Case Report

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Meningiomatosis, the Experience of the "Marie Curie" Medical Clinic on a Case

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ABSTRACT

Meningiomatosis or multiple meningiomas are defined by the presence of at least two meningiomas. A meningioma is an extra-axial tumor that develops from the meninges more precisely at the level of the arachnoid. Its multiple characters is encountered in 1-9% of cases, with a female predominance due to hormonal factors, exposure to ionizing radiation and very rare in children. It is most often located at the supra tentorial level intra cranially. We report a case of aggressive cerebral Meningiomatosis diagnosed in the radiology department of the "Marie Curie" Medical Clinic in Bamako-Mali with the aim of describing and recalling the radiological characteristics of multiple meningiomas on Magnetic Resonance Imaging (MRI). It was a young male subject aged 10 years referred for intracranial expansive process. MRI exploration revealed multiple meningiomas (occipital convexity, bilateral temporal and jugum sphenoid). The pathology study confirmed cerebral Meningiomatosis. MRI remains the reference examination in the management of cerebral Meningiomatosis.

Keywords

Meningiomatosis, MRI, Benign tumor, Diagnosis and "Marie Curie" clinic.

Introduction

Meningiomas are classically benign extra-axial tumors, developed from meningothelial cells of the arachnoid and correspond to 13-20% of all intracranial tumors [1-3]. Multiple intracranial meningiomas or Meningiomatosis are characterized by the presence of at least two meningiomas on different intracranial sites in a patient outside the framework of neurofibromatosis [4,5]. These tumors are rare in children. Similarly, the sex ratio is twice as high in women as in men [6]. Meningiomas in children differ from those in adults in their greater aggressiveness, posing problems of care for clinicians and more common in boys than girls [1]. Despite low-grade histology, they may present with frequent recurrences and significant progression. Their appearance on magnetic resonance imaging is very evocative and poses few problems in terms of differential diagnosis [1,6]. Its contribution to the management of this tumor.

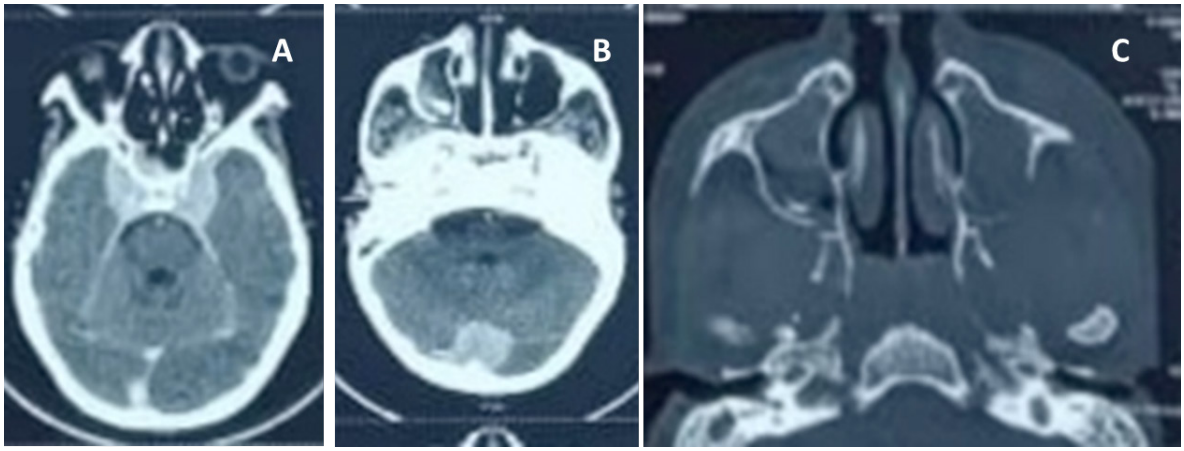

Figure 1 (A, B and C): Cerebral CT with axial reconstruction after injection of contrast product (A and B) and the bone window (B) showing multiple sphenoidal meningiomas encompassing the cavernous (A) and medial occipital (B) sinuses) associated with filling of the maxillary sinuses and lysis of the left wall (C).

