Elemental Composition of Ora Serrata Pearls—A Form of Focal Nodular Drusen

Examination of the peripheral fundus may reveal discrete pearl-like structures at the furthest extent of the retina. They occur throughout the ora serrata region, but only where a tongue of retina tissue in the form of a dentate process overlies the pars plana. In the later stages, the pigment epithelial covering is lost, exposing the pearl as a glistening bead. To our knowledge, the clinical significance and precise composition of these structures have not been described, although there is suggestion that they consist of drusenlike material. In this study, scanning electron microscopy with energy dispersive x-ray spectroscopy (SEM-EDS) was applied in conjunction with classic histochemical staining techniques to characterize the elemental content of these pearls.

Methods | A formalin-fixed enucleated globe was submitted for routine pathological examination and found to contain 3 pearls at the region of the ora serrata. Sections of the gross specimen were photographed, dissected (R.E.E.), and sent to the Barbara W. Streeten Ocular Pathology Laboratory at the State University of New York Upstate Medical University. Specimens were embedded in paraffin and submitted for hematoxylin-eosin, periodic acid–Schiff, and van Gieson elastic stains. A section was placed on a carbon stub for SEM-EDS using the FEI Aspex personal scanning electron microscope (FEI Inc).

Results | On gross examination, 3 pearls were located at the dentate processes of the ora serrata. Light microscopy revealed that the pearls were attached to the Bruch membrane and covered by retinal pigment epithelium cells and neurosensory retina. The pearl-like masses stained positively with periodic acid–Schiff and stained negatively with van Gieson elastic. Scanning electron microscopy with energy dispersive x-ray spectroscopy showed discrete heterogeneous masses that demonstrated emission peaks corresponding to calcium and phosphorus, suggesting the presence of calcium phosphate.

Discussion | Ora serrata pearls have been described in the literature as idiopathic drusenlike material based on their general appearance and simple staining patterns. In this study, we described the histopathological appearance (Figure 1) and elemental composition of ora serrata pearls. As in past studies, pearls were observed by light microscopy as eosinophilic periodic acid–Schiff-positive spheres located within or beneath retinal tissue, projecting from the Bruch membrane and covered by retinal pigment epithelium.¹ Using SEM-EDS, we have demonstrated that the calcium within ora serrata pearls is in the form of calcium phosphate (Figure 2), the same calcium salt seen in dystrophic calcification of drusen.² We found ora serrata pearls to be morphologically consistent with focal nodular drusen, indistinguishable from drusen previously described.³

The presence of drusen in any form in the furthest extent of the peripheral retina may help explain the pathophysiologic mechanism of its formation. We found ora serrata pearls only in elongations of dentate processes, which lie anterior to the termination of the choriocapillaris in an area made up mostly of veins.⁴ The propensity of pearls to form in this low-oxygen tension area, and not in areas of richer oxygen supply, such as the ora bays, is well documented.¹,⁵,⁶ This pattern
of formation suggests a pathophysiologic mechanism involving ischemia-induced retinal degeneration. In light of our SEM-EDS analyses that characterize pearls as calcified drusen, we may infer that a similar ischemic processunderlies the formation of other retinal drusen. Because ischemia is a known trigger for phosphate-dependent calcium accumulation (eg, in mitochondria), the paucity of arterioles at dentate processes would similarly explain the extensive calcification that can be seen in pearls of the ora serrata.

In conclusion, we propose that pearls are not idiopathic developmental bodies, but rather drusen that result from physiologic, age-related, or other ischemic change. While pearls do not affect vision, they remain clinically relevant because their composition and distribution may support an ischemic basis for the development of drusen-related disease.

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OBSERVATION

A Postmortem Ocular Finding of Tache Noire in a Living Patient

Patients in the intensive care unit develop exposure keratopathy in the setting of sedation and severe illness. This chronic drying of the ocular surface can cause corneal ulceration and even perforation.1 In a patient who was being treated in the intensive care unit, desiccation in the exposed part of the eye produced a pattern of scleral discoloration known to forensic pathologists as tache noire de la sclerotic, which is an early postmortem darkening of the sclera where the eye is not covered by the lids.2 To our knowledge, this is the first report of a postmortem eye finding in a living patient.

Report of a Case | The ophthalmology service was consulted for abnormal discoloration of the right eye, thought to represent subconjunctival hemorrhage, in a woman in her late 50s being treated in an intensive care unit for sepsis and multi-organ dysfunction. During her hospitalization, she experienced multiple vascular ischemic events that were likely embolic and an increasing leukocytosis of unknown origin with progressively worsening mental status. She had no known history of inflammatory eye disease or systemic vasculitis.

On ophthalmic examination, visual acuity and visual field assessment were limited by the patient’s mental status. The right pupil was nonreactive and miotic with an afferent papillary defect by reverse testing, and the left pupil was normal. The right eye exhibited lagophthalmos and a perilimbal, dark brown subconjunctival hue was observed at the 3- and 9-o’clock positions in the exposed areas of the eye. The discolored areas were completely covered by conjunctiva and were not associated with scleral thinning (Figure 1). The conjunctiva in the unaffected areas were not injected. Portable slitlamp examination of the right eye revealed superficial punctate keratopathy. The left eye exhibited normal lid closure, and the cornea was clear. In both eyes, the anterior chamber, lens, and dilated fundus examination findings were unremarkable.

A regimen of lubricating ointment and artificial tears for the right eye was instituted and an adhesive was used to close the eyelid. On follow-up examination the next morning, the discoloration of the patient’s right eye had almost completely resolved (Figure 2). The patient died of systemic illness several hours later. An autoimmune laboratory workup result was negative. Pathology corroborated the scleral darkening to be tache noire.

Discussion | In this case, we observed lagophthalmos and corneal surface changes commonly found in severely ill patients. In addition, however, this patient presented with marked scleral darkening in the exposed areas of the involved eye. One of the most serious differential diagnoses for dark discoloration of the sclera is necrotizing scleritis, which is treated with a course of immunosuppressive or immune-modulating therapy.3 This patient had no history of inflammatory eye disease. Given that the discoloration resolved significantly with ocular surface lubrication overnight, we believe that it was due to severe surface dryness rather than an autoimmune or infectious process. While, to our knowledge, there is no similar finding recorded in the ophthalmic literature, this pattern of discoloration in the exposed areas of the eye is a common postmortem finding known in forensic medicine as tache noire, which is French for black spot.4 This finding can be observed...