Short communication

Iris mammillations. Three case reports∗

B. Sánchez Marugán, M. Acebes García, J. García Hinojosa,
M.J. León Cabello, M. Casal Valiño

Servicio de Oftalmología, Hospital Universitario Infanta Leonor, Madrid, Spain

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ABSTRACT

Clinical cases: We report the cases of 3 young patients who were seen in our hospital with the diagnosis of iris mammillations. Two of them were bilateral without familial association. The third child had iris mammillations as part of an ocular melanocytosis.

None of them had abnormalities in the posterior segment or the pachymetry.

Discussion: The discovery of iris mammillations during an examination requires a long-term follow-up of these patients due to its association with uveal melanoma. This is required whether it is a casual discovery or in the presence of ocular melanocytosis.

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Mamelones de iris. A propósito de tres casos

RESUMEN

Casos clínicos: Se presentan tres pacientes jóvenes vistos en nuestro servicio con diagnóstico de mamelones de iris. Dos de los casos eran bilaterales, sin asociación familiar. El tercero presentó mamelones de iris en el contexto de una melanocitosis ocular. Ninguno mostró asociación con anomalías en el polo posterior o en la paquimetría.

Discusión: La presencia de mamelones de iris en una exploración, tanto como hallazgo casual como en el contexto de una melanocitosis ocular, requiere del seguimiento a largo plazo de estos pacientes por su posible asociación con el melanoma de úvea.

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Introduction

Mammillations are congenital anomalies of the iris characterized by dome-shaped elevations of the iris, regularly distributed over a part or the entire surface of a character-istically velvety iris (without surface crypts).

Iris mammillations appear isolated or associated with ocular or dermal-ocular (Nevus of Otta [sic: Ota]) melanocytosis, in such cases with increased risk of uveal melanoma.


∗ Corresponding author.
E-mail address: beasmarugan@hotmail.com (B. Sánchez Marugán).

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Therefore, it is important to know these lesions for long-term patient follow-up, preventing uncontrolled development of this severe ocular complication. This is a report of three cases of children who were seen in our department and had iris mammillations during eye examination.

**Case 1**

5-year-old male patient, Caucasian, seen for pigmentation in the sclera of his left eye, from neonatal period. On examination the patient showed 20/25 uncorrected visual acuity in both eyes. Previous biomicroscopy shows scleral pigmentation and iris hyperpigmentation with elevated pigmented nodules and regularly distributed in the left eye (iris mamelons) (Fig. 1).

Fundus examination showed no changes. Intraocular pressure could not be measured due to lack of cooperation of the patient; however, it was possible to measure corneal thickness; results were 564 μ in the right eye and 567 on the left.

**Case 2**

8-year-old Hispanic female patient who is seen for reduced sight. Examination reveals: AV: right eye (−2.00; 180°): 1 and left eye (−1.00, −4.00 to 5°): 0.3. Biomicroscopy shows bilateral iris mammillations diffusely distributed (Fig. 2). Fundus examination was within normal limits. Pachymetry was around 540–545 μ, in the right and left eye respectively.

**Case 3**

16-year-old female patient, white race, who comes in for headaches. Examination reveals: AV: right eye (+1.75; −0.50 to 170°): 1 and left eye (+1.75; −0.75 to 5°): 1. The previous biomicroscopy has bilateral mammillations of the iris (Fig. 3). Fundus examination is normal. Pachymetry values for her right eye were 550, and 540 for the left one.

**Discussion**

Iris mammillations are raised nipple-shaped protrusions that fully or partially cover the surface of the iris. The term was coined by Coats in 1912 to describe the iris of a subject with ocular melanosis. They are characterized by being raised lesions, soft on their surface and variously dome-shaped, conical or stellate at their base, regularly covering the surface of all or part of the iris. When the surface of the iris is fully covered with mammillations, these become larger as they approach the pupillary margin. The iris where they are located is usually more pigmented, appearing as heterochromia in subjects with unilateral mammillations.

Congenital and sporadic lesions have been reported in family cases with autosomal dominant or recessive, polygenic and multifactorial heredity.

Most of the cases reported in literature are unilateral, although there are also bilateral cases.

Iris mammillations appear more frequently in subjects with ocular melanocytosis (hyperpigmentation of uveal tissue, iris, sclera, etc.) but can also occur without other coexisting ocular or dermal pigmentary conditions. It has been suggested that iris mammillations could be either a low expression of ocular melanocytosis or an isolated disorder. If we consider them as a variant of ocular melanocytosis, then we can also consider them as premalignant lesions; however, in most reported cases associated with an intraocular tumor, a preexisting melanocytosis is the most important etiological factor.

Differential diagnosis includes Lisch nodules (neurofibromatosis type I), tapioca melanoma, granulomatous inflammation of the iris and the Cogan-Reese syndrome. Lisch nodules have a cushiony appearance, are unevenly distributed and have a color lighter than the iris surface. They tend to be larger than mammillations and are more variable in size and number. They look brown in blue or green iris, and paler than underlying iris in brown eyes. By contrast, iris mammillations are the same color of the underlying iris in brown eyes and are a medium brown on blue or green eyes. Tapioca melanoma is a pale or amelanotic tumor. It grows slowly and is typical in young patients. Cogan-Reese syndrome is characterized
by diffuse nodules, peripheral anterior synechiae, iris atrophy and dyscoria. Granulomatous inflammatory nodules occur in uveitis as Koepppe and Busacca nodules.

Prevalence of iris mammillations in the general population is unknown. We can ensure that these lesions are rare, probably underdiagnosed, to be considered as a sign of ocular melanocytosis and be followed by the long-term risk posed by developing uveal melanoma.

Conflict of interest

Authors declare having no conflict of interest.

REFERENCES