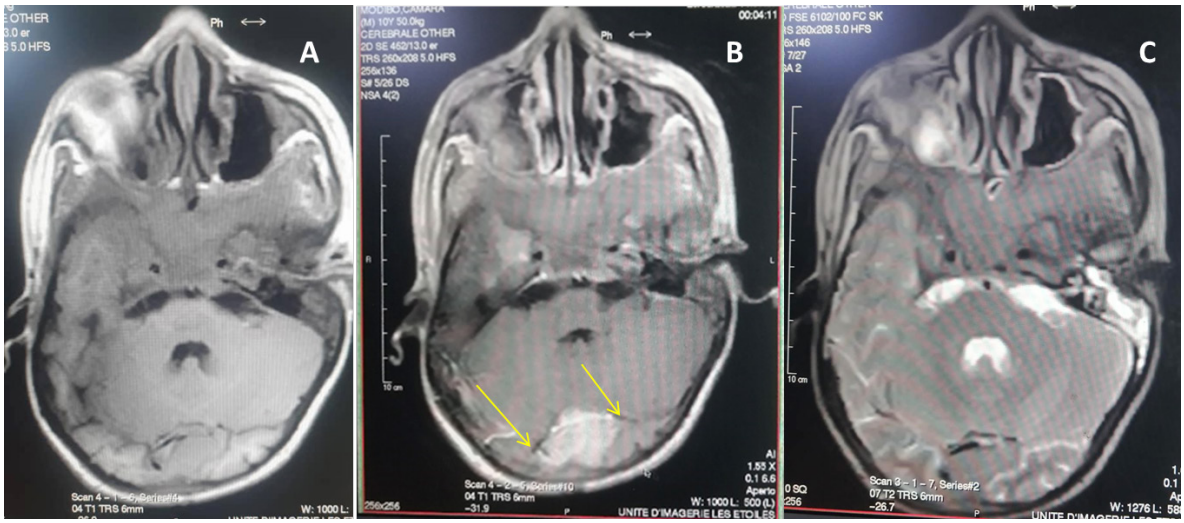

Figure 2 (A, B and C): Cerebral MRI with T1 (A), T1 after injection of gadolinium (B) and T2 sequences showing the occipital meningioma in T1 and T2 iso signal intensely and homogeneously enhanced with contrast uptake meningeal (yellow arrow).

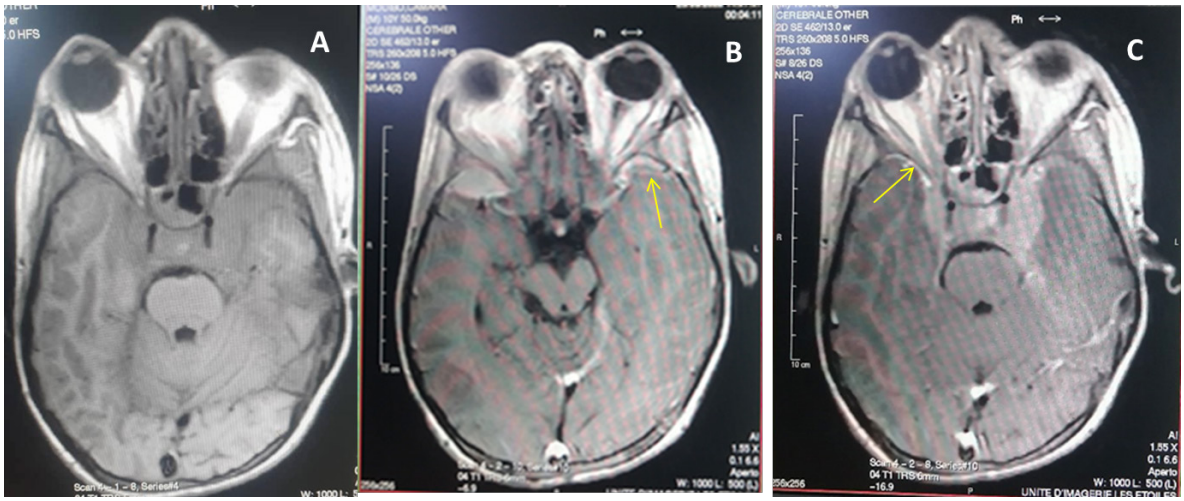

Figure 3 (A, B and C): Cerebral MRI with T1 (A) and T1 sequences after injection of gadolinium (B and C) showing the meningioma infiltrating the jugum sphenoidalis surrounding the cavernous sinuses and extending temporally bilaterally in iso signal and enhanced intensely and homogeneously with meningeal enhancement (yellow arrow).

Observation

It was a 10-year-old male child with no known medical and surgical history. The beginning of the symptomatology will go back approximately 02 months ago, marked by dental pain and swelling which motivated a consultation in the center of their locality (Koulikoro) following the aggravation of the symptoms marked by a painful palpable oral mass. Extending to the head, bucco-cerebral swelling, impossibility of mouth closure and visual disturbances found on clinical examination. The radiological examinations and anatomopathologies were requested and carried out at the medical clinic "THE STARS" of Bamako in Mali.

Cerebral computed tomography with reconstruction without and with injection of the contrast product in the bone and parenchymal window found an extra axial sphenoid isodence mass syndrome infiltrating the optic chiasmas and the cavernous compartment with almost total filling of the bilateral sphenoid-maxillary sinuses associated with a filling of ethmoidal cells and lysis of the left posterolateral wall of the left maxillary sinus observed in the bone window. Another median occipital expansive process pushing back the tent of the cerebellum which was in isodense as previously described above. These processes had rapid, intense and homogeneous contrast enhancement associated with opposite meningeal contrast enhancement (sign of Dural tail) (Figure 1).

