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Malignant Meningioma: Two Cases Observed at the Hospital Center of Soavinandriana, Antananarivo

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Abstract

Introduction: Meningiomas are tumors formed by arachnoid cells, typically attached to the inner surface of the dura mater. Malignant forms are rare and no case has been reported in the Malagasy literature. The objective of our study is to report two Malagasy cases of malignant meningioma and to discuss the epidemiological and anatomical-clinical particularities of this tumor. **Observation:** The first patient, a 41-year-old woman, presented with a rapidly progressive intracranial hypertension syndrome. The patient had undergone surgery two years earlier for a grade II meningioma and had no family history of meningioma, neurofibromatosis, or personal history of brain irradiation or head trauma. Her brain scan showed a heterogeneous polylobed left parieto-occipital mass with a meningeal implantation base. The anatomopathological examination of the samples revealed a malignant meningioma. The second patient was a 33-year-old man, operated for grade I meningioma eleven months before admission, with no other personal or family history. The patient was hospitalized for tumor recurrence with signs of intracranial hypertension. The brain computed tomography (CT) scan showed a heterogeneous extra-axial tumor in right temporo-parietal lobe. Surgical excision was performed. On histological examination, a proliferation of tumor cells of meningothelial appearance with papillary architecture was observed, leading to the diagnosis of malignant meningioma. **Conclusion:** Malignant meningioma is a rare and serious entity. The clinical manifestations are non-specific and imaging may mimic a low-grade meningioma. The diagnosis of certainty is histological and is based on essentially morphological criteria. The latter condition the overall survival of the patient and the therapeutic conduct.

Keywords

Central Nervous System, Malignant Meningioma, WHO Grade III Meningioma

1. Introduction

Meningiomas are tumors formed by arachnoid cells, typically attached to the inner surface of the dura mater [1]. Malignant meningioma is a rare entity representing 1% to 3% of meningiomas [2]. The tumor is fatal with an average survival of 2 to 5 years [3]. Currently, no case has been reported in the Malagasy literature. The objective of our study is to report two Malagasy cases of malignant meningioma and to discuss the epidemiological and anatomical-clinical particularities of this tumor.

2. Observation

The first patient, a 41-year-old woman, presented with a rapidly progressive intracranial hypertension syndrome. The patient had undergone surgery two years earlier for a grade II meningioma and had no family history of meningioma, neurofibromatosis, or personal history of brain irradiation or head trauma. Her brain scan showed a heterogeneous polylobed left parieto-occipital mass with a meningeal implantation base (**Figure 1**). She was operated for a surgical removal of the tumor. On histology, the surgical specimen was found in multiple beige, firm fragments. Microscopic examination showed a tumor proliferation of diffuse architecture, in some places lobulated, formed by cells with regular oval nuclei with a small nucleolus and eosinophilic cytoplasm, sometimes with a syncytial appearance. The mitotic index was 21 mitoses/10HPF. The diagnosis was malignant meningioma. Adjuvant radiotherapy was prescribed but not carried out by the patient. The last follow-up brain scan on 14 months post-surgery showed a resumption of tumor growth with visual manifestations. She presented a resumption of tumor growth with homonymous lateral hemianopsy but she could not receive any other adjuvant treatment after surgery, lack of financial resources.

The second patient was a 33-year-old man, operated for grade I meningioma eleven months before admission, with no other personal or family history. The patient was hospitalized for tumor recurrence with signs of intracranial hypertension. The brain computed tomography (CT) scan showed a heterogeneous extra-axial tumor highly vascularized in right temporo-parietal lobe. Surgical excision was performed. Macroscopically, the surgical specimen was found in the form of multiple fragments. On histological examination, a proliferation of tumor cells of meningothelial appearance with papillary architecture and large necrosis was observed, leading to the diagnosis of malignant meningioma (**Figure 2** and **Figure 3**). The patient had succumbed to his disease, in the third week (24

