

Case Report

Gigantic Exteriorized Frontal Meningioma: An Exceptional Clinical Entity



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ABSTRACT

Background and Aim: Meningiomas are usually benign tumors. Depending on their location and size, dealing with them can be problematic. We report an exceptional case of gigantic frontal meningioma. Its diagnosis was misleading and the excision laborious.

Case Presentation: A 29-year-old patient who presented gradually over 5 years, developed behavioral disorders then swelling of the vertex at the frontal level and decreased visual acuity. Physical examination on admission revealed a median frontal swelling slightly lateralized to the left axis of approximately 10 cm, hard, covered with the healthy, severe frontal syndrome, and decreased visual acuity. Computerized tomography (CT) scan showed a voluminous medial frontal lesion lateralized to the left, with irregular limits, superior sagittal sinus invasion, brain engagement under the scythe, heterogeneous contrast taking, and significant osteolysis first evoking tumor metastasis. Magnetic resonance imaging (MRI) suggested a meningioma. The resection was macroscopically complete. Dural and cranial plasty was performed. The postoperative follow-up was simple. Histology concluded that the meningioma was grade 1 based on the World Health Organization (WHO) classification.

Conclusion: Late-diagnosed meningiomas can take on very large proportions and pose management problems while seriously complicating the patient's functional and vital prognosis.

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Highlights

- The tumor size is important as it can cause severe psychic disorders and regress after the surgery.
- The medical imaging was very atypical so the diagnosis was almost a surprise on histology.
- There was a significant operative risk due to the large size of the tumor and its invasion to the upper sagittal sinus, and neovascularization.

Plain Language Summary

Meningiomas are generally benign tumors. They can evolve quietly for several years and take on significant proportions that can compromise the functional and vital prognosis. Depending on their location and size, treating them can be difficult. We report an exceptional case of a 29-year-old patient whom we treated after 5 years of the progressive appearance of swelling on the skull, severe psychic disorders, and visual disturbance. The mental disorders that worried the patient's family the most were initially taken care of in a traditional way, which favored this long time in treatment by modern medicine. Upon admission to the hospital, diagnosis, and surgical treatment were difficult due to the very large and unusual size of this tumor. In terms of medical imaging, the cerebral CT scan, the most common examination, no longer evoked the diagnosis. MRI was performed which is less available. The surgical treatment could be carried out at the cost of significant sacrifices of certain anatomical structures which were replaced by synthetic material. However, after treatment, the evolution was satisfactory, marked by a regression of psychic disorders. This case illustrates the fact that late diagnosis of meningioma can take on very large proportions and pose problems of management while seriously affecting the functional and vital prognosis of the patient.

1. Background and Importance

Meningiomas are benign tumors that grow very slowly over years before becoming symptomatic. Their discovery can be fortuitous or in the presence of certain symptoms (headaches, epilepsy, or neurological deficit) [1]. Their excision is sometimes laborious, especially when they sit near or invade a vital anatomical structure [2]. We report an exceptional case of gigantic frontal meningioma which posed not only diagnostic but also therapeutic difficulties due to its size and invaded or compressed anatomical structures.

2. Case Presentation

A 29-year-old patient, a farmer resident in a rural environment, presented behavioral disorders over 5 years, followed by swelling of the vertex at the frontal level and decreased visual acuity. No vomiting or nausea was observed. The patient reported a long history of moderate and intermittent headaches as a pathological history. In addition, the patient was kept at the traditional practitioner's home several times for an average of 1 to 2 months for intensive care. Physical examination on admission to our department in August 2021 noted a median frontal swelling slightly lateralized to the left

of approximately 10 cm long axis, hard, covered with healthy skin traversed by large dilated veins, a severe frontal syndrome (aggressiveness, refusal to cooperate, incoherent talk, urination without discomfort: and visual acuity of 2.10 on the left and 3.10 on the right).

Computerized tomography (CT) scan revealed a voluminous median frontal lesion lateralized to the left, measuring 101 x 85 x 77 mm, with irregular boundaries, peripheral edema, invasion of the superior sagittal sinus, and brain engagement under the scythe. This lesion took up the contrast product in a heterogeneous way and caused significant osteolysis. The whole thing first evoked a tumor metastasis. The thoraco-abdomino-pelvic CT scan done in search of a primary tumor was normal. Magnetic resonance imaging (MRI) showed a fleshy lesion with more regular boundaries and more homogeneous contrast uptake than CT, firstly suggesting an aggressive meningioma (Figure 1).

