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NOTE

Surgery

A case of unusual persistent pupillary membrane with total anterior capsular pigmentation

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ABSTRACT. A 5-year-old castrated male poodle presented with blindness. Ophthalmic examinations including slit-lamp biomicroscopy, tonometry, ultrasonography, and electroretinography were performed. The anterior lens capsule of the right eye (OD) was totally pigmented, with persistent pupillary membranes (PPMs). Ultrasonography of the same eye showed severe lens atrophy and retinal detachment. Electroretinography revealed flat a- and b-waves in OD, but normal amplitudes in the left eye (OS). No ocular defects were detected in OS except mature cataract. In this case, it was determined that hypermature cataract with PPMs caused both lens-induced-uveitis and total anterior lens capsule pigmentation. This condition needs to be differentiated from absent pupil. Notably, PPMs with total anterior lens capsular pigmentation are extremely rare in dogs.

KEY WORDS: canine, lens capsule, persistent pupillary membranes, pigmentation

Animal studies show that fetal intraocular vasculature (FIV) provides nutrition to the lens and anterior segment during the period of rapid eye differentiation [4]. Once the ciliary body begins actively producing aqueous humor, the hyaloid system is no longer needed and starts to regress [2]. Atrophy of the FIV occurs initially through apoptosis, and later through cellular necrosis, and is usually completed by the time of eye lid opening, 14 days postnatally. Persistent fetal intraocular vasculature (PFIV) is a congenital anomaly attributable to the persistence of embryonic vascular vestiges within the eye that did not undergo normal involution during the prenatal or early postnatal period of ocular development [8]. In humans, numerous morphologic variants of PFIV may occur including persistent pupillary membranes (PPMs), anterior or posterior tunica vasculosa lentis (TVL), Mittendorf dot, persistence of the hyaloid artery, and Bergmeister papilla with malformations in the shape and size of the globe [5].

Although lens capsule pigmentation due to uveitis is very common, in this case, we observed total anterior lens capsule pigmentation, and the pigment color was the same as that of the iris. It could easily be misdiagnosed as absent pupil, and therefore, it is important to differentiate between the two.

Total persistence of pupillary membrane covering the entire pupil is extremely rare, and is associated with other ocular anomalies such as microphthalmos and cataract [6].

In this case report, we present a rare case of persistent pupillary membranes with total anterior lens capsular pigmentation, in a poodle.

A 5-year-old male castrated Toy Poodle, weighing 6 kg, presented with a 1-week history of severely decreased vision. A neuro-ophthalmic examination revealed the absence of dazzle reflex and menace response in the right eye (OD), and a slight menace response and normal dazzle reflex in the left eye (OS). Direct pupillary light reflex (PLR) in OD was absent, and indirect PLR in OD was normal; whereas the direct PLR in OS was normal, and indirect PLR in OS was absent. Palpebral and corneal reflexes were normal in both eyes (OU).

A complete ophthalmic examination was performed in this patient, before and after mydriasis induced with 0.5% tropicamide (Mydrin-P®, SANTEN OY, Tampere, Finland). Slit-lamp biomicroscopy (Topcon Model SL-D7®, Topcon Corp., Tokyo, Japan) was performed, which showed pigmented anterior lens capsule and mild persistent pupillary membranes (PPMs) at the bottom of the pupil (Fig. 1a) in OD. After mydriasis, iris-to-lens PPMs were detected everywhere except at the 3, 9 and 12 o'clock positions; total anterior lens capsular pigmentation was also seen in the same eye. Mature cataract with slightly irregular iris margins was noticed in OS.

Rebound tonometry (TonoVet®, Tiolat, Helsinki, Finland) revealed intraocular pressures (IOPs) of 8 and 17 mmHg in OD and

