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Clinical case

An unusual pituitary stalk lesion: What is the place of surgery?

J. Todeschi [a], S. Chibbaro [a], J.-B. Clavier [b], B. Lhermitte [c], B. Goichot [d], F. Proust [a,*]

[a] Service de neurochirurgie, hôpital Hautepierre, hôpitaux universitaires de Strasbourg, 1, avenue Molière, 67098 Strasbourg, France
[b] Service de radiothérapie, centre de lutte contre le cancer Paul-Strauss, 67065 Strasbourg, France
[c] Service d'anatomo-pathologie, hôpital Hautepierre, 67098 Strasbourg, France
[d] Service de médecine interne, endocrinologie et nutrition, hôpital Hautepierre, 67098 Strasbourg, France

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ABSTRACT

Background. – Sellar and suprasellar primary melanocytic tumors are exceptional occurrences. Besides the difficulty of differential diagnosis between a primary and secondary lesion, treatment of these pathologies is still unclear and controversial.
Case report. – We describe the case of a 36-year-old woman with no relevant previous medical history who presented with 1 month history of diabetes insipidus, blurred vision and generalized weakness; a brain MRI disclosed an atypical pituitary stalk lesion; initially the tumor was biopsied through an endonasal endoscopic approach that revealed a melanocytic tumor; the patient was afterwards managed by a second stage extended endonasal endoscopic approach achieving a subtotal tumor removal. The overall survival was of 14 months due to the multidisciplinary management including surgery, radio and chemotherapy.
Conclusion. – If a biopsy is essential to deal with these invasive lesions, treatment including surgical resection should be part of a multidisciplinary approach.

1. Introduction

Primary brain melanocytic and/or melanoma tumors are extremely rare entities constituting less than 0.1% of all CNS tumors [1]. In the pituitary region these tumors are often metastasis. Nevertheless, some cases reported in the literature are primary tumors [2–12]. Despite the current performing level of imaging, the diagnosis of primary melanocytic tumors remains not possible because of the signal is not pathognomonic [13]. Also the biopsy remains the crucial step to plan the best management. In order to confirm the primary nature it is important to make an accurate clinical procedure based on three main features: MRI, histopathological findings with positivity of specific immunohistochemical markers, negativity of an extensive total body work up to exclude a metastatic lesion [1,14]. Prognosis remains still very poor despite any aggressive treatments. Recently the place of surgical excision has become a matter of debate and its role to be determined [7,12]. We report a case of a primary pituitary stalk melanoma extending to the sellar and suprasellar region, and also discuss the clinical decision making process along with relevant literature.

2. Case report

A 36-year-old woman with no significant previous medical history presented to the endocrinology outpatient clinic with a one-month history of progressive headache, diabetes insipidus (DI), generalized weakness and visual disturbances. Physical examination was unremarkable, but her neurological exam bi-temporal hemianopia. Complete hormone work-up showed a mild hyperprolactinemia along with hypothyroidism and hypocortisolism. Therefore, the patient was immediately supplemented by hydrocortisone and L-Thyroxine replacement therapy. Brain MRI with pituitary protocol revealed a heterogeneously enhancing lesion involving the pituitary stalk extending to the sellar and suprasellar region causing an upward chiasmatic displacement and particularly optic tract infiltration; the heterogeneous component of the lesion was interpreted as an hemorrhagic feature. No other intracranial lesions were detected (Fig. 1).

Given the atypical radiological appearance of the lesion, a biopsy was performed via an endoscopic endonasal approach; during surgery, a brown dark and highly pigmented lesion was directly visualized. Histological analysis of the specimen confirmed to be a melanocytic tumor consisting of globular cells with melanin pigment, a round nucleus and a clear and prominent nucleolus;

Abbreviations: DI, diabetes insipidus; CNS, central nervous system; MRI, magnetic resonance imaging; BBB, blood brain barrier; PET CT, Positron Emission Tomography–Computed Tomography; DS-GPA score, diagnosis-specific Graded Prognostic Assessment; RPA score, Recursive partitioning analysis.

* Corresponding author.
E-mail address: f.proust@neurochirurgie.fr (F. Proust).

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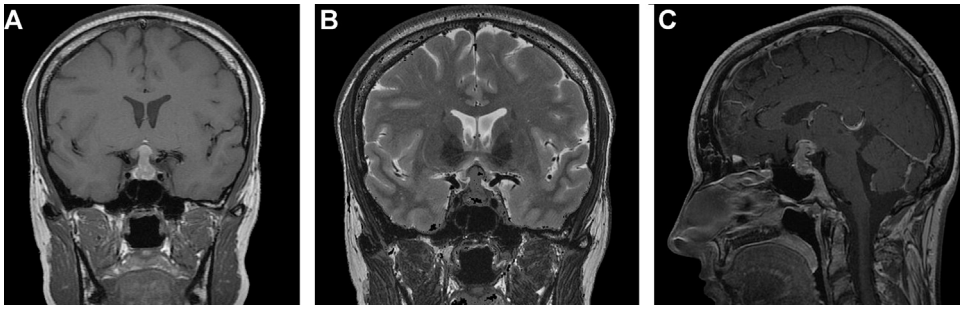

Fig. 1. A–C. Coronal T1 (A), coronal T2 (B) MRI and Sagittal T1 enhanced (C) weighted image showing a heterogeneous lesion with mainly hyper-intensity areas involving the pituitary stalk and extending to the sellar and suprasellar region, causing chiasmatic compression and optic tractus infiltration.

