

# Cystic meningiomas: beyond the solid tumor, a challenge in diagnosis

## *Meningiomas quísticos: más allá del tumor sólido, un desafío en el diagnóstico*

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### ABSTRACT

**Background.** Cystic meningiomas (CMs) are rare variants of meningiomas characterized by atypical radiological and histopathological features. These lesions imply significant diagnostic challenges due to their resemblance to other cystic brain lesions, such as gliomas, metastases, arachnoid cysts, and neurocysticercosis in endemic areas. This report aims to highlight the clinical and imaging findings that aid in distinguishing cystic meningiomas from other lesions, and to discuss their surgical management. **Case Presentation.** We describe two cases of cystic meningiomas in adult patients. The first case involves a 59-year-old patient who presented with tonic-clonic seizures, initially misdiagnosed with racemose neurocysticercosis. Imaging studies revealed a type IV cystic meningioma, successfully treated with surgical resection. The second case describes a 58-year-old patient with progressive lower limb weakness and diplopia. Imaging identified a type III cystic meningioma, which was also managed with complete surgical resection. Both patients recovered well postoperatively, while histopathological diagnoses confirmed benign transitional meningiomas. **Conclusions.** Cystic meningiomas are uncommon but crucial differential diagnoses for cystic brain lesions. Early recognition and appropriate surgical management are essential to ensure favorable outcomes. Imaging techniques, particularly MRI, play a paramount role in diagnosis, while histopathological evaluation confirms the nature of the lesion.

**Keywords:** meningioma; cystic lesion; brain tumor; neuro-oncology; atypical; neurocysticercosis.

### RESUMEN

**Antecedentes.** Los meningiomas quísticos (MC) son variantes raras de meningiomas que se caracterizan por características radiológicas e histopatológicas atípicas. Estas lesiones implican desafíos diagnósticos significativos debido a su semejanza con otras lesiones cerebrales quísticas, como gliomas, metástasis, quistes aracnoideos y neurocisticercosis en áreas endémicas. Este informe tiene como objetivo resaltar los hallazgos clínicos e imagenológicos que ayudan a distinguir los meningiomas quísticos de otras lesiones y discutir su manejo quirúrgico. **Presentación del caso.** Describimos dos casos de meningiomas quísticos en pacientes adultos. El primer caso involucra a un paciente de 59 años que presentó convulsiones tónico-clónicas, inicialmente diagnosticado erróneamente con neurocisticercosis racemosa. Los estudios de imagen revelaron un meningioma quístico tipo IV, tratado exitosamente con resección quirúrgica. El segundo caso describe a un paciente de 58 años con debilidad progresiva de las extremidades inferiores y diplopía. Las imágenes identificaron un meningioma quístico tipo III, que también se trató con resección quirúrgica completa. Ambos pacientes presentaron una recuperación satisfactoria después de la operación, mientras que el diagnóstico histopatológico confirmó meningiomas transicionales benignos. **Conclusiones.** Los meningiomas quísticos son diagnósticos diferenciales poco frecuentes, pero cruciales para las lesiones cerebrales quísticas. El reconocimiento temprano y el tratamiento quirúrgico adecuado son esenciales para asegurar un pronóstico favorable. Las técnicas de imagen, en particular la resonancia magnética, desempeñan un papel fundamental en el diagnóstico, mientras que la evaluación histopatológica confirma la naturaleza de la lesión.

**Palabras clave:** meningioma; lesión quística; tumor cerebral; neurooncología; atípico; neurocisticercosis.

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## 1 INTRODUCTION

Meningiomas are tumors described since the 17th century as a pathological entity. However, it was not until the 18<sup>th</sup> - 19<sup>th</sup> centuries that the diagnosis of this pathology began, in relation to documented morphological changes on the surface of the cranial vault secondary to the hyperostosis they cause. It was not until 1915 that Cushing and Weed employed the term “meningothelioma”, later in 1922 in Cavendish, Cushing renamed them as “meningiomas”.<sup>1</sup>

These lesions comprise a group of neoplasms derived from meningotheelial cells of the arachnoid. They usually constitute grade 1, 2, or 3 lesions according to the World Health Organization (WHO). They represent about 37.6% of all central nervous system (CNS) tumors, thus being the most frequent CNS tumor; however, it remains as the least common in children. Meningiomas affect women 2.32 times more than men, particularly in premenopausal period, and 9.25 times more common in black individuals compared to 7.88 times in white individuals.<sup>2</sup>

Meningiomas have multiple classifications according to their site of origin, histological characteristics and immunohistochemical features, only to mention few. Their most common presentation is described as slow-growing solid lesions, which are predominantly benign. It is rare to find a cystic meningioma (CM) with atypical behavior.

