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Non-Inflammatory Disease of the Uvea in Horses

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Take Home Message

Abnormalities of the equine uvea are relatively uncommon and often of minimal consequence to vision or comfort. However, some innocuous conditions can mimic serious disease and therefore proper assessment is important to avoid misinterpretation and inappropriate clinical action. The purpose of this lecture is to review noninflammatory diseases of the equine uvea.

Congenital Diseases

Aniridia

While the term aniridia infers a complete absence of an iris, it has been used to describe a clinical condition reported in horses where only a rudimentary, non-functional ridge of iridal tissue is present. Aniridia has been reported in a Belgian Draft stallion (along with 65 of his offspring), a Quarter Horse stallion, a Thoroughbred colt and a Welsh/Thoroughbred cross filly. Aniridia is thought to be heritable via an autosomal dominant mode. Horses affected with aniridia are usually presented within the first few months of age because the client has noticed such signs as an unusual appearance of both eyes, squinting excessively when in bright sunlight, and/or overreacting to flashes of light. On ophthalmic examination, the pupils are widely dilated and non-responsive. The extent of the pupillary dilation is so marked that the lens equator and ciliary body processes are visible. The corpora nigra is absent. Posterior segment examinations in horses with aniridia have been uniformly unremarkable.

Anterior Uveal Cysts

The cysts discussed here may not be congenital, but may be present as very small cysts at birth only to enlarge and become apparent later in life. Anterior uveal cysts may be found in four locations: 1) the corpora nigra, 2) along the margin of the pupil or free floating in the anterior chamber, 3) attached to the ciliary body, and 4) within the iris stroma (Fig. 1).

When the corpora nigra becomes cystic, the normally roughened appearance of the corpora nigra becomes smooth and spherical. The condition may be unilateral or bilateral, and the size of the cysts can vary markedly. Most are sufficiently small that they do not cause significant interference with vision. However, depending on the specific location, even moderate-sized cysts can partially inhibit the visual field, especially when the horse is in bright light and the pupil is miotic. Differential considerations include inflammatory or neoplastic changes to the iris in the area of the corpora nigra. Although ultrasound could be used to differentiate cystic corpora nigra from inflammatory or neoplastic infiltrates, this is rarely necessary as the clinical appearance is
usually sufficiently characteristic. Treatment is not ordinarily necessary, the most effective and non-invasive treatment is deflation of the cystic corpora nigra with a laser.\textsuperscript{7}

Iridal cysts may be found distinctly separate from the corpora nigra along the margin of the pupil or free floating in the anterior chamber. These cysts are thought to develop similarly to iridal cysts in dogs and represent a failure of the two layers of neuroectoderm to completely fuse, thus allowing fluid to accumulate in areas between the 2-layered posterior iris epithelium. Because iridal cysts can enlarge, it has been theorized that some of the posterior iridal epithelial cells that comprise the lining of the cyst retain secretory ability.\textsuperscript{6}

![Fig. 1. Cystic corpora nigra.](image1)

![Fig. 2. Ciliary body cyst.](image2)

Ciliary body cysts (Fig. 2) are a feature of multiple congenital ocular anomalies of Rocky Mountain horse\textsuperscript{8} and are thought to originate in a manner similar to that of iridal cysts, except in this instance the failure of the two layers of the neuroectoderm to fuse is in the area of the ciliary body. Other potential causes of ciliary body cysts include inflammatory processes, traction from zonules, anterior segment dysgenesis, and age. They tend to be oval and relatively large, often extending from the pars plicata to the ora ciliaris retinae. Their most common location is ventrolaterally and they can be difficult to detect without pharmacologic mydriasis. As with the other anterior uveal cysts discussed, differential considerations would include inflammatory and neoplastic disease. While ocular ultrasound can be used to confirm the diagnosis of ciliary body cyst, the shape, contour, and ability to transilluminate make this differentiation straightforward on the basis of clinical signs alone. These cysts do not routinely cause pain or visual impairment and treatment is not necessary.\textsuperscript{6}

\textit{Iris Hypoplasia}

Historically, dark and bulging areas in the anterior surface of the iris have been interpreted as cysts within the iris stroma.\textsuperscript{9} These dark, bulging areas are seen predominately in the dorsal region of eyes with blue irides, although they can be present in the ventral aspect of the iris and in eyes with brown irides. These lesions represent areas of iridal stromal hypoplasia and that the resulting protrusion is a result of aqueous pressure pushing the relatively weaker portion of iris
forward. The affected eyes are not painful and there is no discernable effect on vision. The fact that these protruding areas actually represent thinning of the iris stroma is confirmed by passing a bright, focal beam of light through the pupil and observing the retroillumination of the fundic reflection through the lesion. When the pupil is subsequently dilated either by the use of mydriatic agents or dark adaptation, the bulging area disappears, taking on a wrinkled appearance. The most important rule out for this clinical presentation is iridal neoplasia, specifically melanoma. Ocular ultrasound can aid in making this differentiation, but careful examination under conditions of both miosis and mydriasis as described above should allow for an accurate diagnosis.

