Are You Giving Me The Hairy Eye?

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Introduction:

A choristoma is normal tissue in an abnormal location, and ocular dermoids are choristomas that have features of cutaneous tissue[4]. They do not show signs of neoplastic or dysplastic changes, and consist of various dermal tissue components, including sebaceous glands, hair, hair follicles, keratinized stratified squamous epithelium, and other adnexal structures[5]. According to Mann, dermoids can be classified into three different types. Limbal dermoids represent the most common and least severe type and affect the temporal limbus region. The second type is characterized by its expanse and presence in deeper layers of the cornea, involving most of the surface and getting as deep as Descemet's membrane, but not involving it. The last type is even more diffuse and involves the anterior segment of the eye.[1]

Ocular dermoids have been identified in many species, including rats, guinea pigs, cats, dogs, pigs, cattle, and humans[1,5]. In dogs, the most commonly affected breeds include German Shepherds, Saint Bernards, Bassett Hounds, Dachshunds, and Corgis[4]. Dermoids are considered benign congenital abnormalities in most of the affected species; but, there is some evidence suggesting that they have an autosomally recessive and polygenic mode of inheritance in cats, specifically the Birman and Burmese breeds, and Hereford cattle[1,3]. However, dermoids have been noted to be rare in cats[2]. The locations for ocular dermoids include the eyelids, conjunctiva, nictitating membrane, and limbus, with the limbus being the most common[1,4]. Typically, dermoids are unilateral, but have been reported to occur bilaterally in humans, cattle, and dogs[1]. Fortunately, the presence of an ocular dermoid does not necessarily mean that the animal will be affected with other congenital ocular abnormalities, unlike in 30% of humans who may have an additional congenital ocular anomaly[1]. And despite being present at birth, ocular dermoids may not be recognized in certain species until the animal is older and the eyes are open; it is usually the second week of life or later when a dermoid is recognized in animals[1].

Dermoids may also go unnoticed until they begin to cause clinical signs or secondary complications[2]. The clinical signs that can commonly be associated with dermoids are related to the severity of irritation caused by the structures of the dermoid, particularly how numerous the hair shafts are and where they are located[4]. The hair can cause signs of irritation also seen with ectopic cilia and distichiasis, such as epiphora, keratitis, and blepharospasm. The severity of the signs tends to correlate with the degree of irritation and whether or not the hairs cause ulcerations to the cornea. If the hairs cause corneal ulcers, clinical signs can be more severe and are associated with the ulcer. These signs are neovascularization, aqueous flare, reflex uveitis, and miosis.[2]

Case History and Presentation:

The current case study involves Ella, a 4.5-month old, female, Cavalier King Charles Spaniel. When Ella was approximately 6 weeks old, she was taken by the breeder to an ophthalmologist in Massachusetts. The ophthalmologist diagnosed her with bilateral ocular dermoids based on gross appearance. Ella was adopted shortly thereafter and did well in her new home, having mild irritation from her dermoids, but no corneal ulcers. She presented to the Mississippi State College of Veterinary Medicine Ophthalmology Service on January 2, 2018.

At presentation, Ella was bright, alert, and responsive. Her physical exam findings were unremarkable, with a temperature, pulse, and respiratory rate of 101.9 Fahrenheit, 120 beats per minute, and 32 breaths per minute, respectively. Her lungs had normal bronchovesicular sounds and there was no murmur or arrhythmia noted on thoracic auscultation. She had a small, non-painful, and reducible umbilical hernia that measured approximately 2x2 centimeters.

Ella's gross ophthalmic exam revealed bilateral ocular dermoids, with several hairs growing from them, and epiphora of both eyes. The left dermoid originated from the inferotemporal limbus and extended onto the cornea, while the right dermoid was originating from the inferotemporal conjunctiva and stopped at the limbus. There were also pinpoint areas of central corneal opacity noted bilaterally, near the hair shafts, that were suspected to be subepithelial corneal dystrophy. The intraocular pressure for the right eye was 13 mmHg and for the left eye it was 15 mmHg. The Schirmer tear test was 12 mm/20 seconds in the right eye and borderline at 12 mm/minute in the left eye. Both eyes were negative for evidence of fluorescein stain uptake and the remainder of the ophthalmic examination, including fundic examination was within normal limits.