In these case reports, we describe a CM, clinical manifestations, behavior, imaging and the management provided.

## 2 CASE PRESENTATION

### Case 1

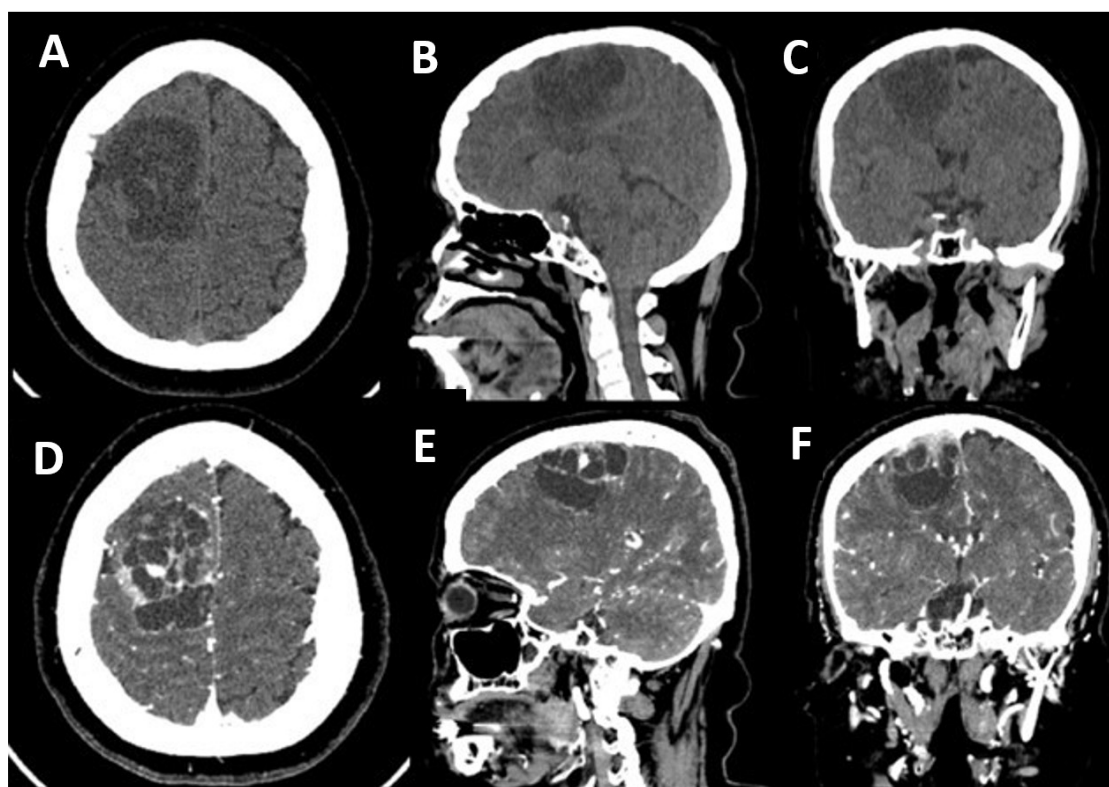
A female 59-year-old patient presented to the emergency department (ED) with generalized tonic-clonic seizures. A computed tomography (CT) scan (Figure 1) identified a heterogeneous cystic extra-axial lesion in the frontal convexity, with a broad base causing displacement of the adjacent parenchyma. The lesion showed intense enhancement of the septa and the solid component, with a broad implantation base