Medulloepithelioma

Medulloepithelioma of the anterior uvea is relatively uncommon and these tumors arise from undifferentiated medullary epithelial cells that line the inner portion of the embryonic optic cup. Normally, these cells differentiate into numerous epithelial cell lines in the adult eye including the posterior iris epithelium, the ciliary body non pigmented epithelium (NPE), and the retinal epithelium. In the horse, medulloepitheliomas most frequently arise from the ciliary body region. Histologically, medulloepitheliomas usually demonstrate nonpigmented neuroepithelial cells arranged in tubules and rosettes. They are classified as either teratoid or non-teratoid depending on whether the neoplastic cell population is characterized by relatively homogeneous, undifferentiated primitive neuroepithelium or contains tissues not normally found in the eye such as cartilage, muscle, and brain tissue. Medulloepitheliomas are most frequently diagnosed in young adult horses presented for a fleshly mass noted within the pupil or anterior chamber. Although these tumors are thought to grow slowly, they may be associated with other ophthalmic changes such as corneal vascularization, corneal edema, corneal fibrosis, anterior uveitis, glaucoma, and buphthalmos. Enucleation is the treatment of choice.

Acquired Diseases

Melanoma

Although relatively rare overall, intraocular pigmented masses are reported in older gray horses, usually with concurrent cutaneous melanoma (Fig. 3). The initial clinical appearance of anterior uveal melanomas differs based on whether the ciliary body or the iris is the tissue of origin, how rapidly the neoplasm is growing, and the length of time before seeking a medical evaluation. When presented early, common clinical signs include focal corneal edema with a dark mass filing the anterior chamber, which is often in contact with the posterior surface of the cornea. As the neoplasm gets larger over time, common clinical signs would include blindness, blepharospasm, epiphora, buphthalmos, diffuse corneal edema, aqueous flare, anterior chamber masses that often obliterate any normal architecture and prevent viewing deeper structures. When structures deep to the iris can be seen, the lens is usually cataractous. If the tissue of origin is the ciliary body, a dark mass may be seen on the posterior surface of the lens extending into the vitreous cavity. Differential considerations for a dark mass in the anterior chamber would include a uveal cyst and iris hypoplasia. The distinction is usually not difficult on the basis of clinical appearance as cysts of the ciliary body, iris, and corpora nigra are all smooth, and oval to spherical.
Surgical ND:YAG laser treatment of uveal melanomas in equine eyes has caused shrinkage of the mass; however, corneal edema and secondary uveitis is a common side effect of laser therapy. Among equine patients, there is one reported instance where sector iridectomy was successfully employed in the treatment of an intraocular melanoma. Oral cimetidine (dose 2.5 mg/kg of body weight, PO, q 8 h) has been used to shrink non-ocular melanomas in horses, but no studies have been done to determine the effect of cimetidine on uveal masses. Generally, intraocular melanoma in horses has been treated by enucleation or exenteration. Success with these procedures has been high, and metastasis has not been reported.

**Lymphoma**

Lymphoma is the most common systemic neoplastic disease with an ocular manifestation in the horse. In one study of 79 horses with confirmed systemic lymphoma, 21 had ocular lesions. Of these, eyelid swelling and inflammation was the most common ocular sign (11/21) with anterior uveal involvement the next most common (4/21). The signs associated with anterior uveal manifestation of systemic lymphoma are non-specific and include blepharospasm, episcleral injection, corneal edema and vascularization, aqueous flare, hypopyon, hyphema, iridal congestion and swelling. Frequently, these non-specific signs are accompanied with a history of chronicity and poor response to anti-inflammatory medication. In addition, the ophthalmic signs are usually accompanied by non-specific signs of systemic disease such as fever, respiratory disorders, weight loss, peripheral lymphadenopathy, and anemia.

On the basis of ocular signs alone it is usually not possible to differentiate intraocular lymphoma from other potential causes of anterior uveitis. The diagnosis of systemic lymphoma is to be suspected in any horse with anterior uveitis especially when accompanied by systemic signs of illness such as fever, weight loss, lethargy, and swollen lymph nodes. The diagnosis of lymphoma can be confirmed by identifying neoplastic lymphocytes from samples of solid tissues such as the spleen, lymph nodes, or skin nodules, or by centesis of affected body cavities.
(thoracic or abdominal). In general, success rates for treatment have been low for systemic lymphoma. The ocular portion of the treatment regimen would be the same as discussed for anterior uveitis.

References