The latter appeared to be located in the left lateral ventricle, where it obstructed the foramen of Monro, thus creating mild hydrocephalus. Subsequently, the patient was transferred to the Neurosurgical Department with the presumptive diagnosis of left ventricle neurocytoma for further management.

Because the patient suffered progressive clinical decline, characterized by dysphasia and paresis of the lower extremities, we placed a ventriculostomy to control his malignant intracranial hypertension. Twenty-four hours later, the patient underwent biparietal craniotomy. We performed a transcallosal approach with microsurgical technique; an anterior longitudinal callosotomy was performed, and a fibroelastic intraventricular mass of whitish coloration, limited by a vascular pedicle, measuring approximately $6 \times 3 \times 4$ cm was resected.

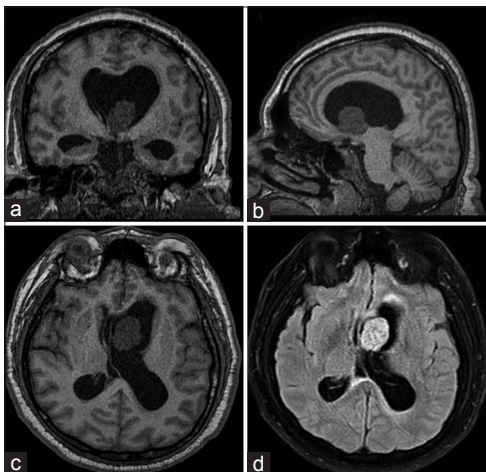


Figure 1: (a) T1-weighted coronal view. Hypointense nodular image in the floor of the left lateral ventricle, producing mild-moderate, noncommunicating hydrocephalus. (b) T1-weighted sagittal view. Note the involvement of the brainstem, which explains the symptoms of the patient. (c) T1-weighted axial view showing a nodular, hypointense lesion with small cystic areas in its interior. (d) Axial view, FLAIR. Note the absence of perilesional edema, and the lack of contrast enhancement

Gross total resection was achieved, and there were no postoperative complications.

Biopsy revealed a tumor with small, rounded nuclei and scarce cytoplasm, and cell groups surrounded by abundant fibrillary matrix and cystic areas. Immunohistochemistry was positive for glial fibrillary acidic protein (GFAP) and negative for neurospecific enolase, which was compatible with grade I subependymoma, according to the World Health Organization (WHO) classification [Figure 2].

The patient is currently asymptomatic of his neurosurgical condition, with complete resolution of his speech and sphincter disorders, and he is completing rehabilitation at the Costa Rican National Rehabilitation Centre for his sensory and motor deficits in the lower extremities. Postoperative imaging did not show tumor recurrence a year after surgery.

DISCUSSION

Initially described by Scheinker in 1945,^[18] subependymoma is a rare, low-grade intracranial tumor, according to the WHO classification. The largest case series published to date describe an incidence of 0.4-0.7%, which is larger in middle-aged or senior males, with a mean age of presentation between 48.5 and 53.6 years.^[3,12,16,19] Epidemiologically, our patient coincides with the international observations reported in the literature.

The fourth ventricle and the lateral ventricles are among the most common locations of subependymomas, with approximate incidences of 60% and 40%, respectively. Nevertheless, the lesions that tend to express themselves clinically are those located in the lateral

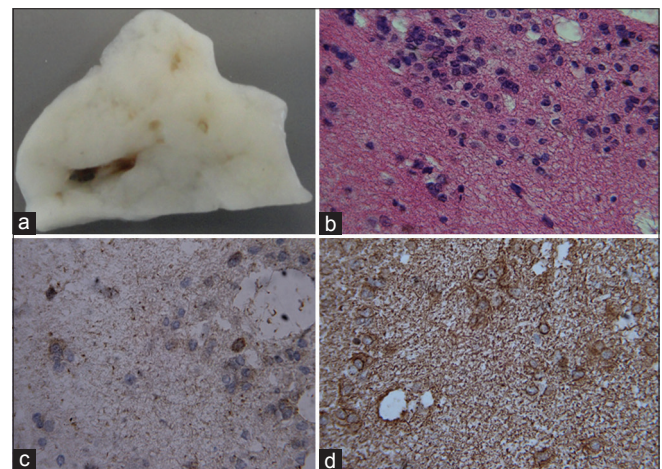


Figure 2: (a) Gross anatomy of resected tumor, showing a fibroelastic, whitish mass. (b) H and E stain of the mass, exhibiting the characteristic rounded cells with small nuclei and scarce cytoplasm over an abundant fibrillary matrix and cystic spaces. Note the absence of atypical mitoses. (c) Immunohistochemistry stain for neurospecific enolase, showing negativity in the majority of the cells. (d) Immunohistochemistry stain for glial fibrillary acidic protein (GFAP), exhibiting strong positivity

