



**CHOROIDAL MELANOMA IN PREGNANCY: A DELAYED ENUCLEATION**

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**ABSTRACT**

**Purpose:** Through this paper we report a case of choroidal melanoma (CM) diagnosed during pregnancy in which the treatment was postponed until after delivery on the demand of the patient and review the literature about this association.

**Case report:** A patient aged of 28 year-old, presented for CM while 26 weeks-pregnant. Eye enucleation postponed on patient demand until delivery. Pathology examination Confirmed CM and showed extra-ocular involvement. Chemotherapy was then Indicated.

**Discussion:** We discuss epidemiological, physiopathology, treatment and prognosis of CM when associated with pregnancy.

**Conclusion:** Delaying treatment of CM in pregnant woman is only advisable in cases where conservative surgical treatment is an option or when the patient demands it. Otherwise; treatment should be carried out as promptly as possible.

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**INTRODUCTION**

Choroidal melanoma (CM) is a relatively rare disease , and is even rarer in the pregnant woman. it poses many challenges concerning physiopathology and treatment.

Through this paper we report a case of CM diagnosed during pregnancy in which the treatment was postponed until after delivery on the demand of the patient. We emphasis the clinical findings, ancillary tests results, treatment decisions, pathology results, and survival until last examination.

we then review the literature to discuss epidemiology, physiopathology and treatment of this association.

**Case Report**

Our patient is 28 year-old, she was in her 26 weeks-pregnancy. She presented for ophthalmic complaints made of blurred vision with temporal field scotoma. Her past medical history was unremarkable, her ophthalmic examination of the left eye found a decreased vision (3/10), the anterior segment was normal, and on eye fundus examination we found a brownish mass of the posterior segment, elevated without subretinal exudation or hemorrhage. The fellow eye examination was unremarkable. Her general physical examination didn't find evidence of systemic involvement. We performed ultrasonography in the left eye and found a mass of the posterior segment with base implantation measuring 12mm and height measuring 8 mm with medium internal reflection suggestive of choroidal melanoma (Figure: 1).

We performed magnetic resonance imaging (MRI) of the eye and orbit which showed a mass of the posterior segment with no involvement of the sclera ,the orbit, or the optic nerve (Figure: 2) . Systemic work up didn't found evidence of systemic involvement; mainly there was no sign of liver, pulmonary or bone metastasis. All these findings were suggestive of CM. We discussed treatment options with the patient, and proposed proton beam treatment as conservative treatment or enucleation as a radical treatment explaining pros and cons of both alternatives. She opted for enucleation , but wanted to postpone any treatment until delivery against our recommendation to treat the tumor as promptly as possible since we don't know the exact dynamics of metastasis. The patient was monitored for 2 months and the size of tumor remained unchanged. Delivery occurred 2 months after initial diagnosis, the patient gave birth to a healthy baby, and enucleation was scheduled 1 week later. While performing enucleation , and upon macroscopic examination of the enucleated eye we found scleral invasion by the tumor . The diagnosis of CM was confirmed by pathology examination and classed as epithelioid type with scleral effraction (figure: 3).The patient was put on systemic chemotherapy with close follow up. The patient was monitored and remained metastasis free up to a last follow up of 18 months.

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Figure 1 ocular ultrasonography showing posterior segment process with medium internal reflectivity.



Figure 2 MRI of the orbit showing an intraocular process with no extraocular structures or optic nerve involvement.



Figure 3 enucleated eye with scleral effraction by choroidal melanoma

## DISCUSSION

CM is a rare condition in 30 year-old patients or younger [1] ; and it is even rarer in the setting of pregnancy. Out of 3706 patients diagnosed with CM: 0.4 % were pregnant women in a series by Shield et al.[2]. This is far different from skin melanoma which is more frequently reported during pregnancy[1].

interestingly 7 out of 16 pregnant women with CM were known to have choroidal nevus before pregnancy in a series by Shields et al[2]. This is probably linked to melanocyte stimulating hormone (MSH) high levels during pregnancy[1] . A formal relationship between MSH and other pregnancy hormones (e.g. progesterone and estrogen ) with CM has never been formally established[3][1]. Indeed research of oestrogen receptors in CM specimen failed to show evidence of this relationship [3].

However in a cross-sectional population based study including 2505 american women aged 40 years or older ; the authors found that the earlier the pregnancy , the more there is a risk of choroidal nevus[4]. Many other theories try to explain pathogenesis of melanoma during pregnancy. There is the immunological theory which advocates that immunity is altered to favour type 2 helper cell pathway over type 1 helper cell pathway which could promote rapid tumour growth[5]. There is also the vascular approach in which fetal-induced angiogenesis with increased tumor vascularity may be implicated [5][6].

And finally the foetal cell microchimerism theory in which foetal progenitor cells are supposed to enter the maternal circulation, engraft in maternal cutaneous melanoma and differentiate into endothelial cells[5]. Many authors reports a rapid growth of CM owing to these various possible mechanisms [5].

Clinical signs made of blurred vision , ocular pain or secondary glaucoma and exophthalmos in advanced cases coupled with ancillary tests (ocular ultrasonography and MRI) allow 99% of diagnosis ; but since the decision could be of huge impact, extra care must be practiced in atypical presentations ; as reported by fabian et al[7]; a case of scleral cyst in a pregnant woman raised a difficult differential diagnosis with CM.

In a series by shields et al. [2] ; 16 pregnant women with CM were treated by enucleation in 10 cases, and by brachytherapy in 4 and 2 were observed as doubtful cases. The survival rate is comparable to non pregnant woman with CM for most authors [2][6][8]. Conservative treatments frequently used are photocoagulation that can be safely performed during first months of pregnancy and brachytherapy toward the end of pregnancy [9]. However the most widely practiced treatment is proton beam therapy; and when the patient is in her 8th month of pregnancy , labor activation is an option to administer this treatment and reduce teratogenicity [8]. In more advanced cases enucleation is the most practiced surgery. For some surgeons delaying of treatment increases the risk of metastasis [8] ; while for others in cases of large tumors, tumors involving the ciliary body and epithelioid cell type CM ; the delaying of does not influence prognosis since any metastatic spread would have commenced at an earlier stage [10]. However this delaying of treatment could be justified in two cases: when surgical resection of the tumor under general anesthesia with hypotension to avoid enucleation is an option and when the patient asks for postponing the surgery until after delivery.

Placental metastasis is possible, hence pathologic examination of placenta after delivery is mandatory[11]; unfortunately we didn't perform this examination in our patient because of lack of communication with the obstetrician team.

## CONCLUSION

Many questions still unanswered regarding relationships between pregnancy and development of choroidal melanoma. The main concern in treating choroidal melanoma is avoiding teratogenicity when considering conservative treatment by radiotherapy. while for radical treatment (enucleation), it should performed as promptly as possible to avoid extra-ocular involvement except when the patient asks for postponing surgery.

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