

Ehlers-Danlos syndrome in a dog

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Ehlers-Danlos syndrome, also called cutaneous asthenia, is a rare connective tissue disease reported in humans, dogs, cats, mink, cattle, and sheep. The skin is hyperextensible and may also be very thin and fragile with greatly reduced tensile strength. The hair coat is often dry and fine (1). The disease complex may include joint laxity and ocular abnormalities (2). Histologically, the dermis is thinner than normal, with fragmentation and alterations in size and orientation of collagen bundles. There is a decrease in the number of collagen fibers (3). The disease is hereditary and appears to be autosomal dominant (1,3,4).

This case involved a male dog of mixed breeding from a litter of five; it had the general appearance of a spaniel cross with a medium brown hair coat of very fine texture. The dam was reported to be normal. The status of the father and of the four littermates is unknown. The pup was clinically normal at the time of the puppy vaccination visits. At four months of age, slight cloudiness of the corneas was noted. An episode of otitis externa and one of tonsillitis occurred during the first year of life.

At eleven months of age, mild lameness of the right hindlimb was diagnosed as due to a medial patellar luxation. The right patella could be luxated easily with digital pressure but would return to the trochlear groove when pressure was released. The left patella felt loose but could not be luxated. Both hock joints could easily be overextended; however, this joint laxity did not interfere with walking nor did the hindleg conformation appear abnormal while the animal was standing. All other joints were normal as measured by palpation. The lameness was treated conservatively with rest, and did not recur.

The dog was referred for ophthalmic consultation for the ocular abnormality at eleven months of age. Subjective visual function testing and pupillary light reflexes were normal, though the dog appeared mildly photophobic. Each globe was assessed to be of normal size. However, the diameter of clear cornea was small (microcornea) and the corneal-scleral limbus was indistinct (sclerocornea). A fine, granular opacity in the rostral corneal stroma was visualized with the aid

of the biomicroscope. There was absence of corneal vascularization or cellular infiltrates. Both gonioscopic and indirect ophthalmoscopic examinations were normal. No treatment was prescribed.

Between 12 and 35 months of age, five cutaneous lacerations occurred. These included two separate episodes caused by a dog groomer teasing mats from behind the ears. Two other wounds, one on the lateral tarsus and another on the paw of a front leg, occurred indoors and were of unknown cause. In each case there was very little bleeding and the wounds tended to gape. The skin appeared very thin and was extremely friable. Horizontal mattress sutures pulled through the skin less easily than did simple interrupted sutures. The most recent laceration was to the bridge of the nose and was repaired with a tissue adhesive (Vetbond, Animal Care Products/3M, St. Paul, Minnesota, USA). The wound margins held together very well and healing occurred uneventfully. Of the methods used to repair the skin, the tissue adhesive seemed to work best.

An important clinical sign of Ehlers-Danlos syndrome is cutaneous hypersensitivity. The extensibility index is used to quantitate the extensibility of the skin and is the ratio of the vertical height of the dorsolumbar skin, when traction is applied, compared to the body length from occipital crest to the base of the tail. Normal values are 8–15% while those of affected animals are 17–25% (4). Cutaneous hyperextensibility was present in this case (Figure 1).

Full thickness skin biopsies were collected from the lateral chest wall at the level of the mid-shaft of the eighth rib. Histological examination revealed a very thin epidermis of one to two cell layers thick. The superficial and deep dermis were also very thin and pale-staining due to thin and haphazardly arranged collagen fibers. There were no inflammatory changes. The pathological diagnosis was severe epidermal atrophy and dermal collagen atrophy with probable collagen dysplasia. The clinical picture and the histopathological findings were consistent with Ehlers-Danlos syndrome.

It is very unusual to see the complete syndrome of cutaneous fragility, articular laxity, and ocular abnormalities in the same animal. The cutaneous form is most commonly seen. Some animals may exhibit only hyperextensibility or fragility although most exhibit both (2). A constellation of possible ophthalmic manifestations including blue (thin) sclera, strabismus, microcornea, keratoconus, lens subluxation, severe

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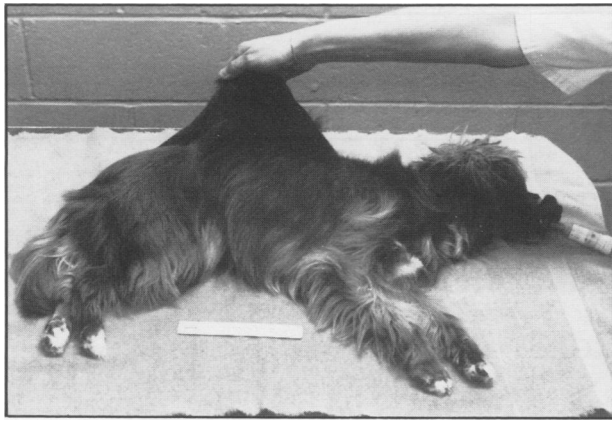


Figure 1. The skin could be stretched very easily and had an extensibility index of 28% measured while the dog was anesthetized.

myopia, angioid streaks (breaks in Bruch's membrane), and retinal detachments have been described in human Ehlers-Danlos patients (5). There are two published reports of Ehlers-Danlos in dogs with ocular manifestations. Barnett and Cottrell (3) described bilateral lens subluxations, cataract, corneal edema, and poor pupillary light reflexes. The corneal opacity observed in our case was presumed to be caused by altered arrangement of collagen fibrils or by interfibrillar deposits. Histological examination with appropriate histochemical staining would be required to define this dystrophy. In the case reported by Anderson and Brown (6), only brief mention was made of bilateral corneal opacity. Sclerocornea, an unreported finding in the dog, is thought to arise from a disturbance of mesenchymal and surface ectodermal growth at the rim of the embryological optic cup (7). Sclerocornea has been described in people in association with congenital ocular defects as well as with nonocular con-

genital abnormalities, such as polydactyly. It appears that, much as in people, it is rare to have the full complement of ocular manifestations of Ehlers-Danlos syndrome in a single patient.

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