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Case Report

Primary cerebral melanoma: An exceptional localization in a case report ☆

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ABSTRACT

Cerebral melanoma is often related to a secondary location of a cutaneous or mucosal melanoma. However, primary cerebral melanoma is a very rare clinical situation, representing less than 1% of all melanomas and 0.07% of all cerebral tumors. The diagnosis of a primary cerebral melanoma therefore requires rigorous clinical and paraclinical investigations. We report a case of primary cerebral melanoma treated in our onco-radiotherapy department.

Introduction

Melanocytic cells - the precursors of melanomas - are present in the skin, mucous membranes, uvea and central nervous system, particularly in the leptomeninges [1], which explains the topographical spectrum of melanomas. Primary cerebral melanoma is a rare entity compared to other primary brain tumors on the one hand, and secondary cerebral melanomas on the other hand. The positive diagnosis of a primary cerebral melanoma is often difficult, requiring a meticulous clinical and paraclinical investigation. The therapeutic management is based on surgical resection of the tumor followed by adjuvant radiotherapy. We report a clinical case of a primary cerebral melanoma treated at the oncology center in OUJDA, Morocco.

Case report

A 65-year-old female patient with a single kidney and no particular pathological history. She was admitted to the regional oncology center in Oujda for the treatment of primary cerebral

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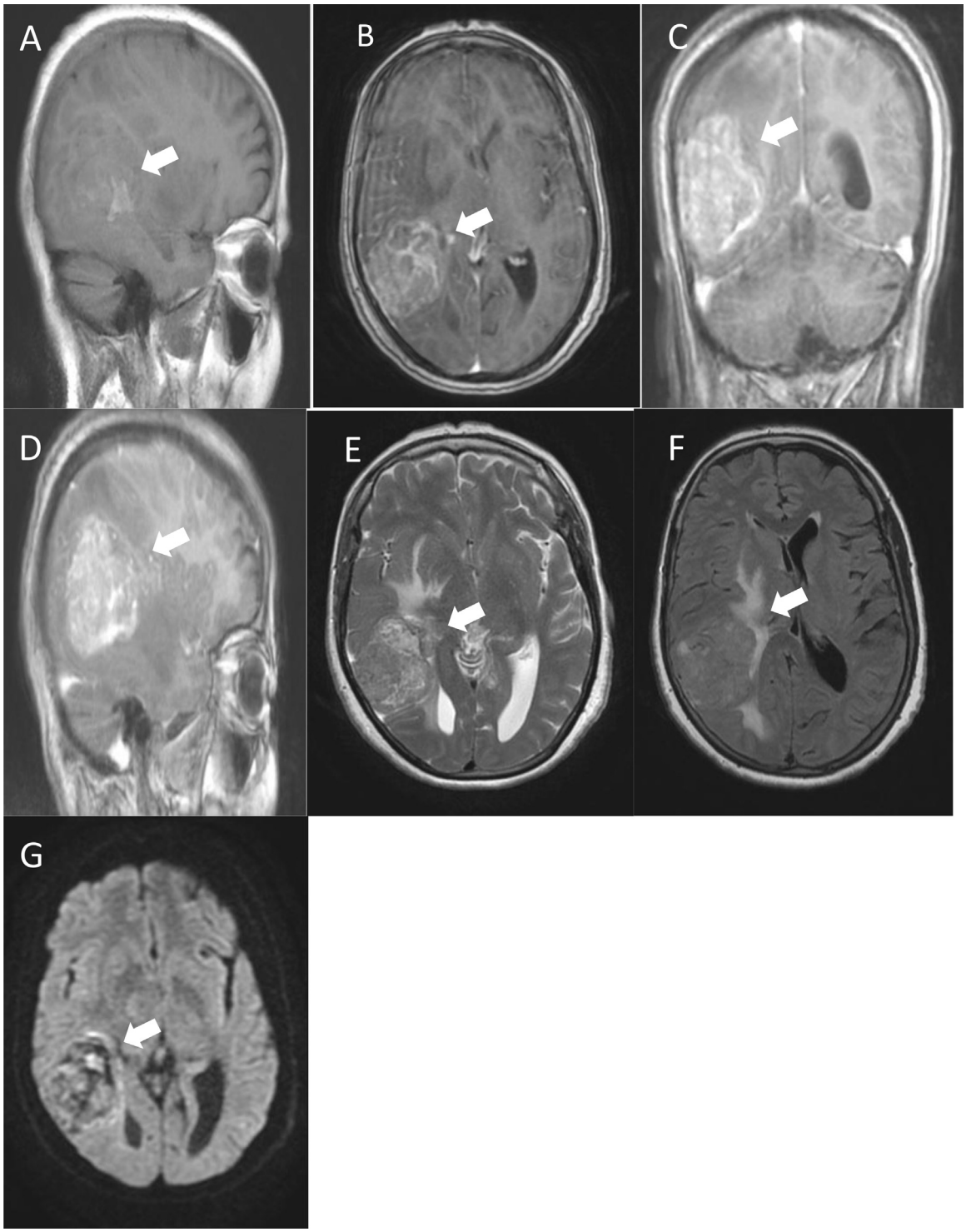