adjacent to the *falx cerebri*. The patient was diagnosed by a neurologist with racemose neurocysticercosis and was treated with antiparasitic medication (Albendazole 400 mg orally *tid* for 4 weeks) which showed no response. Subsequently, the patient developed hemiparesis. Neuroimaging by magnetic resonance imaging (MRI) (Figure 2) revealed an extra-axial broad-based lesion, hypointense in T1, hyperintense in T2 due to the extensive cystic component with multiple septa, hyperintense in FLAIR, with marked enhancement of the septa and central solid portion in contrast-enhanced T1. Diffusion-weighted imaging (DWI) showed few peripheral areas of restriction, without hemorrhagic residues or calcific deposits on susceptibility-weighted imaging (SWI). These findings categorized the lesion as a type IV cystic meningioma according to Nauta et al. classification. Spectroscopy (Figure 3) with short echo time showed an alanine peak, decreased Cho and Cr peaks, and absence of NAA, a characteristic pattern of meningiomas. The patient underwent surgical intervention, revealing an irregular cystic lesion with xanthochromic content, with multiple septa and dural implant towards the falx. A grade 1 Simpson resection was achieved. The patient was discharged 48 hours after surgery without additional deficits. Ulterior pathology analysis reported a WHO grade 1 transitional meningioma (Figures 4 and 5).

### Case 2

A 58-year-old patient presented to the ED with decreased strength in the lower limbs, associated with diplopia and drowsiness. Imaging studies were requested. Cranial non-contrasted CT scan (Figure 6) identified a nodular lesion with a solid appearance and a peripheral cystic component at the midline below the falx, molding the inner table. After the administration of contrast agent, the solid component showed intense and heterogeneous enhancement with small central cystic areas, maintaining an adequate interface with the cystic component. This caused a caudal displacement of the anterior cerebral arteries and ventral displacement of the corpus callosum, and was associated with edema of the surrounding white matter.

Non-contrasted MRI (Figure 7) confirmed the cystic component as isointense to fluid in all sequences. The solid component showed low signal intensity on T1 and intermediate signal intensity on T2, with hemosiderin deposits visible on SWI. Contrasted MRI revealed heterogeneous enhancement of the solid component and the adjacent pachymenings.



**Figure 1.** Non-contrasted CT scan in axial (A), sagittal (B), and coronal (C) views. A heterogeneous extra-axial broad-based cystic lesion is identified in the frontal convexity, causing displacement of the adjacent parenchyma. Contrast-enhanced CT scan, in axial (D), sagittal (E), and coronal (F) views, shows intense enhancement of the septa and the solid components, with a broad implantation base adjacent to the *falx cerebri*.

Extensive vasogenic edema of the bilateral frontal white matter was also noted. These findings categorized the lesion as a type III cystic meningioma according to Nauta et al. classification. Surgical intervention was performed, achieving complete resection of the lesion without deficits, and the patient was subsequently discharged.