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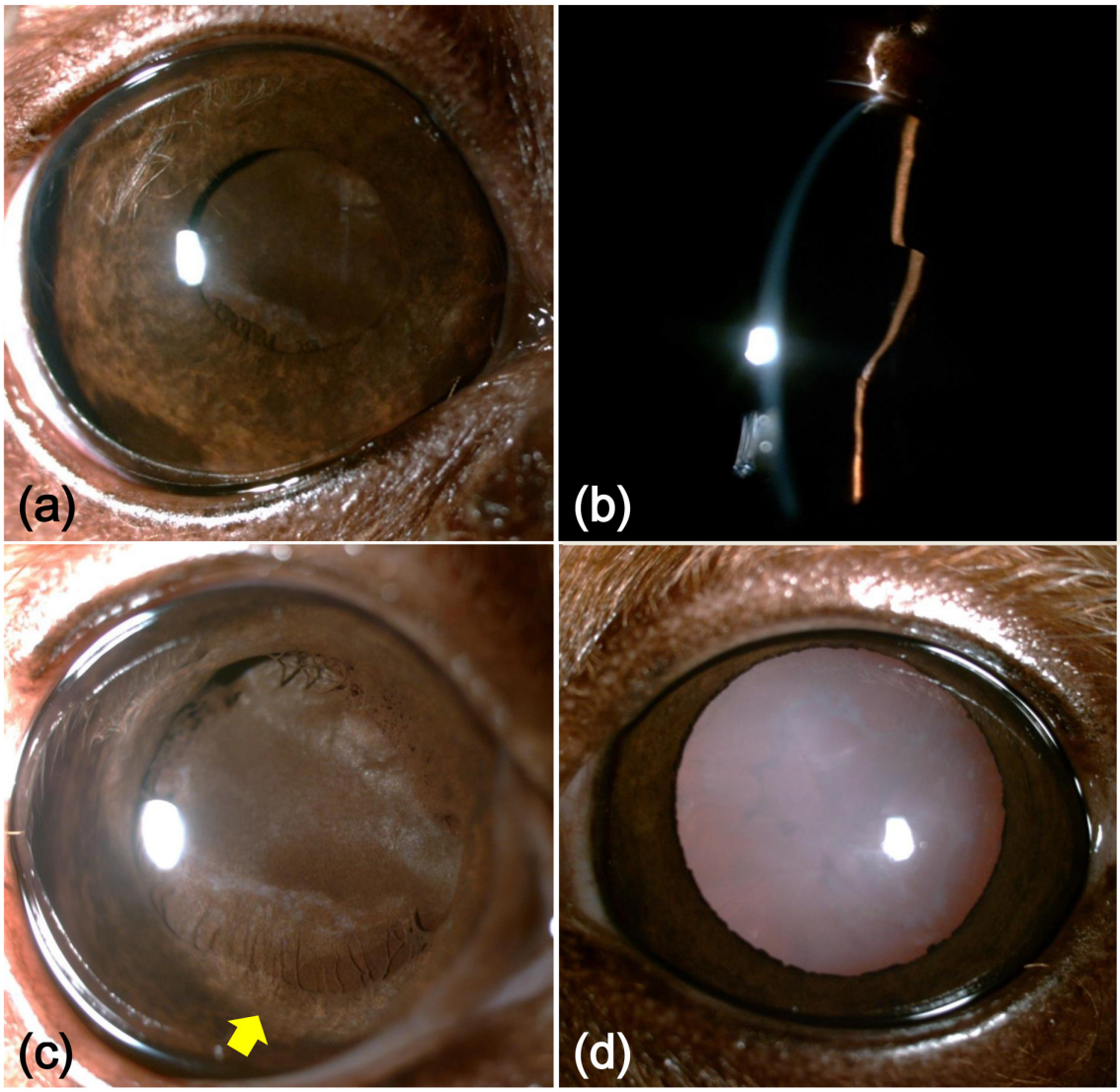

Fig. 1. Slit-lamp biomicroscopy images: Pigmented anterior lens capsule and mild persistent pupillary membranes (PPMs) were present (a), and the slit beam did not penetrate through the pupil (b) at the right eye (OD). After mydriasis, general iris-to-lens PPMs were detected, except at the 3, 9 and 12 o'clock positions. Total anterior capsular pigmentation was detected. Fine iridial strands, similar to blood vessels, originated near the iris collarette between the 6 to 8 o'clock positions (arrow) (c). A mature cataract with mild irregular iris margins was observed in the left eye (OS) (d).

OS, respectively. The posterior parts of the eyes were not visible with indirect ophthalmoscopy (Vantage®, Keeler Instruments Inc., Broomall, PA, U.S.A.) with a 30-diopter indirect lens (Classic BIO lens®, Volk, Volk optical Inc., Mentor, OH, U.S.A.), because of the PPMs and pigmentation on the anterior lens capsule in OD and mature cataract in OS.

Ocular ultrasonography (ProSoundα6®, Hitachi Aloca Medical, Ltd., Tokyo, Japan) revealed a resorbed lens, moderate vitreal degeneration, and retinal detachment in OD; and mature cataract and mild vitreal degeneration in OS (Fig. 2). The axial lengths of the eye globes were 17.6 and 17.7 mm in OD and OS, respectively. The lens thicknesses were 2.4 and 8.3 mm in OD and OS, respectively. The lens diameters were 11.7 and 11.9 mm in OD and OS, respectively. The results of the scotopic electroretinography (ERG, RETIport®, Roland Instrument, Brandenburg, Germany) in response to a bright flash are shown in Fig. 3. The light stimulus was transmitted through a contact lens with a high luminance diode (LED-electrode, MAYO Corp., Inazawa, Japan). ERG showed flat appearances of the a- and b-waves in OD. The values for b-wave amplitude were 9.48 μV for OD, and 248 μV for OS.

