An uncommon case of anterior segment dysgenesis in a domestic shorthair cat

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SUMMARY

A case of anterior segment dysgenesis in an 18-month-old, one-eyed, domestic shorthair cat is reported. This congenital anomaly presented with a central corneal coloboma, a microspheric lens with a partial cataract, pupillary membrane remnants and goniodysgenesis. Treatment consisted of lens removal by phacoemulsification and a corneal graft using a porcine small intestinal submucosa biomaterial (Vet BioSISt®) to repair the cornea and preserve the vision. Intraocular pressure remained within the normal range.

Features of this case warrant classification as Peters’ anomaly, first in humans in 1887 and which consists of defective development of the posterior cornea caused by abnormal migration and differentiation of neural crest cells impeded by the late separation of the lens vesicle from the surface ectoderm.

Keywords: Anterior segment dysgenesis, cat, coloboma, Peters’ anomaly, biomaterial graft.

Clinical history

The 5 kg castrated male domestic shorthair cat was referred by a colleague, as an emergency, for a suspected anterior luxation of the left lens associated with a corneal ulcer. The cat had undergone an enucleation of the right eye at 8 months of age for an unknown reason; the condition was described by the owners as similar to the one now affecting the left eye.

The cat was in good general condition and had no previous history of ocular trauma. Vision appeared normal. No sign of ocular pain could be detected and there was no periorbital inflammation. Pupillary light reflex was present. The globe was of normal size. Intraocular pressure, measured using a rebound tonometer (Tonovet®, Icare Finland Oy, Espoo, Finland), was 19 mmHg. Examination of the cornea and anterior chamber using a biomicroscope (SL15®, Kowa Europe GmbH, Düsseldorf, Germany), revealed no signs of inflammation or neovascularisation. The cornea was of normal size and convexity.  A full thickness grey opacity 6 mm in diameter was present in the paracentral region surrounding a small central ectatic opalescent zone, with the epithelium covering the cortex of the lens. Punctuate

Revue Méd. Vét., 2010, 161, 4, 173-177
fluorescein staining in the most central zone without extension into the adjacent tissues was observed.

The anterior chamber was of normal depth. The spherical lens, identified in the anterior chamber, was of small size. Multifocal opacities were observed in the anterior cortex and nucleus. The lens was centred on the optic axis with corneal adhesions at the zones of modified transparency, but did not contact the iris. Strands of residual pupillary membrane were present on its surface. The pupil was normal in shape and very mobile without any detectable synechia (Fig. 1).

Because of the increased risk of corneal perforation in its central zone, a corneal graft using a porcine small intestinal submucosa biomaterial (Vet BioSISt®, Cook Inc., Hertfordshire, UK) combined with partial lens removal was performed. The aim was to reconstruct the cornea whilst preserving its transparency and conserving vision. The pre-anæsthetic blood test (complete blood count, serum biochemistry and serum protein electrophoresis) showed no abnormalities. Serological and antiviral assays (Speed-Tests® FELV-FIV-PIF, BVT-Virbac, La Seyne sur Mer, France) for feline immunodeficiency virus (FIV), feline infectious peritonitis (FIP) and feline leukaemia virus (FeLV) were negative, as was the bacteriological examination performed on a sample from the central zone of the cornea. Following a 4-day-course of local antibiotics (ofloxacin ointment, Exocine®, Laboratoire Allergan France SAS), surgery was performed under isoflurane gaseous anaesthetic using an operating microscope (Zeiss OPMI 6, Carl Zeiss S.A.S, Le Pecq, France).

Immediately prior to surgery, the anterior segment had a significantly different appearance from that of the initial examination 4 days previously. The lens had lost its spherical appearance and appeared crumpled. There was an axial corneal defect with the base comprised of the anterior capsule of the adherent lens. The depth of the anterior chamber remained unchanged, with the lens remaining adhered to the posterior corneal surface in the intermediate and paracentral zones (Fig. 2).