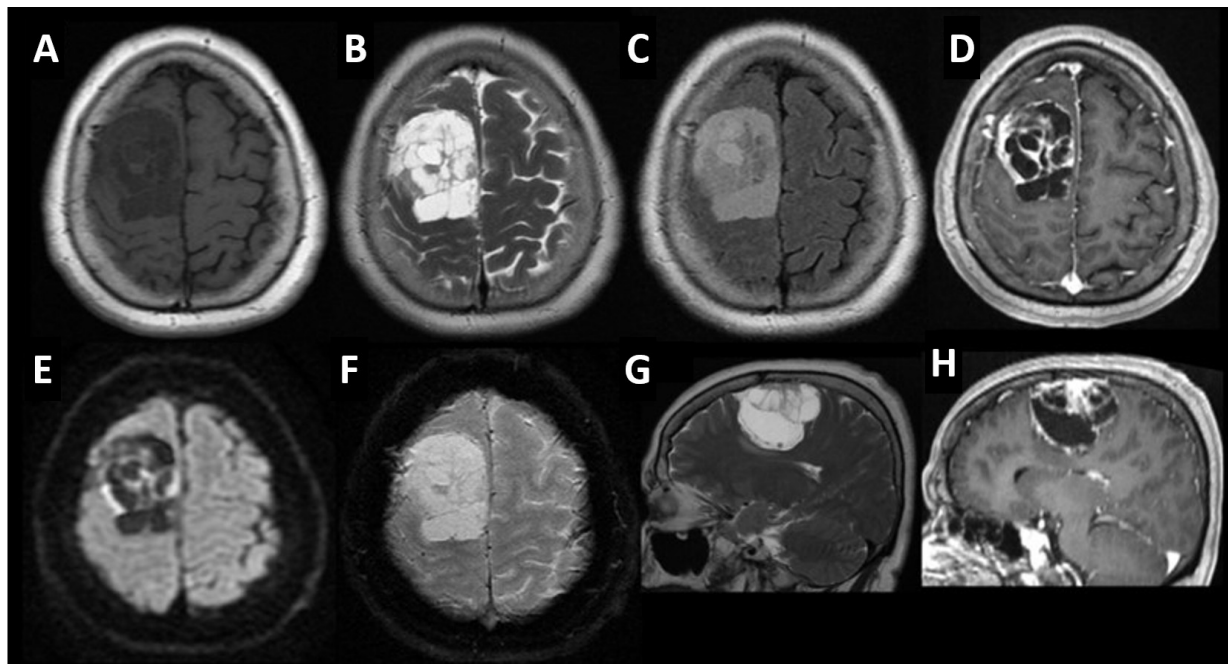
### 3 DISCUSSION

Meningiomas are the most frequent CNS tumors, accounting for one-third of all such tumors, representing 36.6% of all CNS tumors and 53.2% of benign CNS tumors<sup>3</sup>; however, cystic presentation is uncommon. The first cystic meningioma (CM) was reported by Penfield<sup>4</sup> in 1932. They represent 2-7% of all meningiomas in adults, and 10-18% in children. Radiological diagnosis is challenging due to multiple differential diagnoses with other cystic lesions such as hemangiopericytoma,

high-grade gliomas, hemangioblastoma, neurocysticercosis, and metastases<sup>5</sup>.

Anatomically, CMs are primarily located in the frontoparietal region<sup>6</sup>, predominantly towards the convexity. Clinically, their most frequent manifestations are headache, seizures, and behavioral disorders<sup>7</sup>.

Meningiomas are often seen as solid masses with typical radiological features. However, as mentioned before, CM show atypical findings<sup>8</sup>. They can be classified as benign, malignant, or atypical based on histological characteristics, which determine their aggressiveness and invasion level. Several studies have documented that aggressive behavior of the lesions is highly associated with the presence of cysts or invasion through the skull base foramina<sup>9</sup>, making these lesions difficult to diagnose due to their high correlation with high-grade gliomas, metastatic tumors, hydatid cysts, hemangiopericytomas, and arachnoid cysts, thus making the diagnosis of CM a substantial challenge<sup>10</sup>.



**Figure 2.** MRI identifying the extra-axial broad-based lesion hypointense on T1 (A), hyperintense -due to the extensive cystic component with multiple septa- on axial T2 (B) and sagittal T2 (G), hyperintense on FLAIR (C), with notorious enhancement of the septa and the central solid portion on contrast-enhanced axial (D) and sagittal (H) T1 sequence. Diffusion-weighted imaging shows peripheral areas of restriction (E), without hemorrhagic residues or calcific deposits on SWI (F).

Various authors have described the characteristics of CMs heading to classifications such as the one made by Nauta et al.<sup>11</sup>. The team classified CMs into four types (Figure 8): In type 1, the cystic component is completely contained within the tumor as a core. In type 2, the cyst is located peripherally but remains encapsulated by tumor cells. In type 3, the cyst is located peripherally in the adjacent parenchyma unattached but close to the tumor. In type 4, the cyst is located between the tumor and the brain, resembling cerebrospinal fluid in the subarachnoid space, thus found outside the tumor or brain parenchyma (Table 1)<sup>11,12</sup>.

Another classification is described by El-Fiki et al.<sup>13</sup>, who correlated the location of the cyst and its content to divide them into 4 types.

In type A, the cyst is extratumoral, and its content is similar to cerebrospinal fluid (CSF). It can be subdivided into type A1 if the fluid is surrounded by an arachnoid membrane and type A2 if it is not. In type B, the cyst content has xanthochromic