PPM is a congenital ocular disorder with early fetal manifestations [7]. PPMs occur commonly in dogs and are usually incidental findings. The association between persistent hyperplastic tunica vasculosa lentis and persistent hyperplastic primary vitreous (PHTVL/PHPV), with PPMs or microphthalmos, has also been described in dogs [7] and cats [1]. Iris-to-lens type PPMs can cause cataracts. Also severe uveitis induced by hypermature cataract can result in lens capsule pigmentation. PPMs have their origins near the iris collarette region [9], whereas posterior synechia always presents with pigment clumps on the anterior lens capsule [6]. In our case, the pupil was not fixed, and normal miosis and mydriasis were observed on indirect PLR in OD. Fine iridial strands, similar to blood vessels, were seen to originate near the iris collarette between the 6 to 8 o'clock positions.

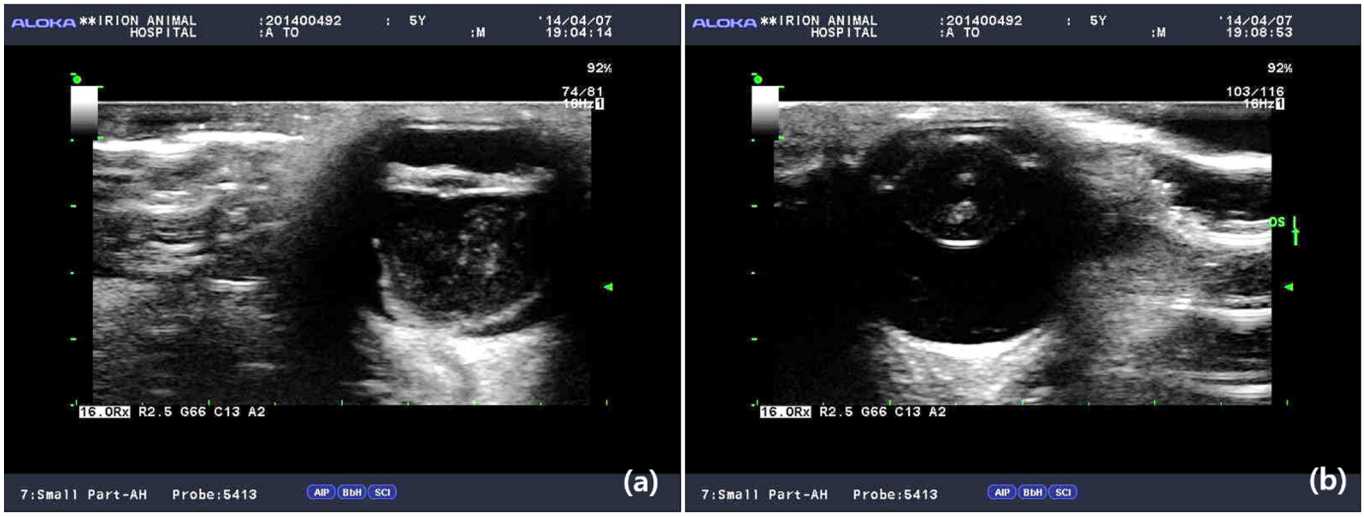

Fig. 2. Ocular ultrasonography: Small resorbed lens, moderately degenerated vitreous, and retinal detachment were detected in OD (a). OS was normal except for cataractous lens (b).