Faced with the expansion of the tumor process, a cerebral MRI was performed at the clinic for confirmation, extension assessment and exact characterization of the tumor. We did with classic sequences (sagittal T1, sagittal T2, coronal and axial and STIR T2), sequences with Gadolinium injection (axial, sagittal and coronal T1). The diffusion and spectral sequences were not carried out given the capacity of our MRI (0.35 Tesla). This MRI found extra-axial tissue formations in T1 and T2 iso signal intensely and homogeneously enhanced after injection of gadolinium located at the median occipital level at the bilateral temporal level and at the level of the sphenoid jugum with infiltration of the optic chiasma surrounding the cavernous sinuses. It was associated with polyploid thickening of the maxillary sinuses (Figures 2 and 3).

The pathology study confirmed the grade II epithelial type meningioma. Our patient did not have time to undergo treatment; the evolution was crowned by the death of the child immediately after intracranial biopsy.

Discussion

Socio-Epidemiological Aspect

Multiple meningiomas were first described by Anfirmow and Blumenau [7], it is important to differentiate between meningiomatosis linked to neurofibromatosis and multiple meningiomas in the absence of family cases [1-3,7]. Pediatric meningiomas are extremely rare and it is difficult to know their natural history with precision, like our case, the series published in the literature being small in size and the age limit for inclusion of pediatric cases varying according to the authors. It is estimated that meningiomas occurring before the age of 18 represent only 0.4 to 4.6% of intracranial tumors in children and between 1 and 4% of all intracranial meningiomas [8]. Our work makes it possible to establish the poor prognosis of this tumor given its location, its aggressiveness and above all the young age of our patient, which was 10 years old.

Clinical Signs

The average duration of symptoms before diagnosis varies from 4.5 months to 1.6 years. Grade II-III meningiomas develop more rapidly [9]. The initial clinical presentation depends above all on the site of tumor localization; the most frequently encountered clinical manifestations are neurological deficits. In our case, we found sensory deficit (visual disturbances), meningiomas in children develop more frequently in the ventricles and can therefore easily obstruct the circulation of cerebrospinal fluid responsible for intracranial hypertension syndrome (ICH) [10]. Our patient had no subtentorial location or close to the intracerebral ventricles. The localizations were bilateral temporal, sphenoid jugum and medial occipital without any notion of HIC syndrome.

Imaging Means

The cerebral scanner with injection of contrast product makes it possible to specify the size and location of the tumor. MRI is the reference examination with injection of gadolinium to clarify the relationship of the tumor with the vessels of the brain and possibly with the cranial nerves. Multiple meningiomas present on MRI as an iso-dense tumor on T1 and more often hyperdense on T2 with enhancement after injection of gadolinium. Aurélie WEIBEL et al. states that pediatric meningiomas are frequently multi cystic (15% versus 2 to 4% in adult series), which makes their diagnosis difficult, especially in adolescents [11]. In our case, the MRI did not find any cystic component and the enhancement was intense and homogeneous after injection of gadolinium. In the literature, there is therefore a correlation in children between the presence of these images and the grade of malignancy of the tumour. Gao et al. showed that malignant meningiomas more frequently took contrast heterogeneously [12]. In our patient, the enhancement was rapid, intense and homogeneous with no sign of visible heterogeneity. The MRI examination made it possible to assess the exact extension of the tumor, confirm the diagnosis of multiple meningioma and deduce a prognosis despite its limit in terms of sequence. MRI in general remains an essential means of imaging in the management of meningiomas. In addition, the low field can still be used in countries like ours given the non-accessibility of high field MRIs in Bamako in MALI.

Treatment and Evolution

Meningiomas in children differ from those in adults in their greater aggressiveness, posing management problems for clinicians. Surgery is the first treatment implemented in all cases. Many factors can influence the quality of surgical excision, such as deep location, large tumor size, and excessive blood loss, adhesion to vital structures or extensive invasion of adjacent structures [12]. Our patient did not survive his surgery given the deep location. Radiotherapy should be less recommended in pediatric meningiomas. Our patient was unlucky for possible radiotherapy. Pediatric meningiomas are considered to have a poorer prognosis

than adult meningiomas with overall survival rates of 35% at 10 years in the series by Menon et al. [14]. Mirimanoff et al. reported survival rates of around 83% after 5 years, 77% after 10 years and 69% after 15 years [15]. Our patient died at the age of 10.

Conclusion

Meningiomatosis or multiple meningiomas remains a rare pathological entity, even very rare in children. Their deep anatomical location can be a bad factor. Magnetic resonance imaging made it possible to make the diagnosis of aggressive cerebral Meningiomatosis, to assess the extension and to give the prognosis despite its limitations. It remains the reference examination in the management of meningiomas.

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