A lateral canthotomy was performed. The lens fragments were extracted by phacoemulsification (Universal II; Laboratoires Alcon S.A., Rueil- Malmaison, France) following a keratotomy 1 cm from the central zone of the cornea directed in such a way as to penetrate directly into the lens. An ocular ultrasound probe (Atiop®, Alcon France S.A) was used to aspirate and remove any part of the lens capsule which was not adhered to the cornea. The anterior lens capsule was left in place with the exception of the perforated area. A high-density viscoelastic product (Healon GV®, Amo, Mougins, France) was used to fill the anterior chamber. The corneal epithelium was then removed beyond the limit of the 6 mm diameter paracentral zone. To prepare the site for the biomaterial graft used to reconstruct the cornea, the underlying tissue and the exposed superficial layer of normal stroma of the intermediate zone were excised by lamellar keratectomy.

Two corneal grafts derived from porcine small intestinal submucosa (Vet BioSISt, Cook Inc., Hertfordshire, UK) were then superimposed using simple interrupted absorbable sutures (Vicryl® 10/0, Ethicon, Janssen, Noisy-le-Grand, France). The first graft, of 6 mm diameter, was sutured onto the exposed superficial layer of normal stroma of the intermediate zone. To prepare the site for the biomaterial graft used to reconstruct the cornea, the underlying tissue and the exposed superficial layer of normal stroma of the intermediate zone were excised by lamellar keratectomy.

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Reconstruction of the lateral canthus of the eyelid was performed and the nictitating membrane was sutured to the lateral aspect of the upper lid. Finally, a blepharorrhaphy was performed for additional protection. Ten mg of marbofloxacin (Marbocyl®, Vetoquinol, Lures, France) and 1.5 mg of meloxicam (Metacam®, Boehringer Ingelheim, Paris, France) were injected intravenously and subcutaneously respectively.

An Elizabethan collar was fitted and the antibiotic was continued as a 10-day oral course (marbofloxacin at 2
mg/kg/day). After 20 days’ isolation at the owners’ home, the sutures were removed from the eyelids and nictitating membrane. The anterior chamber was formed, and the biomaterial corneal grafts were incorporated in a zone of corneal granulation. No anterior synechia were present. An indometacin-based anti-inflammatory treatment (Indocollyre 0.1%; Bausch & Lomb, Lab. Chauvin, Montpellier, France), and a vitamin A ointment to promote healing (Faure®, Laboratoires Europhtha, Monaco) were prescribed for 1 month.

At the end of this period, the eye was visually functional. The cornea presented with a small central opacity, which was relatively transparent and only slightly vascularised (Fig. 5).

**Discussion**

The congenital abnormalities of this eye present similarities with anterior segment dysgeneses described in both animals and humans (corneal opacity, remnants of persistent pupillary membrane, malformation of the iridocorneal angle, ectopic microspherophakia). [2,3,5,7-10,13,15-19,21,22] The corneal coloboma and anterior chamber lesions were non-inflammatory. [2,3,9,16,17] The central corneal perforation was not of traumatic or infectious origin. [2,9,16,17] The presence of microspherophakia adherent to the defective cornea could not be confused with the well-documented lens luxation of...
familial and congenital origin in the Siamese cat, [3,12,14] which is most commonly inflammatory in adult cats, and degenerative in elderly cats. [4,11-13,15,19,24] This case is similar to keratolenticular dysgenesis described by PEIFFER in 1983. In that case, a Persian kitten presented with a lens, containing a dumbbell-shaped partial cataract, embedded in a central keratocornea. Histology showed that the Descemet’s membrane and anterior capsule of the lens were absent, causing the corneal stroma and the lens cortex to be in close contact.[16] A similar corneal coloboma was described in a newborn infant affected by a Peter’s anomaly, heart malformation and hydrocephalus in 1996. [9]

In many species of animal, a persistent pupillary membrane is the most common manifestation of anterior segment dysgenesis. [4,13,17,21] More rarely, clinical signs associate, in varying degrees, anatomical defects affecting the optic axis. These can be found at the level of the corneal stroma and endothelium, the iris, and the anterior aspect of the lens. In all probability, this is the result of a defect in the separation between the surface ectoderm and the lens vesicle. [3,8,9,16,18]

