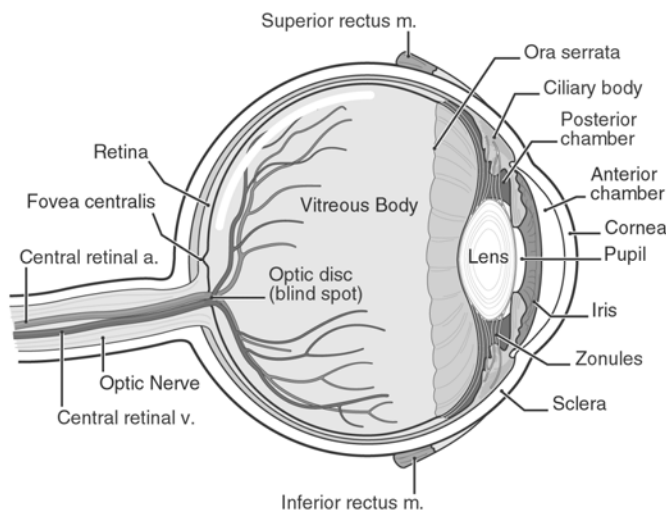


What You Should Know About Pigmented Lesions of the Retinal Pigment Epithelium

By David J. Browning, MD, PhD

Occasionally, a routine eye examination will reveal a dark lesion of the multi-layered lining in the back of the eye, and the patient will be referred for a consultation with a retina specialist to determine its nature. The question boils down to this: Is the lesion a cancer, a melanoma, or is it a benign lesion? Pigmented, benign lesions of the back of the eye are most often moles, or nevi, which develop in the choroid, a layer of blood vessels and connective tissue just inside the sclera, the white wall of the eye. Less



often, they are lesions of the RPE, an acronym for retinal pigment epithelium, which is a layer of cells lining the inside of the choroid. The retina lies just inside the RPE and is the nerve layer which transforms light landing on the back of the eye into a nerve signal traveling to the brain. These layers of the eye are illustrated in figure 1 to help you visualize the concepts we will discuss.

Figure 1. Anatomy of the Eye

The pigmented lesions of the RPE are of three types. By far the most common is congenital hypertrophy of the RPE, found in 2% of patients. Less frequent is the lesion known as Bear Tracks, found in 0.12% of patients. Least common are lesions associated with Familial Adenomatous Polyposis, also known as Gardner's Syndrome, found in 0.004% of patients.

Congenital Hypertrophy of the RPE (CHRPE)

This lesion is usually solitary, and can be found anywhere in the RPE, varying in size from 0.1 mm to 24 mm in diameter. There is no thickness to

the lesion, an important point in distinguishing it from a melanoma. It is usually gray to black in color, and may have a depigmented halo surrounding the lesion and often depigmented spots within the lesion called lacunae. The retina overlying these depigmented spots is often dead, and careful visual testing can show blind spots in the field of vision corresponding to these lacunae. I have never seen a patient who noticed

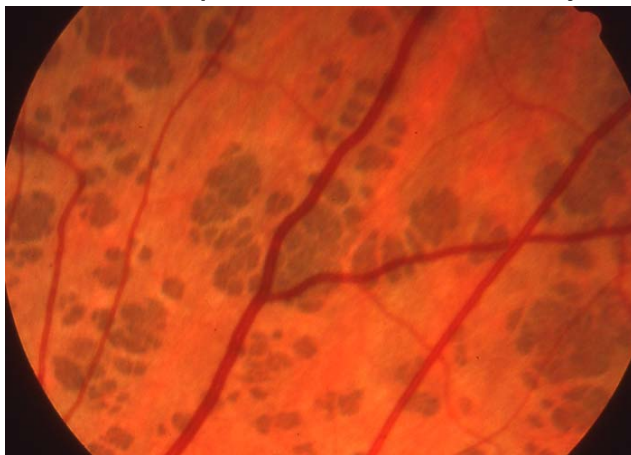


this in day-to-day life. Over a period of decades, these lesions may expand slightly, but they never uniformly thicken. Rarely, a localized nodule of thickening can develop. These lesions simply need to be photographed as a baseline measure for future comparison. They do not develop into cancers later in life. There is no known tendency for CHRPE to be inherited. A photograph of CHRPE is shown in figure 2.

Figure 2. CHRPE

Bear Tracks

Sometimes patients have multiple smaller patches of pigment in the RPE. Because these patches are often clustered together, they can look like the footprints of a bear in the mud, hence the name Bear Tracks. Under a microscope, these lesions look just like CHRPE. These lesions do not

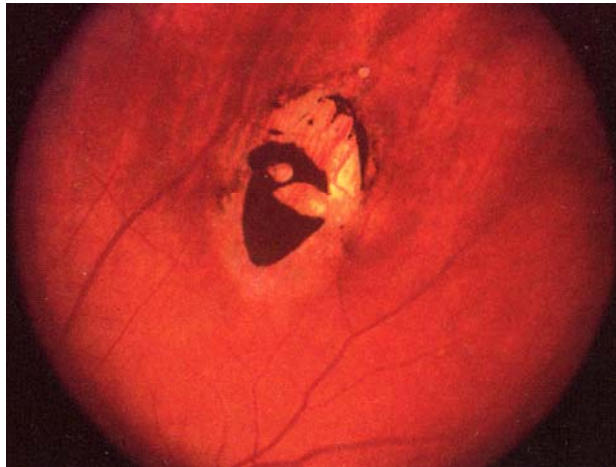


turn into cancer and they never cause loss of vision. A baseline photograph of the lesions may be recommended for future comparison, and no extraordinary follow-up is needed. Bear Tracks are not an inherited condition. An example of Bear Tracks is shown in figure 3.

Figure 3. Bear Tracks

RPE Lesions of Familial Adenomatous Polyposis (Gardner's Syndrome)

Familial Adenomatous Polyposis is a genetic condition caused by a mutation in chromosome 5q. Each cell of the body has 26 pairs of chromosomes, one of each pair inherited from the mother, and the other of each pair inherited from the father. This mutation causes a number of abnormalities, the most important being development of polyps in the colon, the large bowel. These polyps always turn cancerous; therefore a way to predict who will get them is significant. One of the best ways to predict development of the polyps is to look in the eye, since two-thirds of affected patients will have more than four black lesions of the RPE. These



lesions superficially resemble CHRPE, but there are important differences in closer study. They are not as large on average, and they usually have a spindle shape. They frequently have a white tail on one end. All patients who have this finding must have yearly sigmoidoscopy of the colon to look for polyps. A lesion associated with Familial Adenomatous Polyposis (FAP) is shown in figure 4.

Figure 4. FAP

Final Comments

Almost all lesions of the RPE are benign, and only one, the rarest, is associated with a cancerous syndrome. If you have questions after reading this brochure, please call my office, at (704) 295-3180. If you would like to learn more about Pigmented Lesions of the Retinal Pigment Epithelium or another medical condition, an excellent resource is the PubMed site within the National Library of Medicine website. This site can be accessed via any search engine, or at the following link:

<http://www.ncbi.nlm.nih.gov/entrez/query.fcgi>.