Fig. 1 – cerebral MRI images, T1sequence (sagittal section (A), injected axial section (B), injected coronal section (C) and injected sagittal section (D)), then T2 sequence axial section (E), FLAIR sequence axial section (F) and diffusion sequence axial section (G) objectifying a right parieto-temporo-occipital intra-axial process.

melanoma. The clinical symptoms dated back 12 months prior to admission, by the installation of morning headaches not relieved by analgesic treatment without neurological deficits and without deterioration of the general condition. The clinical examination found a patient in good general condition with no detectable abnormality, particularly neurological or dermatological. Cerebro-medullary MRI revealed the presence of a right intra-axial parieto-temporo-occipital tumor process measuring 56*43*46 mm, in heterogeneous isointense T1, intermediate hyperintense T2 and FLAIR, including areas

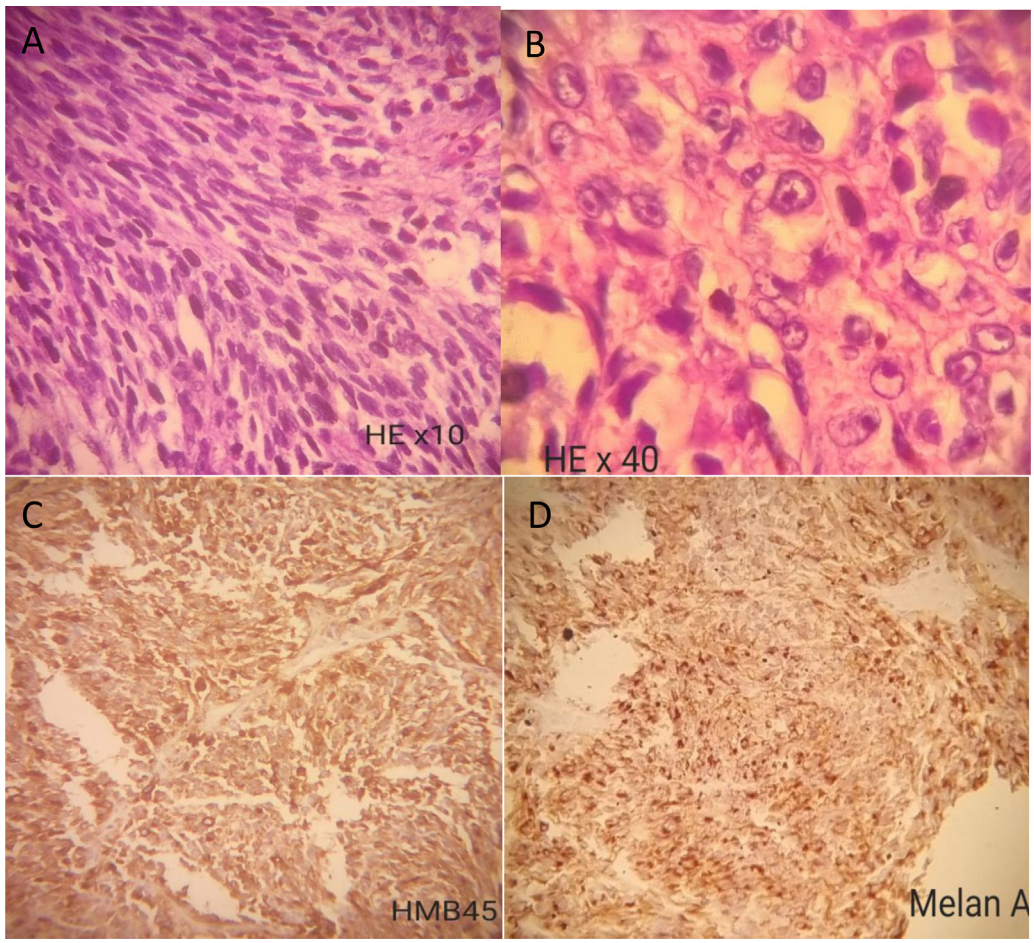

Fig. 2 – Histological and immuno-histochemical appearance of the surgical specimen. Hematoxylin and eosin staining at x10 (A) and x40 (B) magnification. Diffuse positivity of tumor cells to anti-HMB45 (C) and anti-MelanA antibodies (D).

of hyperintense T1 and T2 enhanced in a moderate and heterogeneous manner after the injection of the contrast product, in relation to hemorrhagic remodeling, this process is surrounded by a glove-like edema without spinal cord lesion (Fig. 1). Surgical resection of the tumor was performed. Histological and immuno-histochemical analysis was in favor of a melanoma, showing notable expression of the PS-100, HMB45 and Melan A antigens by the tumor cells (Fig. 2). Ophthalmological and dermatological exploration did not reveal any abnormality, likewise, 18FDG