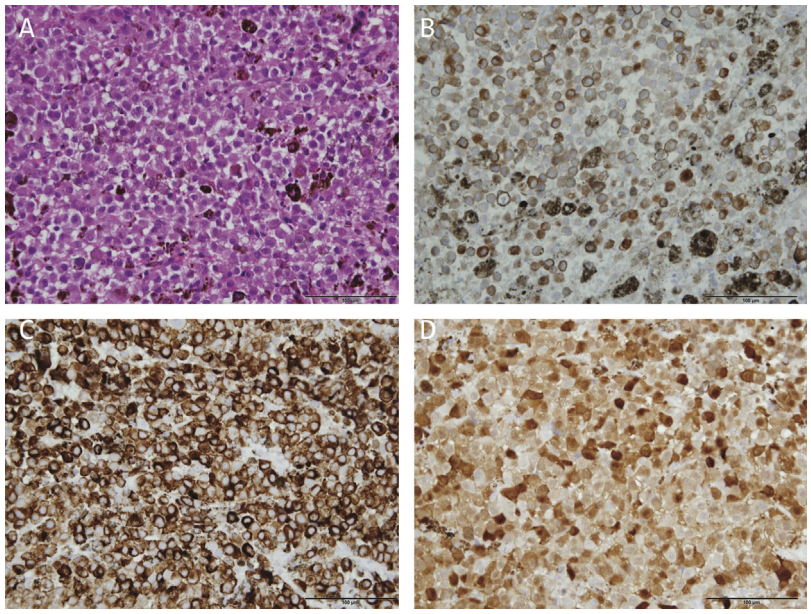

Fig. 2. A–D. Histologic photomicrographs (400/magnification) of surgical specimen. Standard staining with hematoxylin - eosin (A) and immunostaining with antibody Melan A (B), HMB-45 (C), protein S100 (D). The HMB-45 and Melan-A staining produce a cytoplasmic granular marking, and PS100 of both the nucleus and the cytoplasm.

the rapid tumor proliferation explains necrotic foci and immunolabeling confirmed the melanocytic nature of the tumor (Fig. 2). The proliferation index "Ki67" (clone SP6) was very high, estimated at 40%. Molecular biology study disclosed the BRAF V600 mutation.

Furthermore, in order to exclude a primary cancer, an extensive total body work-up was performed including: full ophthalmology examination to rule out eventual ocular localization as well as dermatological examination and digestive endoscopy; a total body 18 FDG PET scan along with a very thin slice CT-scan was performed. Considering that all complementary investigation did not show any other and/distant lesion it was sensible to conclude for a primary sellar lesion.

A MRI performed 2 weeks later revealed a significant tumor volume increase with additional upward extension to the third ventricle (Fig. 3); despite the tumor aggressive nature with a rapid growth mass, surgical resection by extended endonasal endoscopic route was performed. Perioperative observation suggested an extensive infiltrating, adherent and very bloody lesion. The awakening was uneventful although the patient, a few weeks later, developed a hypothalamic syndrome with sleeping problems and bulimia. An early postoperative brain MRI did not show either a ventricular dissemination (being also the 3rd ventricle clear) or hydrocephalus. Finally, a spinal MRI did not show any drop metastasis. Fractionated radiotherapy was performed within 4 weeks. A total dose of 30 Gy was given to the patient. Regular and frequent follow-ups were carried out; 6 months after surgery the patient started to be confused and disoriented in time and space. Lumbar puncture was performed and confirmed the suspicion of carcinomatous meningitis with diffuse ependymal metastatic spread. Due to a positive BRAF V600 mutation, chemotherapy with Vemurafenib (Zelboraf®) was started. Two months after this treatment, the patient's neurological examination was essentially unchanged although repeated spinal taps were unremarkable with no tumoral cells. At 12 months follow-up, MRI showed significant disease progression and a new onset of obstructive hydrocephalus; at this stage Temozolomide was introduced as second line chemotherapy being unfortunately not very effectively administered, probably due to the large tumoral spread; the patient was then treated conservatively and ultimately only palliative care was provided and she passed away 14 months after surgery.

3. Discussion

The present report concerns a young lady, who was harboring a rare primary pituitary stalk melanoma, who presented with a poor prognosis of 14 months survival despite a microsurgical resection coupled with chemo- and radiotherapy. Primary melanocytic neoplasms of the CNS are rare lesions arising from melanocytes of the leptomeninges [1]. They include diffuse leptomeningeal melanocytosis or melanomatosis, melanocytoma and primary malignant