The patient was operated two months after admission. In the absence of the possibility of the tumor preoperative embolization, to reduce blood loss, the scalp's large dilated veins were ligated before the skin incision. Similarly, the superior sagittal sinus was ligated in its anterior third. This ligation of the superior sagittal sinus was done immediately after skin detachment by

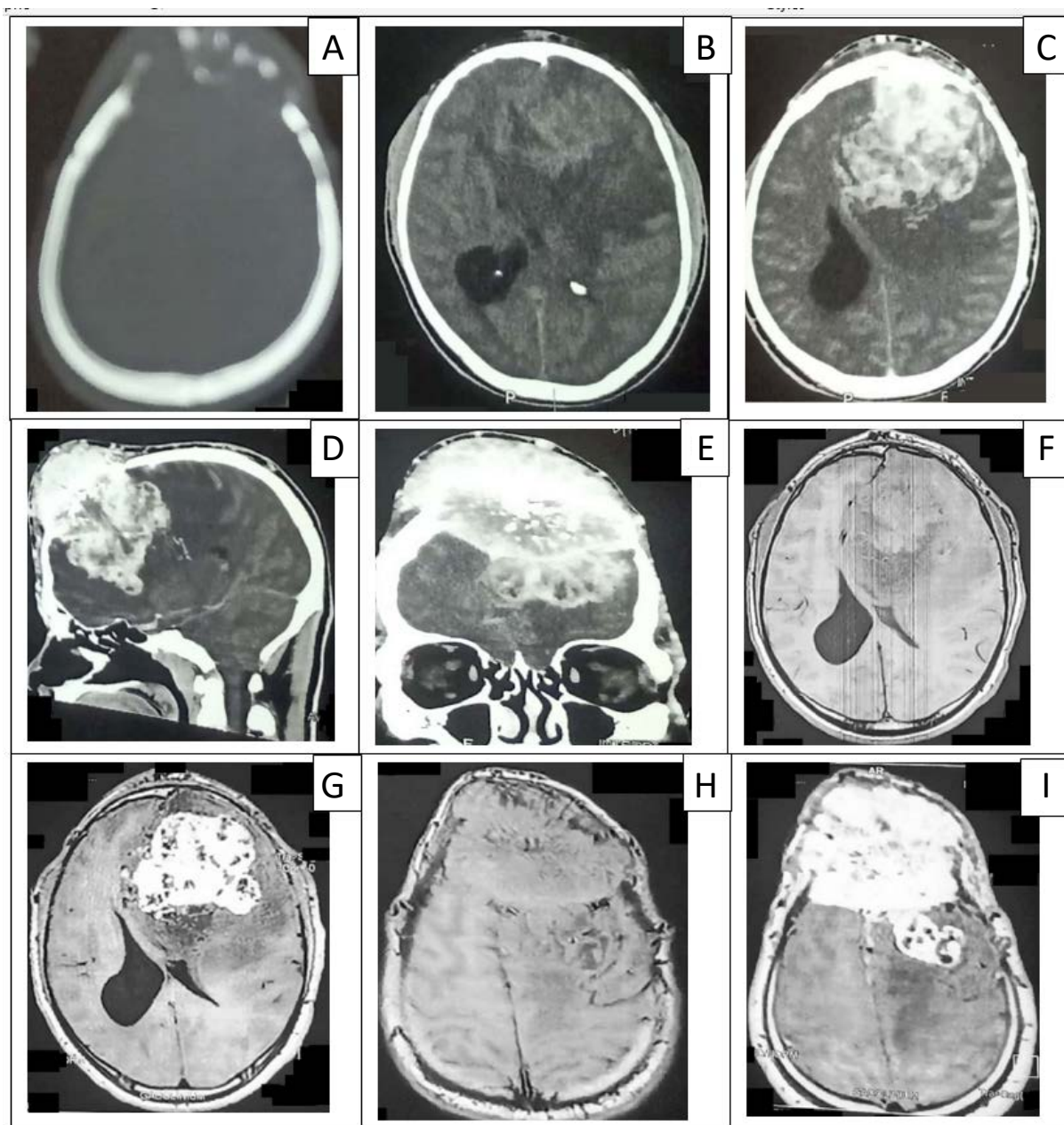


Figure 1. Pre-operative imaging. Preoperative computerized tomography (CT) scan, axial section in bone window (A); parenchymal window without (B); and with (C) contrast product injection; computerized tomography (CT) injected in sagittal (D) and coronal (E) reconstruction; magnetic resonance imaging (MRI) in T1-weighted sequence in axial section passing through the Ventricles without (F) and with (G) contrast products injection; magnetic Resonance imaging (MRI) in T1-weighted sequence in axial section passing over the ventricles without (H) and with (I) contrast products injection