PET-CT did not show any hyper-metabolic focus suspicious for lymph node, visceral or osseous neoplastic extension (Fig. 3). The patient subsequently underwent adjuvant radiotherapy to the tumor bed at a total dose of 60 Gy in 30 fractions of 2 Gy by 3-Dimensional conformal radiotherapy. After a 3-month follow-up, the patient is alive and in good control of her disease.

Discussion

During embryogenesis, the neural crest gives rise to melanocyte cells, which migrate to the skin, mucous membranes, eyes and leptomeninges. This topographical distribution of melanocytes in the human body explains the usual locations of melanomas. However, primary cerebral melanomas are a rare clinical situation; they represent less than 0.07% of all brain tumors and form a clinical and histological entity distinct from other melanomas, particularly secondary cerebral melanomas [2]. Given that the contribution of the literature is poor, the epidemiological analysis of existing data identifies the concentration of cases of cerebral melanoma reported in Japan, the eastern United States and Europe, and a profile of younger male patients [3].

Hughes et al. distinguishes 4 types of intracranial melanoma: isolated primary cerebral melanoma, diffuse leptomeningeal melanoma, discrete medullary melanoma and meningeal melanosis [4]. Primary cerebral melanoma is often difficult to diagnose, sometimes due to the non-specific clinical and radiological presentation and sometimes due to the high frequency of secondary forms. However, Hayward described 3 conditions before opting for a primary cerebral melanoma: no malignant melanoma outside the central nervous system, the absence of lesion in another part of the central nervous system and histological confirmation of melanoma [5].