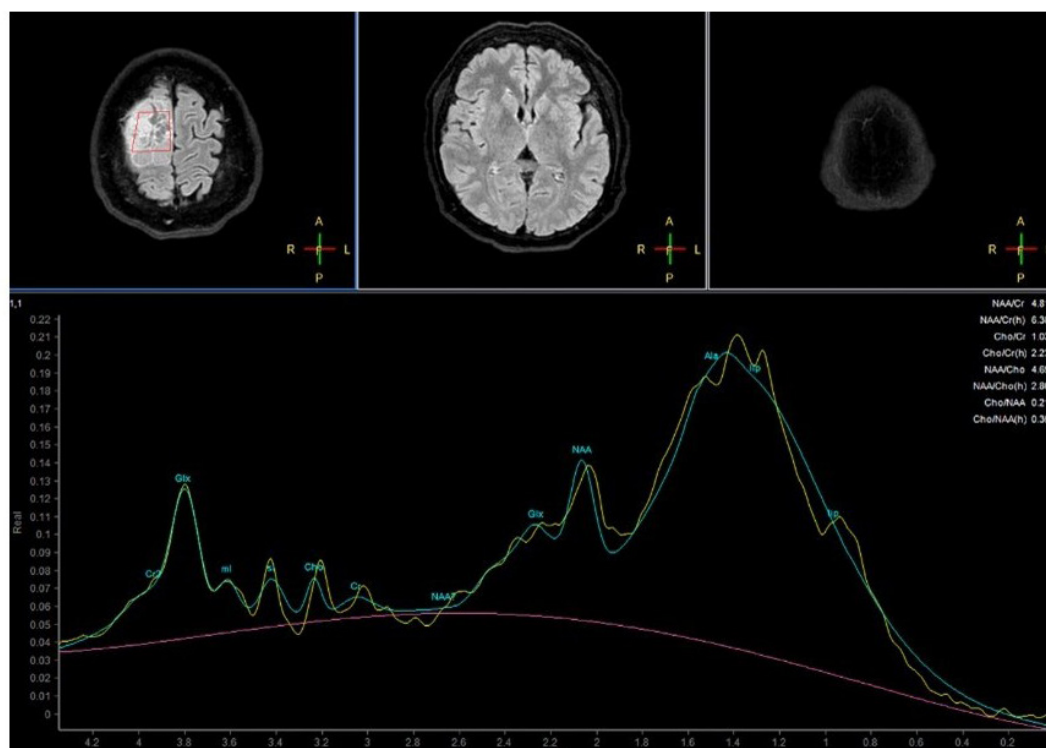
characteristics. It is subdivided into type B1 when the cyst does not have a defined wall and type B2 when the cyst has a thin wall with surrounding edema. In type C, the cyst contains yellow to dark brown fluid accompanied by marked peritumoral edema. Type D includes meningiomas with the presence of extratumoral or peritumoral clear cysts and small intratumoral dark brown cysts (Table 2)<sup>13</sup>.

Jung et al.<sup>14</sup>, based on the proposal by Nauta et al.<sup>11</sup>, classified cystic meningiomas into five types, in relation to their variability:

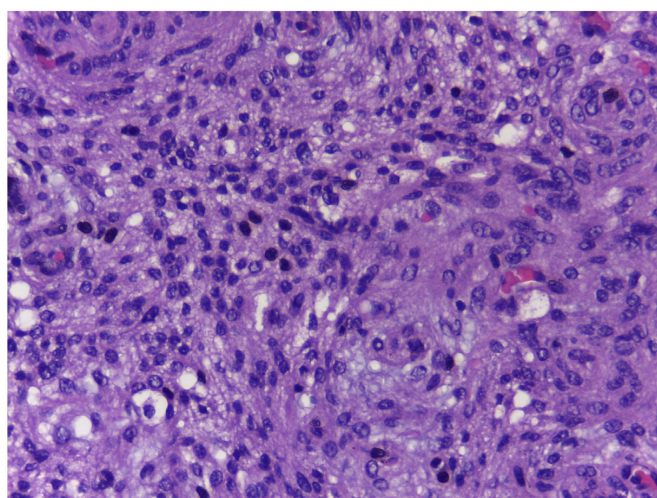
Type I: intramural cysts located centrally within the tumor. Type II: intramural cysts located peripherally and surrounded by a meningioma. Type III: peritumoral cyst in the adjacent parenchyma. Type IV: peritumoral cyst between the tumor and the parenchyma. Type V: a combination of type I and type III (Table 3).

In the diagnostic approach, CT is the first imaging modality used. It allows the identification of bone changes associated with meningiomas, such as hyperostosis (25-49%)<sup>15</sup>.