These preoperative images show a voluminous median frontal lesion lateralized on the left, with irregular borders, heterogeneously enhanced by the contrast product, peripheral edema, invasion of the superior sagittal sinus, brain engagement under the scythe, and significant osteolysis.

an enlarged burr hole on healthy bone centered on the superior sagittal sinus without touching the tumor. After significantly reducing the hemorrhagic risk, we made 6 burr holes around the tumor then we cut the bone

which was between these holes with a Gigli saw. Thus, the invaded bone and part of the tumor were removed. The remnant of the tumor that had burrowed into the brain parenchyma was carefully dissected and removed.

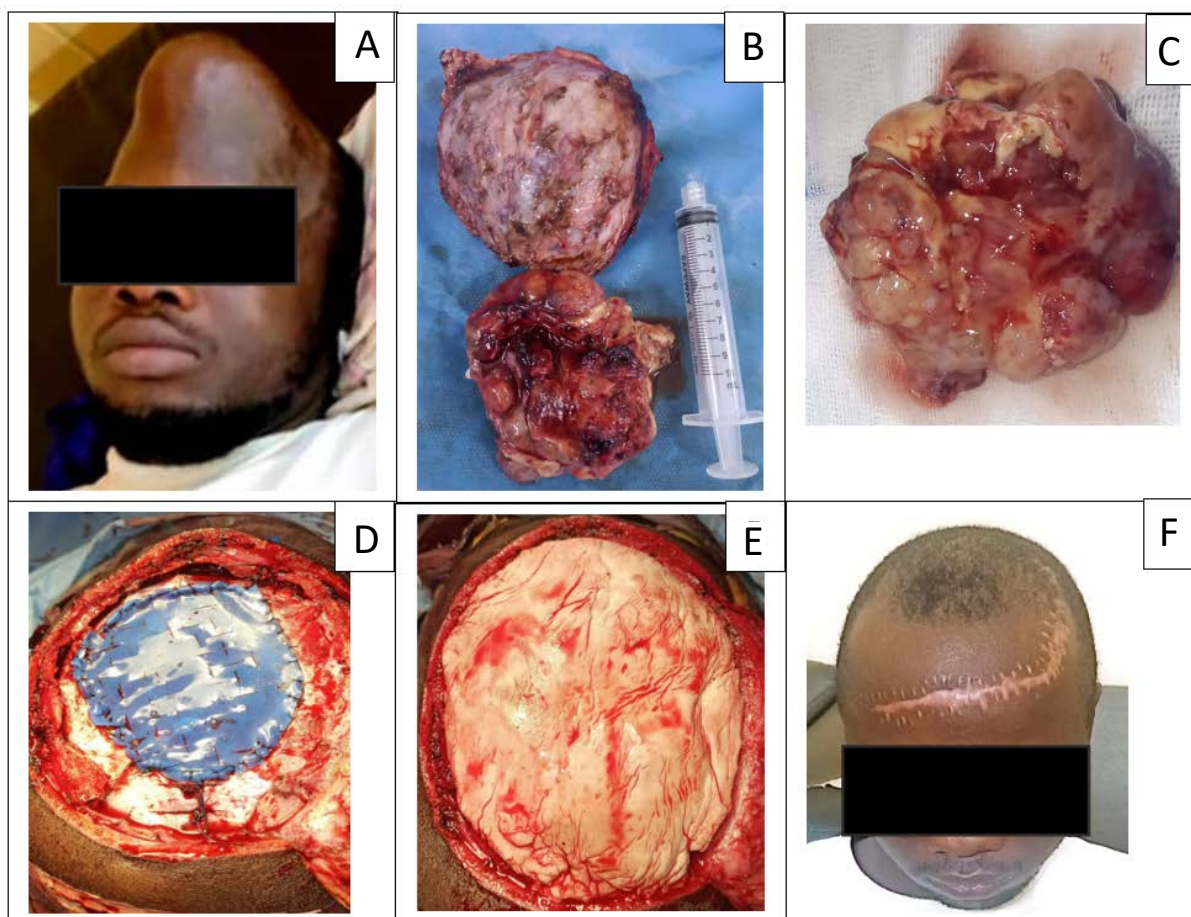


Figure 2. Pre, per, and 4-month postoperative photos, the patient's clinical appearance preoperatively (A) showing a voluminous median frontal swelling slightly lateralized to the left, covered with healthy skin large dilated veins, operative specimen showing the exocranial and endocranial aspect of the invaded Bone (B); aspect of the intra parenchymatous portion of the tumor (C), peroperative images of duroplasty (D) and cranioplasty (E), clinical appearance 4 months postoperative for the patient (F) showing disappearance of the swelling