This anterior segment dysgenesis presents many similarities with Peters’ anomaly in humans. It is an irido-corneo-trabecular dysgenesis, which is normally bilateral, but can be unilateral and asymmetric. [2,5,10,14] It presents as a central corneal opacity caused by the absence of the endothelium and Descemet’s membrane. Iridocorneal adhesions arise from the iris collarette attaching to the periphery of the endothelial deficit. Lesions of the iris can also be associated with this, as well as defects of the iridocorneal angle occasionally associated with a primary glaucoma of more or less early onset. [2,9,10,20] Malformations of the face, heart, genito-urinary tract, central nervous system spinal cord, and retarded growth can equally coexist and have a hereditary nature often. [2,5,9,10,20] The association of a palpebral agenesis and an ocular anterior segment dysgenesis has been described in a domestic cat. The genetic origin of these disorders has not been proven in this species. [13,19]

In human ophthalmology, dysgeneses of the ocular anterior segment are classified into either ocular anomalies caused by defective migration or differentiation of cells of the cephalic neural crest, or more complex forms of dysgenesis, in which the induction mechanisms of the lens, of ectodermal origin, are paramount. [10]

In the case under discussion, the abnormal conformation of the 2 central zones of the cornea may have resulted from an anomaly of embryogenesis of the corneal stroma, the origin of which cannot be determined. The same applies to the defective migration of the lens. Only histological examination of the anterior segment would allow this hypothesis to be tested, but this would have been incompatible with our aim of preserving a functioning eyeball.

In the cat, glaucoma is known to be principally secondary to uveitis, lens luxation or neoplasms of the anterior segment. [11,19,24] Primary glaucoma is rare. [7,11-13,15,17,19,22] In a histological study of 131 cases, WILCOX et al. [24] found only 3 cases of goniodysgenesis responsible for ocular hypertension. A breed disposition to primary glaucoma has been described in the Siamese and the Persian (pectinate ligament dysplasia) and the Burmese (closed angle). [19] No publications have mentioned the angle anomalies described in this clinical case. Today, the cat is 3 years older and has shown no signs of increase in ocular pressure to date.

Phacoemulsification was chosen to excise the lens due to the adhesions between the lens and the cornea. The keratotomy was performed on healthy cornea whilst taking the lens position into account so as to allow easy extraction of the lens fragments. The evacuation of the irrigation fluid was performed at the same time by means of a hand piece, via the perforation in the cornea, thus sparing the anterior chamber from ultrasound waves and debris from the lens.

The preservation of the contact zone between the anterior lens capsule and the cornea supported the application of the biomaterial corneal graft, using a previously published technique. [1,6,23] The superimposition of layers allowed a good quality seal to be obtained. Due to the cat’s lively nature, the operated zone was protected by suturing the nictitating membrane to the upper eyelid, followed by a supplementary blefarorrhaphy.

The central scar opacity had an undeniable transparency, unlike the leucomas observed by BUSSIERES and colleagues [1] in 2 cats with perforated corneal ulcers treated by excision of necrotic tissue and the application of a porcine small intestinal submucosa graft. A pedunculated conjunctival graft had been used to protect the operation site and could explain the appearance of a very opaque central leucoma. In the case under discussion, the biomaterial graft was replaced by tissue which re-covered the area with a functioning transparency, and was sufficient to preserve the vision of this one-eyed cat.

**Conclusion**

Anterior segment dysgeneses are well known in humans. Their pathology is well understood and their origin has been explained using animal models. Their mode of transmission and the genetic mutations responsible for these conditions have been the subject of much research. In more than 70% of cases, they are associated with congenital glaucoma, which carries a poor prognosis for vision. This clinical case closely resembles Peters’ anomaly, first described in 1887. As far as the author is aware, this is the first case describing an anterior segment dysgenesis with corneal perforation that was successfully treated with a biomaterial graft in the cat.

**References**