**Figure 3.** Spectroscopy with short echo time, showing an alanine peak, decreased Cho and Cr peaks, and absence of NAA, strongly associated characteristics of meningiomas.

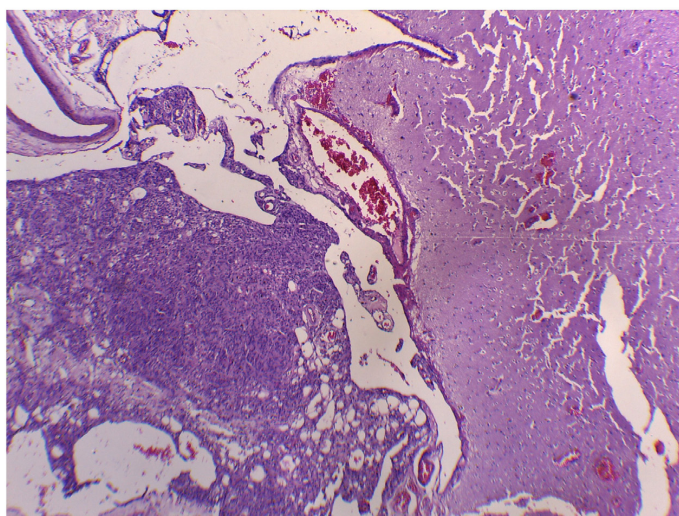


**Figure 4.** Histological study which shows evidence of a lesion composed of solid layers of meningotheelial cells with whorled fascicles.

Cystic meningiomas present different characteristics on MRI. Zhang et al.<sup>16</sup> documented that peritumoral cystic meningiomas are predominantly located in the *falx cerebri*,

while intramural cystic meningiomas are located in the frontal convexity. The cystic component is larger in the intratumoral group, whereas perilesional edema is more pronounced in the intramural cyst group<sup>17</sup>.

MRI is considered the ideal diagnostic method by imaging, not only for the study of brain tumors, but for several SNC lesion. For CMs, the solid component shows intermediate to hypointense signal intensity on T1 and intermediate to hyperintense on T2, with marked enhancement in the post-contrast phase. The cystic component shows isointense behavior to fluid in all sequences. In diffusion techniques, meningiomas exhibit different behavior depending on the degree of cellularity, usually showing restricted movement in the solid component and facilitated movement in the cystic area of the lesion<sup>6</sup>. Generally, the solid content of these lesions consists of hypervascular, highly perfused masses which increase perfusion curves, thus reflecting an increase in relative cerebral blood flow (rCBF), with reported values between 6 and 9 ml/100g<sup>18</sup>.



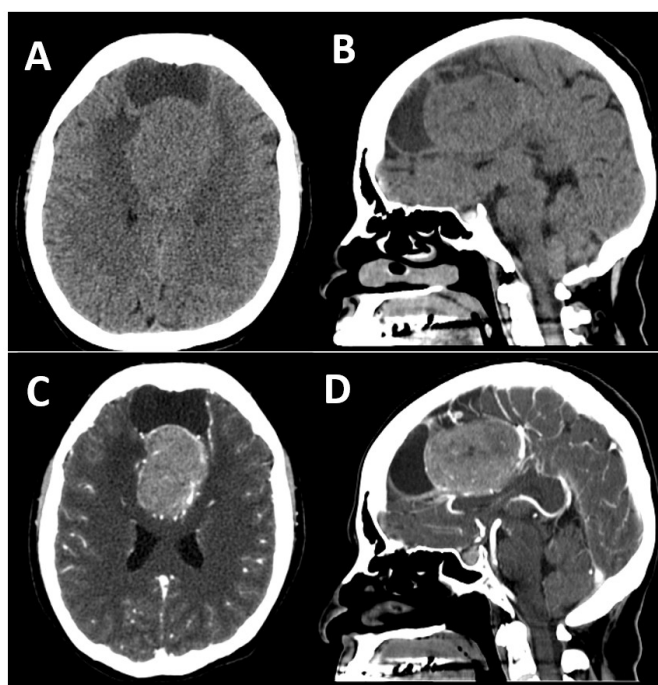
**Figure 5.** Histological study shows no evidence of mitotic activity, necrosis, or cortical infiltration.