The resection was macroscopically complete. Dural and cranial plasty was performed because the dura and the bone were destroyed by the tumor. A transfusion of 2 bags of red blood cells was performed before the patient awoke (Figure 2).

The anatomical-pathological study of the surgical specimen showed under microscopy a tumoral proliferation organized in areas or bays within a richly vascularized fibrous stroma. These vessels had a hyalinized wall and a regular endothelial lining. Tumor cells were rounded or polygonal with ill-defined boundaries and fibrillar cytoplasm. They were provided with oval, vesicular, finely nucleolated nuclei. No necrosis, mitosis, or capsular crossing existed. This histological aspect led to the conclusion of an angiomatous meningioma grade 1 of the World Health Organization (WHO) 2016 (Figure 3). The postoperative course was simple. After a 4-month follow-up, the frontal syndrome almost disap-

peared, no more aggressiveness was observed, and the patient was more cooperative. He just kept a little logorrhea with coherent words. Visual acuity increased to 5/10 in both eyes. At 16 months, visual acuity increased to 8.10 in both eyes; the neurological examination of the patient was normal apart from slight psychic disorders compatible with living in society and the cerebral scanner showed no tumor recurrence (Figure 4).

3. Discussion

Extracranial extension of intracranial meningioma is unusual and is accompanied by skull osteolysis [3]. Few cases have been described, 2 cases in Senegal in 1971 [4], 1 case in Korea in 2014 [3], 1 case in Kosovo in 2015 [5], 3 cases associated with neurofibromatosis type 2 in 2016 in Germany [6]. The largest cases were located on the polar front [3, 5] as was the case in our patient. In our situation, the low socio-economic level of the