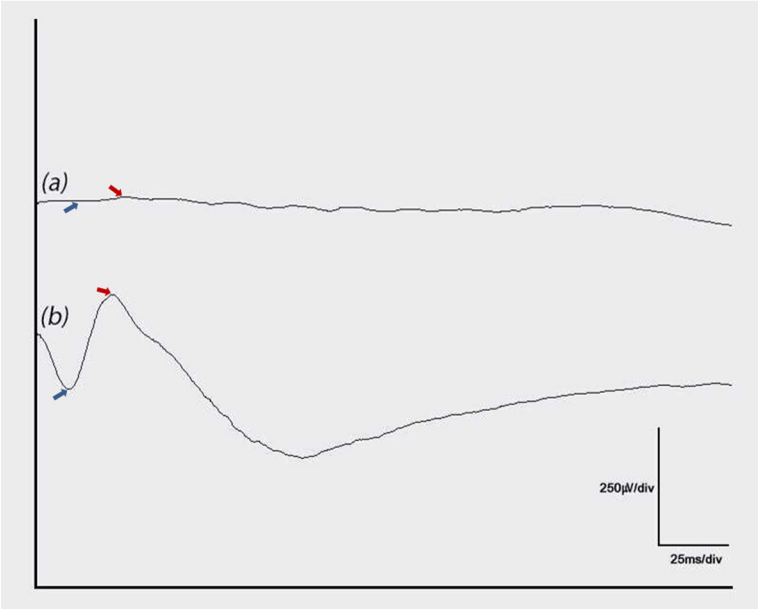

Fig. 3. Results of electroretinography (ERG): The a- and b-waves were flat, and the b-wave amplitude was 9.48 μV in OD (a). The a- (blue arrow) and b- (red arrow) waves were normal, and b-wave amplitude was 248 μV in OS (b).

The patient had a previous history of cataract in OS, approximately a year ago. He came in after the cataract had matured to the point of interfering with his daily life, and he was unable to see properly. The owner was not aware of the condition of the OD until the OS was diagnosed with a mature cataract, at the time of presentation. Although an obstacle test confirmed that the dog had minimal vision, the owner was concerned about the impaired vision. The menace response was decreased and the dazzle reflex was normal in OS, and both reflexes were absent in OD. The reduced vision was probably caused by a mature cataract in OS, and the OD had been blind for a long time. The lens of OD was cataractous and had undergone resorption, indicating chronic lens-induced uveitis (LIU). LIU is an inflammatory response of the ocular uvea against lens proteins, and has been described as a naturally occurring disease in many animal species, including dogs [12]. In the veterinary literature, LIU has been divided into two syndromes named ‘phacoclastic uveitis’ and ‘phacolytic uveitis’ [14]. Phacolytic uveitis is a lymphocytic-plasmacytic anterior uveitis that may occur secondary to the release of lens protein through an intact lens capsule [14]. Miniature Poodles, Toy Poodles, and American Cocker Spaniels are the breeds commonly affected, reflecting the high incidence of cataract in these breeds [13]. Anterior or posterior synechia, pigment on the anterior lens capsule, and iris hyperpigmentation are the clinical signs associated

with chronic uveitis [13]. In our case, resorbed lens size was detected. Consequently, the anterior lens capsular pigmentation arose in part from pre-iridal fibrovascular membranes, and partly as a result of pigment migration due to chronic LIU.

Because of the PPMs with total anterior lens capsular pigmentation, the direct PLR in OD was absent. Normal indirect PLR in OD was indicative of a normally functioning optic chiasm. Cataract induced by the PPMs of OD probably led to the development of microphakia, vitreous degeneration, retinal detachment, and total anterior lens capsular pigmentations, which were probably the result of the combination of PPMs and phacolytic uveitis associated with cataract progression. In this case, the lens resorbed and shrunk, just as in phacolytic uveitis. On the other hand, if phacoclastic uveitis had been present, the patient would have had uncontrolled inflammation, and developed phthisis bulbi or secondary glaucoma [6]; however, no such events were reported in the patient's history. The family members were unaware of the concurrent condition of OD, because there were no external manifestations. Even a veterinarian could probably not diagnose the ocular condition of OD without a complete ophthalmic examination. Considering the current status of OD, we can safely assume that it did not develop recently.

In humans, when vision is impaired by PPMs, surgical treatment options using lasers are available [11]. However, this procedure is uncommon in veterinary medicine, and due to the resorbed lens and retinal detachment in OD, surgical treatment for OD was not an option in this patient. Cataract surgery was recommended for OS, and phacoemulsification was performed. Vision was restored in OS after surgery.

PPMs with other anterior chamber abnormalities may occur due to an autosomal dominant inherited trait [3]. Moreover, PPMs are usually sporadic. Nevertheless, genetic molecular studies may be appropriate in familial cases to identify the genetic locus, and to help explain other cases of isolated anterior segment abnormalities [10]. However in this case, the family history could not be tracked.

Histopathological evidence would have confirmed the etiology in this case; however, the owner was unwilling to pursue enucleation of OD. We concluded that the cause of both LIU and total anterior lens capsule pigmentation, was hypermature cataract with PPM. It is important to differentiate between an absent pupil and total anterior lens capsule pigmentation, to prevent a faulty diagnosis.

In summary, this report presents a rare case of PPM with total anterior lens capsular pigmentation covering the entire pupil, in a poodle.

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