Melanoma of the Ciliary Body of the Young Subject: A Case Report

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Abstract

Uveal melanomas are the most common primary intraocular. We report a case of melanoma of the ciliary body from the left eye of a 21 y old patient. Ophthalmological examination of the left eye revealed the presence of a poorly pigmented tumor that presses back the lower temporal equatorial lens. The bottom of the eye is uninterpretable. The ultrasound measurements of the tumor were 3.5 mm thick over 10 mm in diameter. The diagnosis of ciliary body melanoma was posed. However, the patient has atypical karyotypic abnormalities that might suggest an ocular metastasis of another unknown primitive tumor.

Keywords: Melanoma; Ciliary body; Atypical karyotypic abnormalities; Young subject

Introduction

Uveal melanoma is the most frequent type of intraocular primary tumor. It mostly affects people of the Caucasian race and is rare among black and Asian individuals [1]. Contemporary metastases at diagnosis of the primary tumor are found in 2% of cases, and they affect the liver in the vast majority of cases. We report the case of a young Algerian girl with ciliary body melanoma with contemporary metastases at diagnosis, but with no liver metastases.

Observation

A 21 y old girl was addressed for suspected malignant melanoma of the ciliary body of the left eye, lasting for six months. Visual acuity was 10/10 in the right eye, and there was no light perception in the left eye. Intraocular pressure was 10 mmHg in the right eye and 41 mmHg in the left eye. The ophthalmological examination was normal for the right eye. However, the left eye showed predominantly lower temporal episcleral vasodilation, a neovascularization over the entire iris area, a total white cataract, an iridocorneal angle closed over 360º, and the presence of a lightly pigmented tumor repressing the lower temporal lens equator. The ocular fundus was uninterpretable. Ultrasound measurements revealed the tumor was 3.5 mm thick and over 10 mm in diameter.

Before this picture, the diagnosis was made of malignant melanoma of the ciliary body, and the only therapeutic indication was enucleation because of the blindness and pain associated with ocular hypertension. Enucleation was performed the day after admission. An anatomopathological examination performed on the work piece enucleation confirmed the presence of malignant melanoma of the 10 × 10 mm whitish ciliary body. We also observed a proliferation of epithelioid cells little joined together, and the mitotic index was low. The tumor nibbled the sclera without embolus, and the anterior chamber angle was closed with a synchia without invasion of the iris.

The papilla was not overrun. Immunohistochemical examination revealed mutations of BRAF genes on exon 15 (Figure 1), with exon 11 being normal (Figure 2), but there were no mutations for NRAS (Figure 3), CKit (Figure 4), or PDGFRA. The staging was essentially the head-thorax-pelvis-abdominal scan. This scan highlighted the cerebral stage, the presence of two right and left frontal lesions of progressive appearance (Figures 5 and 6), the thoracic level, the presence of a nodular lesion of 7 mm non-specific right fowler (Figure 7), a right retractable apical infiltrate consistent with intercurrent infection site, and a plural bilateral effusion in histopathological examination, which did not show any histological signs of malignancy. In the subdiaphragmatic stage, the liver was of normal size and appeared homogeneous without a focused, suspicious lesion (Figure 8). However, we found large mesenteric, necrotic masses, which developed in the pelvic area accompanied by a diffuse intraperitoneal effusion. Ultrasound-guided biopsy of the mesenteric masses confirmed the metastic nature of these masses.

Figure 1: Mutations of BRAF genes on exon 15.
As compared with this picture of malignant melanoma of the ciliary body with multi-organ metastases, chemotherapy was indicated in the absence of other therapeutic methods with administration of Muphoran (Fotemustine). The evolution was marked by the rapid death of the patient two and half months later. The genomic analysis performed several months after enucleation from DNA extracted from the uveal melanoma tissue helped to highlight many anomalies, including a mosaic gain of chromosome 8, very numerous copy number variants (CNV) in loss and gain of chromosome 9, a mosaic loss of chromosome 10, and CNV on chromosomes 5, 7, 12, and 22. There was no CNV on chromosomes 3 and 6. Therefore, it is an atypical profile.
Discussion

Choroidal melanoma is the primary eye malignancy most common in adult men. Its usual evolution is slow and its metastases usually reside in the liver in over 92% of cases. Metastases can rarely be found in the lung, bone, at skin and subcutaneous level, lymph node, brain, or adrenal sites.

The positive diagnosis of ciliary body melanoma is based on clinical examination and imaging, and it is confirmed by pathological examination of the tumor either by biopsy or after enucleation. The tumor is revealed by a decrease in vision, but also through ocular pain associated with ocular hypertension [2] due to the invasion of the anterior chamber angle and lens opacification. This description was found in our observation with blindness discovered through the examination associated with eye pain and hypertension.

In our observation, the patient had no liver metastases during staging, in contrast to literature data in which the development of choroidal melanoma and a ciliary body is marked mainly by the occurrence of liver metastases with a time of extremely variable latency [3]. This absence of liver metastases is an exception and an absence of liver metastases and atypical genotypic profile of our patient make this observation exceptional. Faced with this malignant melanoma of the metastatic ciliary body, without liver metastases with BRAF gene mutations and atypical karyotypic abnormalities, we might consider ocular metastasis from another primary tumor that could be an unknown involutive cutaneous melanoma.

Conclusion

This observation reveals once again the seriousness of uveal melanoma and, in particular, that of the ciliary body in young individuals. The absence of liver metastases and atypical genotypic profile of our patient make this observation exceptional. Faced with this malignant melanoma of the metastatic ciliary body, without liver metastases with BRAF gene mutations and atypical karyotypic abnormalities, we might consider ocular metastasis from another primary tumor that could be an unknown involutive cutaneous melanoma.

References


Figure 8: Coronal section of the abdomen showing the homogeneous liver and supra-mesocolic mesenteric masses, including the large vessels of the abdomen, namely the inferior vena cava and the abdominal aorta.