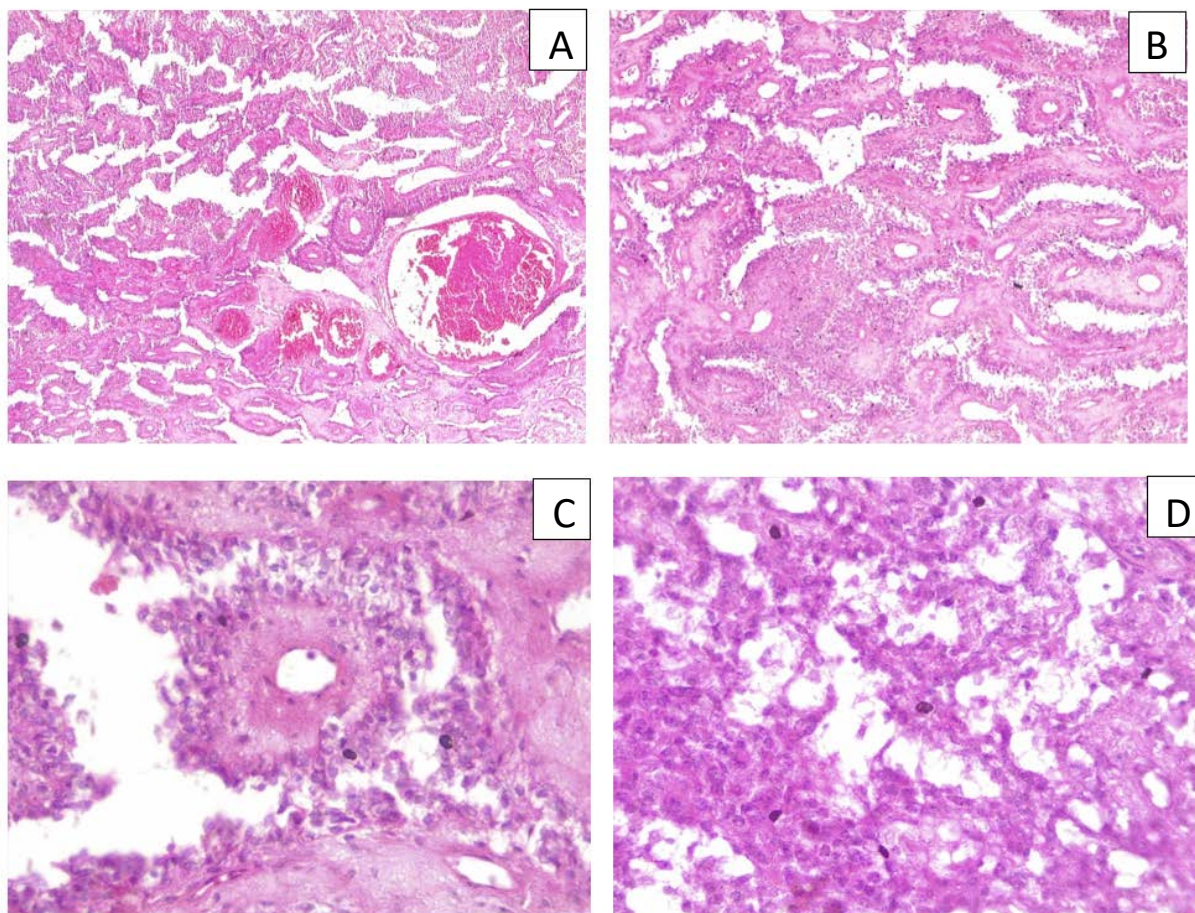


Figure 3. Histological aspect of the surgical specimen; showing an angiomatous meningioma, (A) and (B) low magnification (Gx40): proliferation of round and polygonal cells in fibrous stroma with many vessels hyalinized, congestive and hemorrhage, (C) and (D) high magnification (Gx100): aspect of cells

population favors the false ancestral beliefs, saying that the only treatment for any psychic disorder, also called “madness”, is traditional. Anything that promotes the disease development and will not be diagnosed without difficulty at a very advanced stage. Late diagnosis meningioma can take on very large proportions and pose management problems while seriously affecting the functional and vital prognosis of the patient. Thus, in addition to the local malignancy linked to the meningioma excessive development, distant metastases have been described in patients with atypical or anaplastic meningiomas [7, 8]. First-line treatments are observation and surgery, but adjuvant radiotherapy/radiosurgery is discussed for atypical meningiomas and indicated for anaplastic meningiomas [2]. Surgical excision should be as complete as possible, and the dura and the destroyed bone should be replaced as was the case in our observation. Cranioplasty and duroplasty were performed [3, 5] after subtotal [3] or macroscopic total [5] resection.

On anatomical pathological, this type of aggressive lesion destroying the bone and the dura most often corresponds either to an atypical meningioma (WHO grade II) [3] or to an anaplastic meningioma (WHO grade III) [5]. The grading system in WHO 2021 is comparable to WHO 2016 with three malignancy grades (grades 1-3) based on histopathology or subtype [9]. Despite the aggressive appearance of the lesion on the CT scan and MRI in our case, the pathological anatomy revealed a benign meningioma (WHO grade I). Similar cases have already been described [10, 11]. The use of adjuvant radiation therapy is the gold standard for grade III, debated for grade II, and is not indicated for radically resected grade I meningiomas [1, 12]. The favorable evolution of our case can be explained by the fact that it was a WHO grade I completely resected, therefore no adjuvant treatment was indicated. However, psychic sequelae cannot be excluded in the long term, hence there is a need to insist on the early diagnosis of this type of lesion.