**Table 1.** Nauta et al.<sup>11</sup> Classification.

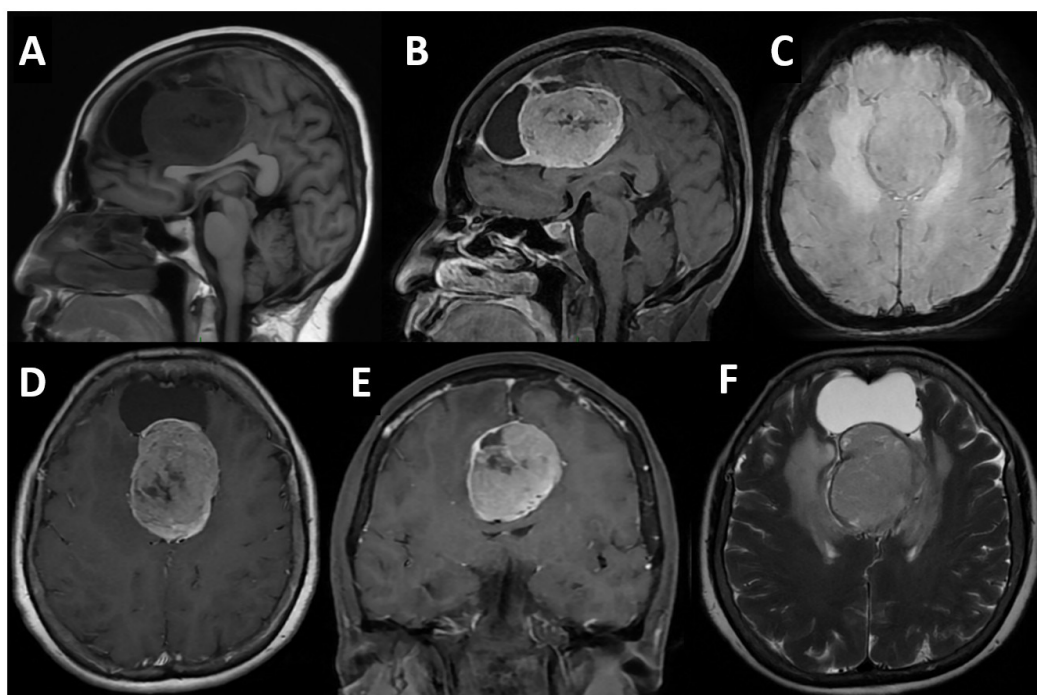
|        |                                                                                                                                                                                     |
|--------|-------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| Type 1 | The cystic component is completely contained within the tumor and located centrally.                                                                                                |
| Type 2 | The cyst is entirely intratumoral but located peripherally and encapsulated by tumor cells.                                                                                         |
| Type 3 | The cyst is located peripherally and found in the adjacent parenchyma.                                                                                                              |
| Type 4 | The cyst is confined to the interface between the tumor and the brain, as cerebrospinal fluid in the subarachnoid space, and is therefore not within the tumor or brain parenchyma. |