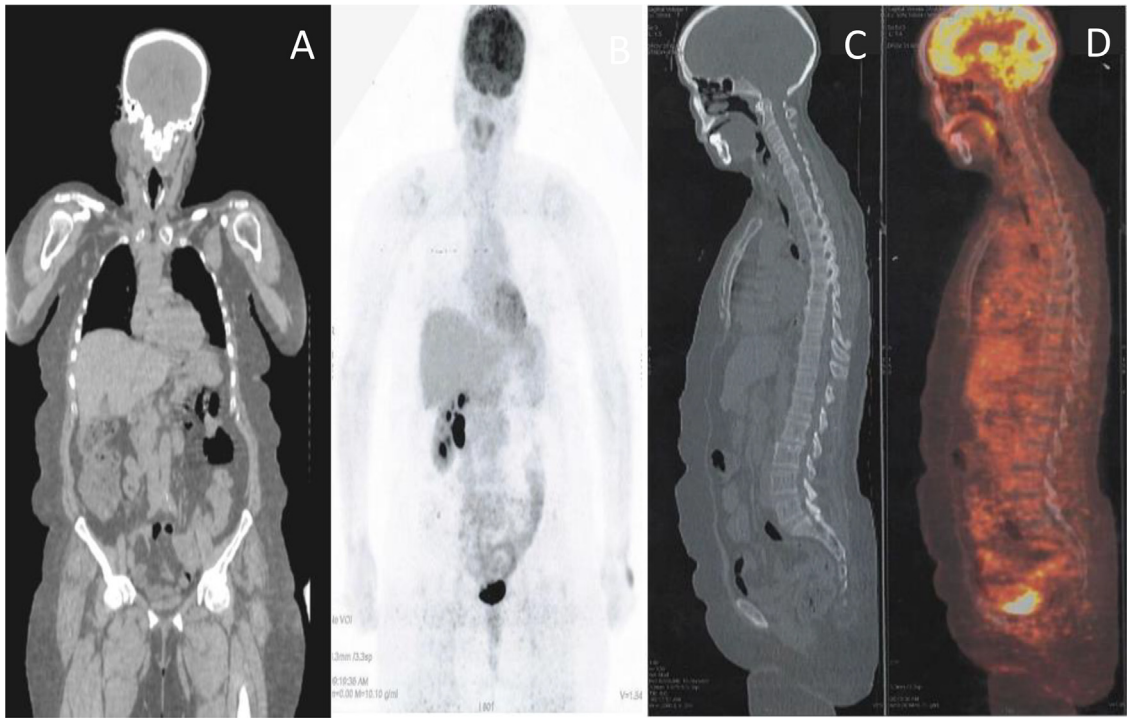

Fig. 3 – FDG-PET images, coronal (A) (B) and sagittal (C) (D) sections showing no extra-cerebral location of the disease.

Most patients present with symptoms related to intracranial hypertension, neurological deficit, or convulsive seizures. The clinical examination is often poor, making it possible to assess the neurological impact of the brain lesion and to rule out a cutaneous or mucosal location of a melanocytic tumor. The radiological presentation of primary cerebral melanomas is variable and nonspecific. The presence of intra-tumoral hemorrhage and melanin gives cerebral melanoma a hyperdense appearance on CT scan, which is accentuated after injection of contrast product with significant peri-tumoral edema [3]. Brain magnetic resonance imaging (MRI) remains the test of choice in the exploration of brain tumors. The MRI presentation of cerebral melanomas varies depending on the type of cerebral melanoma and the possible presence of intra-tumoral hemorrhage. The presence of intra-tumoral melanin leads to a T1 hyperintense and T2 hypointense MRI appearance of melanocytic tumors [6]. However, melanomas without intra-tumoral hemorrhage with less melanin expression present on MRI as T1 iso-hypointense and T2 hyperintense [7].The FDG-PET is increasingly playing an important role in excluding extra-cerebral locations of melanoma with a specificity of approximately 91% [8]. The definitive diagnosis of melanocytic brain tumors is made by histological examination and immuno-histochemical analysis of a tumor biopsy or the surgical specimen. The strong expression of HMB-45, Melan A and S-100 antigens is strongly suggestive of a melanocytic tumor.

At present, there is no standard treatment for primary cerebral melanomas given the rarity of the disease. The most complete surgical resection possible followed by adjuvant radiotherapy seems to give better results according to the available data [7]. Stereotactic radiosurgery, whether or not followed by whole-brain radiotherapy, also improves the quality of life and even survival of patients [2].

The contribution of conventional chemotherapy remains limited given the poor passage of chemotherapy drugs across the blood-brain barrier and the chemo-resistant nature of melanomas. However, immunotherapy molecules have enabled better brain concentration and good tumor response [9].

Clinical outcomes in patients with primary melanoma of the central nervous system depend on the quality of surgical resection, leptomeningeal dissemination, and tumor location [10]. These results remain better compared to those of secondary cerebral melanomas.

Conclusion

Primary cerebral melanoma is a very rare clinical entity and often difficult to diagnose. The diagnostic investigation has a dual interest: firstly, to confirm the melanocytic nature of the brain tumor and secondly to eliminate an extra-cerebral localization of the disease. Surgical resection of the tumor remains the best therapeutic option associated or not with radiotherapy and/or chemotherapy.

Patient consent

Written informed consent for the publication of this case report was obtained from the patient.

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