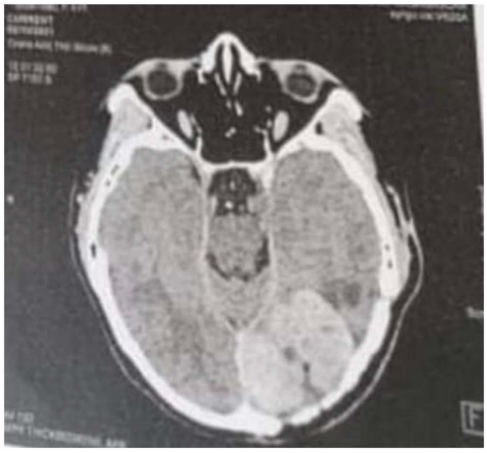

Figure 1. Brain CT scan showing a heterogeneous polylobed left parieto-occipital mass. Source: Department of neurosurgery, Soavinandriana Center Hospital (CENHOSOA), Antananarivo, Madagascar.

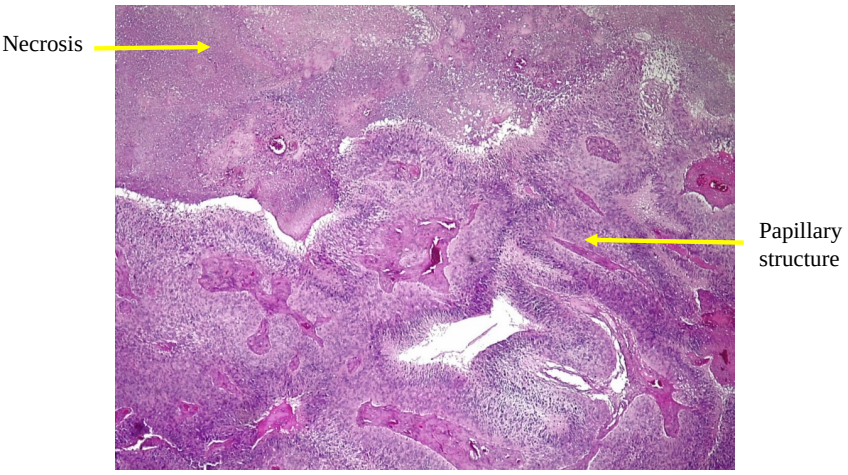

Figure 2. Papillary meningioma with large necrosis, MO, HE, ×400. Source: Department of Pathology, Soavinandriana Center Hospital (CENHOSOA), Antananarivo, Madagascar.

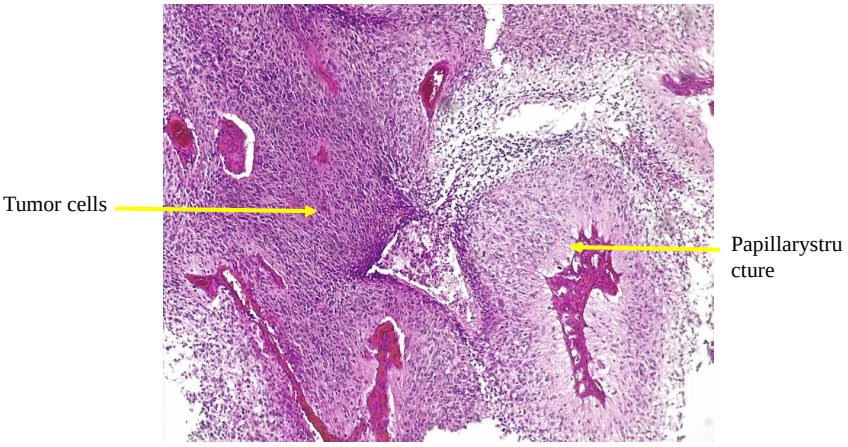

Figure 3. Papillary meningioma with papillary structures, MO, HE, ×100. Source: Department of Pathology, Soavinandriana Center Hospital (CENHOSOA), Antananarivo, Madagascar.

days) postoperative intensive care, with bleeding and infectious complications. He did not receive any other adjuvant treatment after surgery.