ventricles.^[3] Tumors located on the septum pellucidum and the foramen of Monro are more prone to cause CSF obstruction, and typical symptoms include headaches, nausea, vomiting, seizures, ataxia, and vertigo.^[3,4,12,18] The size of the lesion also plays a very important role in the clinical expression of subependymomas; Akamatsu *et al.* found that the majority of symptomatic tumors had a diameter of approximately 4 cm, whereas asymptomatic ones measured 0.8 cm on average.^[1] In their retrospective analysis of subependymomas in the lateral ventricles, Maiuri *et al.* confirmed these data.^[13] Accordingly, our patient presented with symptoms of intracranial hypertension and gait ataxia, and his tumor measured approximately 5 cm in diameter, which is compatible with the data described in the literature.

Radiologically, the appearance of subependymomas can vary depending on their location. According to several studies, subependymomas in the lateral ventricles are reported as well-defined, isodense or hypodense nodular images, and they evince minimal contrast uptake.^[2,11] Imaging differential diagnosis includes ependymomas. One of the factors contributing to their differentiation is that the former ones do not usually present transependymal extension or perilesional edema, whereas the latter ones, in addition to showing contrast uptake, do evince the aforementioned characteristics.^[5] However, there are no pathognomonic imaging signs for one tumor or another, and other differential diagnoses include choroid plexus papilloma, neurocytoma, and giant cell subependymal astrocytoma. In our case, imaging showed an isodense nodular lesion, without perilesional edema, and the first diagnostic possibility was a neurocytoma. These tumors also have predilection for the lateral ventricles, and they can be mistaken for subependymomas because they are isointense in their solid portions on MRI. Furthermore, they tend to present with intracranial hypertension of a few months of course.^[8]

Macroscopically, subependymomas present as fibroelastic, whitish nodules.^[19] One of the intraventricular subependymomas is their growth toward the lumen, with a marked separation between the neoplasm and the normal brain parenchyma. This makes the differentiation between subependymomas and ependymomas easier, since the latter tend to infiltrate the brain parenchyma, while also having ill-defined borders.^[13] The macroscopic characteristics of the tumor of our patient coincided with these classical descriptions. One of the particularities of the case described is that the tumor was attached to the ventricular wall by a vascular pedicle. To date, only five cases of pedunculated subependymomas in the lateral ventricles have been reported.^[9,14,21] It is important to note that in those cases, as well as in ours, the presence of this pedicle made the delimitation of the tumor during the operation much easier, and it became a factor that contributed to the gross total resection of the mass.

Histogenesis of subependymomas is a controversial topic, and several researchers have proposed different cells of origin. Subependymal glia, astrocytes of the subependymal plate, and ependymal cells themselves have all been postulated as likely precursors to subependymomas, but there are not conclusive data.^[4] In spite of this discussion regarding the origin, the histology of subependymomas is well characterized. Nidi of rounded cells with a dense gliofibrillary matrix are classical, and focal microcystic degeneration is not a rare finding, predominantly in lesions located in the lateral ventricles.^[16] In areas of higher cell density, nuclei are sometimes oriented around small-caliber vessels, forming pseudo-rosettes.^[17] Immunohistochemistry of subependymomas is positive for GFAP, S-100 protein, and vimentin.^[20] Markers such as neurospecific enolase, which are commonly positive in neural and neuroendocrine cells, are negative in subependymomas, as well as the markers for mitotic activity MIB-1 and Ki-67.^[16] The biopsy of our patient showed small, rounded cells on a fibrillary matrix with areas of microcysts, as well as positivity for GFAP, which are all compatible findings with this type of tumor.

The treatment of symptomatic subependymomas is complete surgical excision.^[10] Nevertheless, partial resection of these tumors also has favorable results and is acceptable if they are located near critical areas that could become endangered during the surgery;^[6] the surgical goal should be to restore normal CSF flow. Prior to microsurgical techniques, perioperative mortality rates reached up to 28.8%.^[3] Currently, with technological and neurosurgical advances, this is just an anecdotal fact, and patients rarely experience surgery-related complications.^[7,13,15,17] Our patient also underwent a craniotomy without any complications.

In conclusion, subependymomas are rare intracranial tumors of benign behavior. Nevertheless, their size and location can produce symptoms because of CSF obstruction or parenchymal irritation. Even though the imaging characteristics are not pathognomonic, these neoplasms have a distinctive histology. Because of their good prognosis and excellent response to surgical management, prompt and accurate diagnosis is vital to provide our patients with good outcomes.

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