Based on these physical and ophthalmic exam findings, it was recommended for Ella to have a bilateral superficial keratectomies and conjunctivectomies the following day, on January 3, 2018. Since Ella was an apparently healthy puppy, bloodwork consisted only of a PCV, TP, blood glucose, and Azostix BUN reading. She also had a blood glucose reading the morning of January 3rd due to food being withheld since midnight. Her results were within normal limits at 43%, 6 g/dl, 15-26, and 120 mg/dl on January 2nd, and a blood glucose of 111 mg/dl on the morning of January 3rd.

Pathophysiology:

Dermoids are not proliferative or neoplastic in nature, despite their growth associated with the development of the animal[4]. It is not understood whether dermoids are the result of a primary deviation in the corneogenic mesoderm, abnormal influences from embryonic inducers, or normal dermal tissue that simply becomes separated from other tissue destined to produce skin[6]. Although the exact pathophysiology is unknown, there have been speculations regarding their occurrence based on the components and stages of embryonic development[7]. During embryonic development, the germ cell layers, the mesoderm and ectoderm, have been proposed to play an important part in dermoid formation[6].

The most widely accepted theory involves metaplasia of the mesoderm due to excessive corneal exposure to the intrauterine environment from abnormal eyelid development and closure[7]. The mesoderm is the germ cell layer responsible for determining whether the ectoderm will develop keratinized or non-keratinized stratified squamous epithelium, as with skin or cornea, respectively[1]. Combined with signals from the optic cup and lens vesicle, the mesoderm and ectoderm form the cornea[6]. Without these critical influences, the ectoderm and mesoderm would form opaque tissue consistent with the sclera or skin[6]. The lens vesicle closes while the ectoderm on its surface, at the opening of the optic cup, becomes the corneal epithelium. Then, mesenchymal cells around the optic cup move between the lens vesicle and corneal epithelium[6]. The rest of the cornea then forms in waves from caudal to rostral, starting with the endothelium and moving to the stroma. In this theory, the mesenchyme that moves in later waves could contain abnormal or separated dermal tissue that develops into superficial dermoids; whereas, if the displaced mesenchyme were present in earlier waves, the dermoids would appear in deeper stromal layers.[6]

Another theory has been suggested based to the similarity of dermoid pigment and hair texture relative to surrounding facial skin and fur. The eyelids are formed from mesoderm folds that are covered by ectoderm during the same time as corneal stroma formation. In this situation, it would seem possible that mesoderm, destined to form eyelids, could invade the surface ectoderm forming the corneal epithelium and implant islands of mesoderm that can appear as dermoids on the cornea, third eyelid, or conjunctiva.[6,7]

Diagnostic Approach and Considerations:

A tentative diagnosis can be made based on visualizing a normal haired piece of skin at the limbus, on the conjunctival surface (palpebral or bulbar), or on the cornea. This is usually diagnosed in very young animals, but if little irritation is present, a dermoid can be an incidental finding in an adult animal. Though the significance was unknown, the case report by Brudenall noted that the puppy also had a persistent pupillary membrane in the eye affected by the dermoid[1]. This illustrates that it is important to perform a complete ophthalmic and physical exam to ensure there are no other congenital or hereditary abnormalities. The ophthalmic exam should be thorough and should include vision testing, pupillary light responses, fluorescein stain, schirmer tear test, tonometry, and a fundic examination. It is important to note that the depth of the dermoid cannot be determined until surgical removal[1].

Treatment and Management:

Treatment of ocular dermoids can be curative when surgery is employed. Medical management may be attempted, but is not the treatment of choice due to the potential for dermoids to cause persistent corneal irritation[2]. With medical management, removal of the dermoid hairs may be attempted by manual or electro-epilation, but this option is usually only a temporary solution as the hair can regrow[4]. The treatments in medical management often involve measures to prevent self-inflicted damage to the cornea by using an Elizabethan collar, pain control, a topical antibiotic, a mydriatic, and topical artificial tears or other ophthalmic lubrication that could potentially decrease the abrasiveness of the hairs[2]. Pain control may involve non-steroidals, oral opiates, and topical mydriatic agents. A topical antibiotic may also be chosen for treatment of an existing ulcer. However, consideration should be given to dermoid growth with the animal, making surgical removal later in life more difficult with more extensive stromal involvement, potentially requiring additional surgical techniques, such as a tarsorrhaphy, conjunctival flap, or third eyelid flap, to aid in healing[1,4].