3. Discussion

Malignant meningioma is a rare entity representing 1% to 3% of meningiomas [2]. The tumor is fatal with an average survival of 2 to 5 years [3]. In the Malagasy literature, no case has yet been reported.

Kshettry VR and al reported a male predominance of malignant meningioma in people over 75 years of age but a female predominance of the disease was observed in those under 65 years of age [4]. Our two patients were a relatively young woman and man aged 41 and 33 years, respectively. As these were only two cases of malignant meningioma, we could not retain any gender predominance in our study.

Direct exposure to ionizing radiation [5] and alterations in the neurofibromatosis gene [6] are recognized as associated with meningioma development. Some studies suggest that endogenous and exogenous steroid hormones increase risk of meningioma [7]. According to Barnett GH and colleagues, head trauma may contribute in the development of meningiomas in some cases [8]. Alterations in the neurofibromatosis gene can also lead to the development of malignant meningioma [6]. Regarding predisposition to develop malignant meningioma, our patients had no family history of meningioma or neurofibromatosis type 2. In addition, they each had a personal history of meningioma. The woman developed a grade II tumor 2 years before the diagnosis of malignant meningioma, and the second had a grade I tumor 11 months before the diagnosis of malignant meningioma. According to Pallud J and al, half of malignant meningiomas arise from Novo and the other half are secondary to the evolution of a low-grade meningioma (grades I and II) to a high-grade meningioma (grade III) [9] Malignant transformation of a low-grade meningioma is relatively rare concerning 2% of grade I meningiomas [10] and 15% of grade II meningiomas with a median duration of malignant transformation of 5.7 years for grade I meningiomas [11] and 1.4 years for grade II meningiomas [12].

As for the clinical signs, they are not specific. They depend on the location of the tumor and are related to an increase in intracranial pressure. The latter is related to the mass effect of the tumor leading to the triad: headache, vomiting and papilledema [13] which were present in our two patients.

On imaging, malignant meningioma is irregular, heterogeneous, with no distinct interface between the tumor and the brain, necrotic, invasive, and rapidly growing [14]. It may also mimic a benign meningioma with marked edema, massive and homogeneous contrast-taking heterogeneous appearance, irregular or nodular brain surface, mushroom formation on the outer edge of the lesion, bone destruction, and absence of calcification [15]. In the cases of our patients, the first one presented a heterogeneous polylobed tumor of the left parieto-occipital lobe with a meningeal implantation base and the second one, a heterogeneous ex-

tra-axial tumor highly vascularized in the right temporo-parietal region.

Regarding histopronostic criteria, we used those redefined by the World Health Organization (WHO) in 2007 and 2016. These criteria are essentially morphological based on histological subtype, mitotic activity, cortical invasion, hypercellularity, increased nucleocytoplasmic ratio, presence of prominent nucleoli, loss of architecture or sheeting, necrosis and presence of dedifferentiated tumor foci simulating sarcoma, carcinoma or melanoma. There are three grades, namely grades I, II and III. Malignant meningioma is classified as grade III [3]. The grade is a major prognostic criterion since it conditions the overall survival and the therapeutic strategy. It should be noted that patients with grade III meningiomas have an overall survival of 32% to 64% at 5 years and their treatment is based on surgery and radiotherapy [16]. In our study, the first patient had a surgical removal of the tumor and was then prescribed radiotherapy but did not comply with it. She presented a clinical resumption of tumor growth but is alive to this day. The second patient had succumbed to his disease, in the third week postoperative intensive care.

4. Conclusion

Malignant meningioma is a rare and serious entity. The clinical manifestations are nonspecific and imaging may mimic a low-grade meningioma. The diagnosis of certainty is histological and is based essentially on morphological criteria. The latter condition the overall survival of the patient and the therapeutic conduct.

Consent from the Patients

We obtained the consent from our patient for the first case and from his family for the second case study.

Conflicts of Interest

The authors declare no conflict of interest.

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