Surgical Management of Congenital Ocular Dermoid Cyst: A Review

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Abstract

Ocular dermoid is a skin or skin-like appendage usually arising on the limbus, conjunctivae, and cornea. It can be unilateral or bilateral and may be associated with other ocular manifestation or with other malformations. Hair from the lesions is predominantly responsible for the associated irritation resulting in chronic inflammation of the conjunctivae and cornea and may cause visual impairment. Ocular dermoids are rare in cattle, with the prevalence estimated between 0.002% and 0.4%. Excessive lacrimation can be observed due to irritation caused by hair and cyst in the eyes. Superficial keratectomy is required to surgically excise a corneal dermoid although the depth of the dermoid within the cornea cannot be ascertained by ophthalmic examination until surgery is undertaken. In the case of large corneal dermoids, surgical excision should be performed early in the life of the patient to achieve optical improvement and allow functional development of the eye.

Keywords: Dermoid cyst, surgery.
1. Introduction

Congenital cysts are interesting because of their occurrence early in life. The causes of such growths must be different from those of the acquired cysts in adult life. Congenital cysts like dermoid cyst in calves are rare. Corneal dermoid is a congenital lesion observed rarely in newborn animals [1]. It is a skin or skin-like appendage usually arising on the limbus, conjunctivae and cornea [2]. The incidence of corneal dermoids in domestic animals is 3.4% [3]. Grossly the affected cornea appeared to be covered over part of its surface with haired, usually pigmented skin [1]. Hair from the lesions is predominantly responsible for the associated irritation resulting in chronic inflammation of the conjunctivae and cornea and may cause visual impairment [4]. Dermoids may affect the eyelids, conjunctiva, nictitans, sclera and cornea and most commonly present unilaterally. Bilateral ocular dermoids have been reported in cattle [5], sheep, and a camel [6]. Ocular dermoids are rare in cattle, with the prevalence estimated between 0.002% and 0.4% [7].

2. Ocular dermoid cyst

2.1 Disease

Ocular dermoid is a congenital defect recognized in animals Barkyoumb and Liepold [5], characterized by the islands of skin that are histologically normal but misplaced to an abnormal location, usually the lateral canthus, limbus, third eye lid, medial canthus, cornea and eyelid. The site affected by dermoid cysts varies with the species and even breed. In humans, they have been recorded in the brain [8], spinal cord [9], ovary [10], penis [11], tongue [11], orbit [12, 13, 14], nose [15], jaw bone [16], neck and rectum [11], whereas in dogs and cats the brain and spinal cord are most often affected. A single report described a dermoid cyst in the tongue of a German shepherd dog [17]. Gelatt (1991), reported a corneoconjunctival dermoid cyst in a 6-month-old male Hereford calf. Hillyer et al., 2003, reported six dermoid cysts along the dorsal midline of a 3-year-old thoroughbred-cross gelding. Dermoid cysts are early developmental lesions that occur because of the sequestration of embryonic epithelium in deeper layers along the fetal lines of closure [12].
Dermoid cysts have long been thought of as congenital and benign but it has also been reported as being acquired secondary to traumatic displacement of epithelial tissue [11]. In human, dermoid cysts are thought to arise; at between 3 and 5 weeks of gestation, from an abnormal implantation of surface ectoderm along the embryogenic lines of closure that form the facial features [13]. Heritability of dermoid cysts is thought to occur in Rhodesian ridgeback dogs; the condition is overrepresented in this breed and a linkage to one or two pairs of recessive genes is suspected. In horses, no breed disposition is recognised [11]. Being a congenital anomaly, dermoid cyst was recorded in different cattle breeds [7]. Although, it is believed not to be inherited [4], Barkyoumb and Leipold (1984), described the ocular dermoid cyst in Hereford cattle a genetically transmitted defect, in which characteristics of autosomal recessive and polygenic inheritance were observed [5, 18, 19]. The owner of the buffalo calf in this report also owned its sire and of the calves sired by the ox, none had shown any dermatological lesions to his knowledge. To our knowledge, there have been no reported cases of congenital ocular dermoid cyst in the buffalo; nevertheless, its occurrence is recommended to be duly noted in the breeding herds. Dermoid cysts may be unilateral or bilateral [19], but rarely appear bilaterally, except in certain lines of Hereford cattle [4]. Dermoid cysts are sometimes associated with microphthalmos [19]. Microphthalmos is more frequently the result of the involution of the primary optic vesicle or the abnormal closure of the embryonic cleft in which other ocular anomalies are present, including occasional cysts. Pure microphthalmos (nanophthalmos) occurs rarely, and is the result of an arrested development of the globe in all dimensions after the embryonic fissure has closed. The eye is small without other apparent abnormalities [4].

