EUROPEAN OPHTHALMIC PATHOLOGY SOCIETY (EOPS) MEETING 2022

Date of meeting: May 25th – 28th
Place of meeting: Valencia, Spain
Name: Elena Arias García
FISABIO Oftalmología Médica
Valencia
elena.arias.gar@gmail.com
Case number: 3076
Material distributed:2 glass slides

Title:

Iridociliary melanoma in a 55-year-old woman after excisional surgery of a tumor-like lesion on her right eye iris.

Clinical history:

A 55-year-old woman was referred to our center with the diagnosis of a tumor-like lesion in her right eye iris.

Under slit lamp inspection, a 2x3.3 mm lesion was observed in the periphery of the iris, from IX to X hours, with intense vascularization and ectropion uveae sign. Gonioscopy revealed compromise of the anterior chamber angle and ultrasound biomicroscopy revealed compromise of the ciliary body. The posterior pole was normal and the endothelial cell count was in range.

The patient underwent excisional biopsy and the piece was sent for histological examination. An extended study was made that was negative for metastasis.

As the histological results showed borderline margins, another surgery was indicated that finally resulted in non-affected margins. The iris defect was now quite large, so artificial iris prothesis implantation was performed.

Ophthalmic Pathology:

Macroscopy: Iris fragment of 0.6 x 0.4 cm, black colour, irregular surface, irregular margins.

Light microscopy:

Histological slides show malign melanocytic proliferation configuring thecas and nidus of variable sizes. They contain cells with wide cytoplasm and prominent nucleoli. There isn´t significant mytotic activity. The lesion shows heterogeneous pigmentation although there is an important proportion of non-neoplasic pigmented cells (melanophages). There is infiltrative growth that affects some ciliary muscle fascicles and that extends towards the iris until approximately half its length.

Diagnosis:

Malign melanoma affecting iris and ciliary body after excisional biopsy of a macroscopic iris tumor-like lesion.

Comment:

Melanoma of the uveal tract is a cancer that arises from neuroectodermal melanocytes of the iris, ciliary body, or choroid. It is the most common intraocular malignant tumor in Caucasians, with a high potential for hematogenous dissemination[1].

Uveal melanomas may arise in the iris, ciliary body or choroid. Choroidal melanomas are the most common as they account for the 80% of all uveal melanomas, followed in frequency by the ciliary body melanomas that comprise around 12% of all uveal melanomas and the iris melanomas, being the less common, only the 8% of the total[2,3].

Even if the risk factors are common for the three localizations, the prognosis is very different, being the choroidal melanoma the most dangerous with a mortality up to 50% at 10 years. In contrast to this, only about 5% of patients with iris melanomas develop metastasis within 10 years of treatment. The case of the ciliary body melanoma is special because the diagnosis may be delayed as the lesion is easily missed[3]. This way, an anterior uveal melanoma confined to the ciliary body is not generally evident on slit-lamp biomicroscopy of the anterior ocular segment. Because of its hidden location behind the iris, the typical ciliary body melanoma is usually thicker when it is first detected[1]. On the contrary, because the iris is visible to the patient, iris melanomas tend to be detected at relatively small sizes and in patients who are significantly younger. For this reason, and because they are more surgically accessible, they are more likely to be treatable by local excision[2].

The diagnosis of iridociliary-pigmented tumor is primarily clinical. Malignancy is suggested by the increase in tumor size, vascularization, and the effect on adjacent ocular tissues. A firm diagnosis requires a more or less specific series of laboratory investigations. In practice, only histopathology offers a positive diagnosis[4]. Histopathology reveals spindle and epithelioid cell types; the former being arranged in bundles and having a better prognosis[3].

Management options for anterior uveal melanomas include surgical excision (iridectomy, iridocyclectomy, or cyclectomy—usually reserved for cohesive-appearing tumors), plaque radiotherapy and enucleation[1]. If the treatment evolves surgical excision, there will be a tissue defect in the anterior segment. Various conservative treatments are available for iris deficiencies, including colored contact lenses, sunglasses, corneal tattooing, and basic iris prostheses and intraocular lenses (IOLs) with opaque borders. The reconstruction of a pupil using iris sutures remains limited to circum- scribed iris defects of approximately 2 clock hours. A relatively new option for treatment is the implantation of a custom-made artificial iris. The highly versatile implant is made of silicone and can be implanted in various ways, using a variety of techniques, even independent of IOL implantation[5]. In our case, the iris defect was reconstructed using a silicone prothesis with very good esthetic and functional outcome.

We want to emphasize about the importance of the suspect of the ciliary body melanomas, as its diagnosis is often delayed due to its hidden position behind the iris. There are some indirect signs that suggest this diagnosis, the most important one being the sentinel scleral vessel[3]. As they can appear simultaneously with an iris

melanoma that is usually more evident, it is important to explore the ciliary body and see if it is affected. If we are performing a surgery of an iris melanoma suspect, it could be wise to take a sample of the ciliary body adjacent to the iris lesion so the histopathology will determine if we were dismissing the diagnosis of this usually hidden lesions.

References:

1. Yanoff M, Duker JS. Ophthalmology, 4th ed., 2014. Elsevier Saunders.
2. McLean IW, Saraiva VS, Burnier MN Jr. Pathological and prognostic features of uveal melanomas. Can J Ophthalmol. 2004; 39:343–350.
3. Kanski Salmon J. Kanski´s clinical ophthalmology, 9ª ed. Elsevier.
4. Shields JA, Shields CL. Intraocular Tumors: a text and Atlas. 1992, Philadelphia: WB Saunders.
5. Mayer C, Tandogan T, Hoffmann AE, Khoramnia R. Artificial iris implantation in various iris defects and lens conditions. J Cataract Refract Surg. 2017;43(6):724-731