Surgical removal of the dermoid is the treatment of choice. A superficial keratectomy performed under an operating microscope is curative and produces minimal scarring[2]. Based on the size of the dermoid, additional techniques may be used, as mentioned above. As with any procedure or surgery, there are potential complications. Certain complications can threaten sight, such as delayed healing, infection, corneal perforation, scarring, and keratomalacia[2]. Although these complications are uncommon, measures should be taken to avoid or prevent them when possible.

In another study performed by Minamide and Suzuki on a 9-month old Beagle puppy, an enucleation was chosen to remove the eye affected with the dermoid. The puppy was otherwise in normal health and no other littermates were affected. The reason for enucleation rather than a keratectomy was not specified, but it was also a curative, though more drastic, surgical solution.[5]

After removal of the dermoid, an Elizabethan collar should be used to prevent damage to the cornea and complication of the surgically-created corneal ulcer. An antibiotic ophthalmic solution or ointment should be employed to prevent secondary infection of the corneal defect where the dermoid was located. The antibiotic choice varies and may be broad-spectrum, such as a triple antibiotic, or an aminoglycoside given 2-4 times daily for up to a week, at which time the eye should be rechecked and the ulcer healed. In a case report by Lee and others, an antibiotic-steroid combination was suggested for after the ulcer had healed to reduce scarring and improve corneal transparency[4]. Post-operative treatment should also include pain medication, such as a topical or oral non-steroidal anti-inflammatory. A mydriatic, such as atropine, may also be used to help with comfort if there is evidence of reflex uveitis, such as miosis or aqueous flare.[1,2,4]

Case Outcome:

On January 3, 2018, Ella was placed under general anesthesia. Both eyes were surgically prepared with dilute betadine and saline. Cis-atracurium was employed to prevent eye movement during the procedure. A lateral canthotomy was performed and an eyelid speculum was placed in the right eye. Stevens tenotomy scissors were used to remove the dermoid from the lateral portion of the conjunctiva. A figure-8 suture was placed with the knot buried in the right lateral canthus using 6-0 Vicryl. A simple continuous pattern was also used to close the conjunctival defect where the dermoid was removed. The eyelid speculum was then moved to the left eye and a #6400 Beaver blade was used to incise the margins of the dermoid, approximately 1/3 corneal depth. A Martinez corneal dissector was used to evenly remove the dermoid from the corneal stroma. A simple continuous and simple interrupted pattern was also used to close the bulbar conjunctival defect of the right eye where the dermoid was removed and the corneal wound was left open to heal. Removal of the dermoid was evident by the clear corneal surface that remained and the absence of abnormal appearing and textured ocular tissue. Hemostasis was maintained throughout the procedure with a 1:10,000 epinephrine solution. The procedure was successful and did not have any intra-operative complications.

Ella recovered in anesthesia and her recovery was also uneventful. Bacitracin-neomycin-gramcidin ophthalmic solution was started 2-3 times daily to prevent an infection in the superficial ulcer and incisions that were surgically created. Carprofen and tramadol were administered orally for pain control. An Elizabethan collar was provided. The following morning, an ophthalmic exam was performed and there was a superficial corneal ulcer at the temporal limbus of the left eye, as expected. There was mild blepharospasm bilaterally. Miosis was present in the left eye, which suggested the presence of reflex uveitis, for which she was given atropine ophthalmic solution. She was discharged on January 4, 2018 with instructions to have a recheck in 5-7 days with her regular veterinarian in Massachusetts to ensure the surgical sites had healed appropriately. Approximately four weeks after the procedure, the left cornea had minimal scarring, but there was no evidence of recurrence or other complications.

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