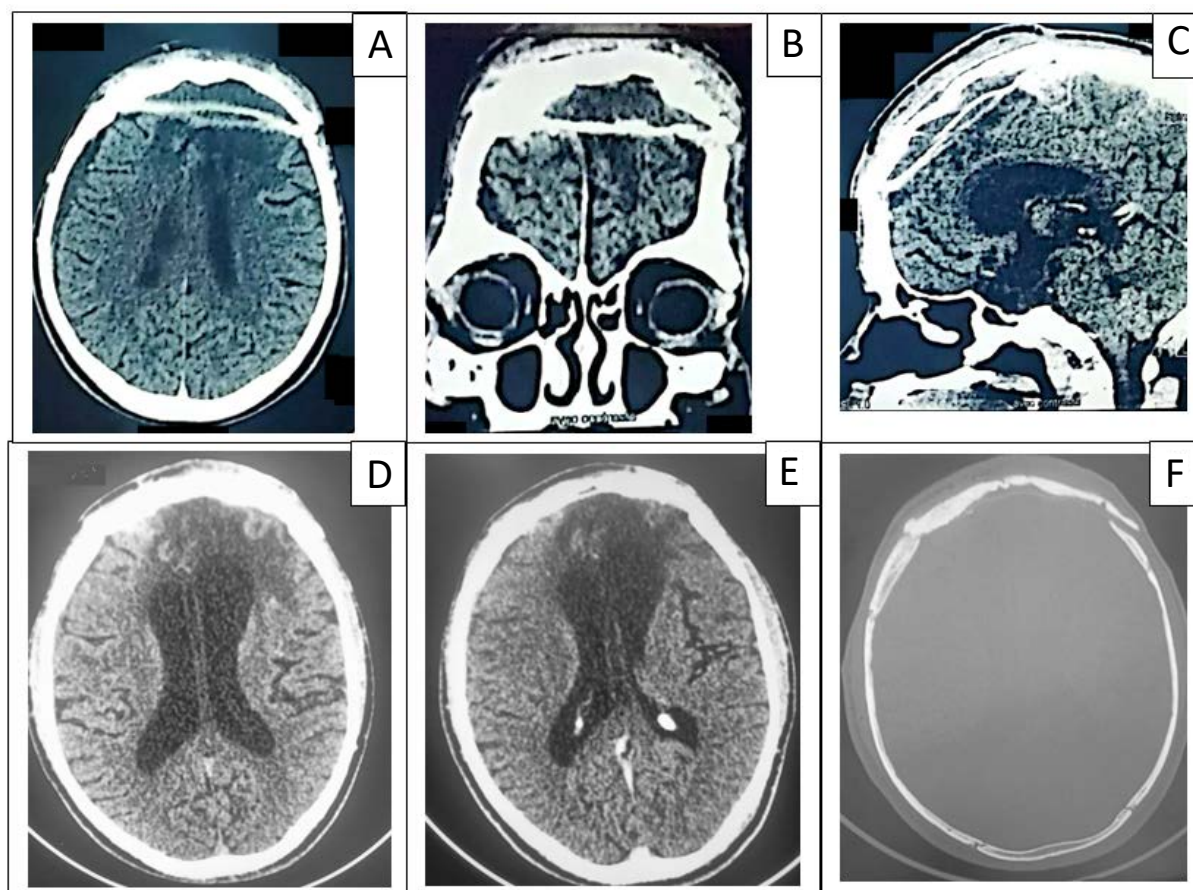


Figure 4. 4-month (A, B, C) and 16 months (D, E, F) postoperative CT scan, CT at 4 months postoperative injected in axial section (J), coronal (K) and sagittal (L) reconstruction showing an absence of tumor residue, CT scan at 16 months postoperative in axial section without (D) and with contrast (E), in bone window (F) showing an absence of tumor recurrence and the bone plasty well in place

4. Conclusion

The low socio-economic level of our population favors false beliefs and the first recourse to the traditional healer. Anything that is at the origin of the disease development will not be diagnosed without difficulty at a very advanced stage. Late diagnosis of meningioma can take on very large proportions and pose management problems while seriously affecting the patient's functional and vital prognosis. The case described in our series experienced a favorable evolution in the short and medium term but sequelae cannot be excluded in the long term, hence early diagnosis should be insisted.

Ethical Considerations

Compliance with ethical guidelines

Written informed consent was obtained from the patient for publication of this case report (including images) following the de-identification of data. The study

followed the ethical guidelines of the 1975 Declaration of Helsinki.

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Authors' contributions

Conception and design: Denlewende Sylvain Zabsonre, Data Collection: Denlewende Sylvain Zabsonre, Inoussa Zoungrana, Julie Marie Adeline Wendlamita Kyelem; Data Analysis and Interpretation: Denlewende Sylvain Zabsonre, Inoussa Zoungrana, Julie Marie Adeline Wendlamita Kyelem; Drafting the article: Denlewende Sylvain Zabsonre, Inoussa Zoungrana, Julie Marie Adeline Wendlamita Kyelem, Yacouba Haro, Souleymane Ouattara, Abdoulaye Sanou, Adama Traore, Abel Kabre; Critically revising the article: All authors; Reviewing the

submitted version of the manuscript: All authors; Improving the final version of the manuscript: All authors.

Conflict of interest

The authors declared no conflict of interest.

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