Dermoid cysts consist of keratinized epithelium and adnexal structures such as hair follicles, sweat glands, and sebaceous glands. It can be unilateral or bilateral and may be associated with other ocular manifestation, or with other malformations. Being a congenital anomaly, dermoid was recorded in different cattle breed. Dermoid cyst is an uncommon developmental anomaly that has been reported in dogs, cats, horses, and cattle. The cyst is usually congenital or hereditary in nature [20]. However, it is believed not to be inherited [7]. It may be solitary or multiple, firm to fluctuant, well circumscribed, smooth, and round and usually the overlaying skin is normal [21].
2.2 Etiology

It is believed that these cysts originate from an incarceration and subsequent growth of embrionary epithelial cells during the closure of the neural tube, and therefore, most of these lesions occur along the median line [22, 23]. However, there are reports of acquired dermoid cysts, secondary to traumatic epithelial dislocations [11]. The increased size of the cyst occurs due to normal cell desquamation within the cyst cavity leading to secondary signs related to the compression of adjacent structures. In most cases, dermoid cysts in dogs are associated with multiple vertebral and spinal malformations and hind limb neurologic deficits [24]. Barkyoyumb and Leipold (1984) described cardiac defects (Tetralogy of Fallot and patent ductus arteriosus), polycystic kidney disease and small tissue masses protruding into the external nares in some of 74 calves reported with ocular dermoids, although they did not specify the number of calves affected and whether calves showed one or a combination of the three abnormalities. Breed predispositions for ocular dermoids are reported in Birman cats, Dachshund, Dalmatian, Dobermann, Golden Retriever, German shepherd and Saint Bernard dogs, and Quarterhorses [7, 25]

2.3 Epidemiology

Ocular dermoids in cattle are not common, with an estimated prevalence of 0.002%–0.4% [26]. Ocular dermoids have been reported in cattle of many breeds worldwide, with a similar low prevalence in all breeds other than the Hereford [7]. Dermoid cysts are uncommon in the horse, with only three reports describing them in the dorsal midline, ventral thorax and distal limbs [11]. There have been single reports of dermoid cysts in other large animals, including the camel and bull [27, 28]. In contrast, these lesions are well documented in humans, dogs and cats [11]. Dermoid cysts represent the most common space-occupying orbital mass in childhood [12]. The apparent predisposition in Herefords is largely based on a report by Barkyoyumb and Leipold (1984) of 74 affected Hereford and Hereford-cross calves from one region of the USA, the majority of calves being bilaterally affected. The dermoid locations in that study were reported in decreasing order as limbus, third eyelid, canthus, eyelid and conjunctiva [6]. There are otherwise few reports of bilateral ocular dermoids in calves, each describing single or low numbers of animals [7], and only one reporting a nasal tumor-like growth. Of these bilateral cases,
inferonasal corneoconjunctival dermoids were most commonly reported, followed by nasal canthal dermoids [7].

2.4 Pathogenesis

The precise developmental mechanisms involved in the pathogenesis of ocular dermoids are not known [7]. Metaplasia of mesenchyme (of primarily neural crest origin), resulting in abnormal differentiation of the surface ectoderm, is considered the most likely mechanism [29]. The resulting dermoid consists of ectodermal elements (keratinized epithelium, hairs, sebaceous and apocrine glands), and mesenchymal elements (fibrous tissue, fat and cartilage) combined in different proportions [30].

Dermoid cysts are formed due to defective epidermal closure along embryonic fissures, which isolates an island of ectoderm in the dermis or subcutis. The cyst usually contains hair, keratin, and sebum, and these materials may produce progressive enlargement of the structure so that it becomes clinically apparent [31]. Dermoid cysts are composed of keratinized stratified squamous epithelium with dermal appendages and adnexal structures, including hair follicles, sebaceous glands, sweat glands, smooth muscle, and fibroadipose tissue. The lumen contains keratin and hair [32]. Cysts that are only lined with epithelium without adnexal elements are termed epidermoid cysts [33].

2.5 Diagnosis

2.5.1 History

History of adverse environmental factors associated with these cases asked from the owner can be helpful. Factors like breed, sex, degree of ocular involvement, age of parents, geographic region, season, type of pasture, soil type, exposure to or suspected exposure to teratogenic plants, feeding and management practices, breeding records, maternal medical and vaccination records, disease status of the herd, periods of stress, drugs administered, congenital defects observed previously, and history of similar congenital defects in neighboring herds should be investigated. Breeding records of defective calves should be collected [33].

2.5.2 Physical examination
History of adverse environmental factors associated with these cases was asked from the owner. The following factors were investigated: breed, sex, degree of ocular involvement, age of parents, geographic region, season, type of pasture, soil type, exposure to or suspected exposure to teratogenic plants, feeding and management practices, breeding records, maternal medical and vaccination records, disease status of the herd, periods of stress, drugs administered, congenital defects observed previously, and history of similar congenital defects in neighboring herds. Breeding records of defective calves were collected [34].