**Table 2.** El-Fiki et al.<sup>13</sup> Classification.

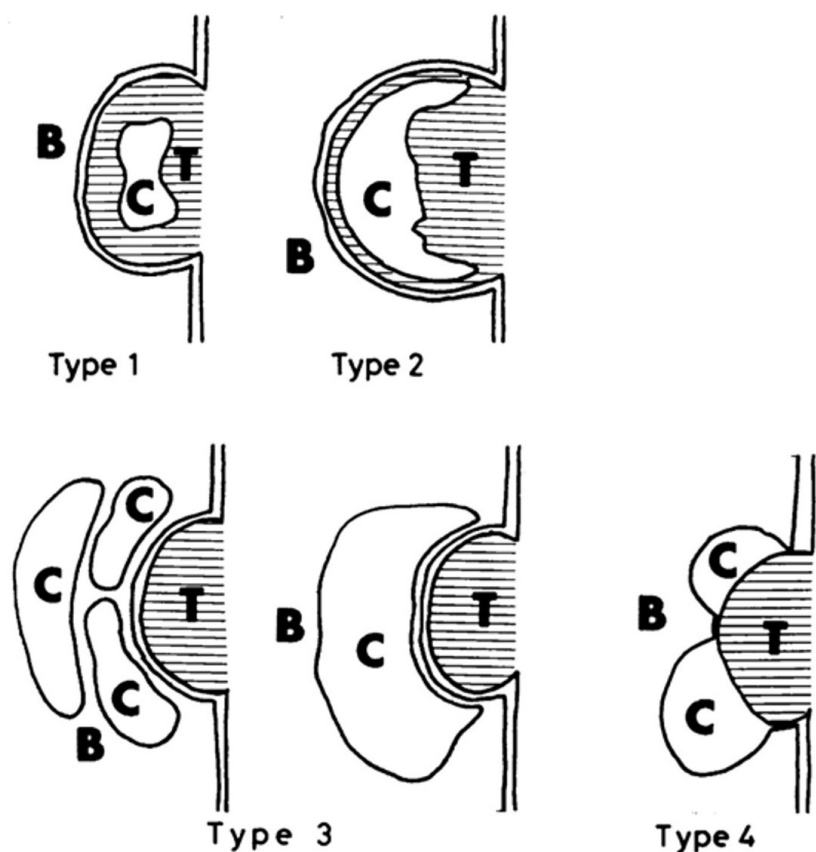
|        |                                                                                              |    |                                                   |
|--------|----------------------------------------------------------------------------------------------|----|---------------------------------------------------|
| Type A | Extratumoral cyst with content similar to CSF                                                | A1 | Fluid is surrounded by an arachnoid membrane.     |
|        |                                                                                              | A2 | Fluid is not surrounded by an arachnoid membrane. |
| Type B | Cystic content with xanthochromic characteristics                                            | B1 | The cyst does not have a defined wall.            |
|        |                                                                                              | B2 | The cyst has a thin wall with surrounding edema.  |
| Type C | The cyst contains yellow to dark brown fluid accompanied by marked peritumoral edema.        |    |                                                   |
| Type D | Presence of extratumoral or peritumoral clear cysts and small intratumoral dark brown cysts. |    |                                                   |



**Figure 6.** Non-contrasted CT scan in axial (A) and sagittal (B) views showing a heterogeneous subfalcine lesion. After contrast administration, the axial (C) and sagittal (D) views show a rostral cystic component and heterogeneous enhancement of the caudal solid component.



**Figure 7.** Non-contrasted MRI T1 sequence in sagittal view (A) shows the lesion causing ventral and caudal displacement of the surrounding structures. Post-contrast sagittal, axial, and coronal sequences (B, D, and E, respectively) showing intense and heterogeneous enhancement of the solid component and dural enhancement. Axial SWI sequence (C) with hypointense foci related to hemosiderin deposits. Axial T2 sequence (F) enlightens the cystic component as isointense to fluid and vasogenic edema of the white matter.



**Figure 8.** The image shows the four possible configurations of a tumor (T), a cyst (C), and the brain (B). Original image acquired from the classification by Nauta et al.<sup>11</sup>.

**Table 3.** Jung et al.<sup>14</sup> Classification based on the proposal by Nauta et al.<sup>11</sup>.

|        |                                                                       |
|--------|-----------------------------------------------------------------------|
| Type 1 | Intramural cysts located centrally within the tumor.                  |
| Type 2 | Intramural cysts located peripherally and surrounded by a meningioma. |
| Type 3 | Peritumoral cyst in the adjacent parenchyma.                          |
| Type 4 | Peritumoral cyst between the tumor and the parenchyma.                |
| Type 5 | Combination of type 1 and 3.                                          |

Regarding the functional spectroscopy sequence with short echo time, elevated levels of alanine (1.3 to 1.5 ppm) and choline are usually detected, with a reduction and/or absence of N-acetylaspartate and creatine. Although elevated alanine has been considered typical in meningiomas, it is not detected in all meningiomas; however, its appearance may inversely correlate with the degree of tumor necrosis<sup>19</sup>.

The treatment is surgical, and complete resection of the lesion along with resection of the cyst capsule is primarily indicated for those classified as type II according to Jung et al.<sup>14</sup>.

#### 4 CONCLUSIONS

Cystic meningiomas (CMs) are rare SNC neoplastic lesions. Imaging findings role paramount aids in differential diagnosis; however they remain a diagnostic challenge for the specialist. Treatment is usually surgical, chiefly when symptoms are present. Histological study is fundamental for the appropriate categorization of the tumor.



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