2.5.3 Clinical diagnosis

The corneal dermoids extended slightly beyond the inferonasal limbus and then merged with a second mass of lightly haired tissue within the inferonasal bulbar conjunctiva of both eyes. The nictitans of both eyes was poorly developed, the left nictitans being particularly rudimentary. Signs like Ptosis Proptosis Restriction in extraocular movements Inflammation can be seen during clinical examination [34].

2.5.4 Histology

Histologically, dermoid cysts/sinuses are lined with stratified epithelium resembling normal skin with adnexa and filled with keratinous material [22]. Dermoid cysts are similar histologically, in that they are well-circumscribed cystic structures that lie within the dermis. Dermoid cysts are lined by a keratinising stratified squamous epithelium [12]. The squamous lining of a dermoid cyst shows the formation of some adnexal structures [11]. Numerous fully mature sebaceous glands, with or without other dermal appendages, are widely scattered in the connective tissue layer of the cyst wall. The presence of sebaceous glands is an indispensable histological element with which to make a diagnosis of dermoid cyst [16]. The contents of dermoid cysts are fat with fluid, and hair ball [35]. Soft tissue dermoid cysts usually contain sebaceous glands, hair follicles, hairs, eccrine glands and, in about 20% of cases, apocrine glands [16]. The cyst occurs most commonly in the horse, especially on the dorsal midline, between the withers and the rump (36, 37). Other common locations of these cysts in the horse are the base of the ear, as dentigerous cysts, or in the false nostril, as atheromas [11]. There is one case reported of retrobulbar dermoid cyst in an Andalusian horse [22]. Variation within and among the numerous
dermoids observed histologically and included epidermal thickness, degree of epidermal keratinization and pigmentation, dermal papillae, degree of subacute subepidermal inflammation, number of epidermal adnexa, dermal thickness, amount of adipose tissue in subcutis, and depth of corneal replacement.

2.6 Differential diagnosis

Medial lesions may be confused with congenital encephaloceles, dacryoceles, and mucoceles [33]. Differential diagnosis of lateral lesions includes lacrimal tumors [34].

3. Management

Depending on the type of the eye tumour, its location, and the equipment available, the tumours may be removed surgically, or by cryotherapy, hyperthermia, or radiofrequency. A combination of these modalities can also be used [38]. Some surgical procedures include eyelid wedge resection, third eyelid resection, enucleation, evisceration, or exenteration of the entire globe and orbital contents [38]. Although the implications of spillage of the dermoid cyst content are controversial, the standard practice is to avoid the spillage of the cystic content [10]. Moreover, malignant transformation of the dermoid cysts that reported in the humans [39, 40] was of practical importance in considering the ocular exenteration to avoid extra-morbid second operation to the calf.

3.2 General treatment

Small, asymptomatic cysts may not require treatment. They may stabilize or even decrease in size over years [32]. However, some surgeons opt for early excision to avoid the risk of traumatic rupture in the future [33].

3.3 Surgery

The mainstay of treatment is surgical. Superficial keratectomy is required to surgically excise a corneal dermoid although the depth of the dermoid within the cornea cannot be ascertained by ophthalmic examination until surgery is undertaken [30]. In the case of large corneal dermoids, surgical excision should be performed early in the life of the patient to achieve optical improvement and allow functional development of the eye.
The animal’s eyelashes will be trimmed, and then the eye washed with normal saline solution to remove the contaminants and then dried with sterilized gauge. Lignocaine hydrochloride will be infiltrated in upper and lower eyelids after controlling the animal in lateral recumbency and/or auriculopalpebral, retrobulbar nerve blocks can be used. Eye speculum will be used for proper exposure of operative field. The dermoid will be grasped with allis tissue forceps and the chromic catgut no.1/0 used for ligation and suturing of stamp of dermoid mass. Cyst will be peeled off along with the dermal layers containing the follicles and bleeding will be controlled by instillation of adrenaline solution. The same procedure will be carried out with another eye if the problem is bilateral. Eye will be flushed with NSS solution 2-3 times until blood clot removed from the eye and temporary tarsorrhaphy was performed. Post-operatively, antibiotics like gentamicin and cortisone eye drop will be intilled in both the eyes followed by systemic administration of injectable antibiotic for 5 days.

3.4 Complications

If the cyst ruptures during surgery, a lipogranulomatous inflammatory reaction may occur. This can be mitigated by copious irrigation at the time of surgery. Cysts extending through bony sutures often cannot be removed without rupture.\[\text{[2]}\] If incompletely removed, cysts may recur or lead to abscess formation. They inflammation from remaining dermoid tissue may also result in an orbitocutaneous fistula.\[\text{[3][4]}\]

4. Conclusion

The effect of congenital cysts on the life of affected animals can be considerable. Congenital cysts in calves, although rare, are of potential importance for future research particularly with the advent of sophisticated tools permitting more precise study of gene and chromosomal aberrations. The introduction of new techniques on the chromosome and gene level may be helpful to unravel the pathogenesis of congenital tumours.

5. References


