Diaphragmatic and perineal hernias associated with cutaneous asthenia in a cat

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Cutaneous asthenia is a rare hereditary disorder that results in altered strength or extensibility of connective tissues.

The most common lesions in affected animals are cutaneous fragility and hyperextensibility.

The diagnosis depends on clinical findings (skin extensibility index) and light and electron microscopic evaluation of affected tissue.

Surgical repair of hernias can be performed successfully on an affected cat, and healing of the incision can occur without complications.

An 11-year-old neutered male domestic shorthair cat weighing 5 kg (11 lb) was examined for an acute episode of dyspnea. The cat had a 1-month history of coughing when it was handled and had been medically treated for a left-sided reducible perineal hernia for 3 months with a high-fiber diet and stool softeners. Additionally, hyperextensibility of the cat’s skin observed since 9 months of age had resulted in a presumptive diagnosis of cutaneous asthenia (similar to Ehlers-Danlos syndrome [EDS] in humans). At 10 months of age, the cat created 2 lacerations on the caudal aspect of its head after scratching with its hind limb, and at that time, onychectomy of all digits was performed to prevent further self-trauma.

Evaluation of thoracic and abdominal radiographs revealed gas-filled intestinal loops within the thorax, consistent with a diaphragmatic hernia. Rectal temperature was 38.5°C (101.3°F), pulse was approximately 180 beats/min, and respiration rate was 60 breaths/min. Examination of the cat occasionally resulted in tachypnea. Borborygmus was auscultated in the thorax, although heart sounds could not be discerned. Intestinal loops were not detected via abdominal palpation. A perineal swelling lateral to the left side of the anus was confirmed to be a left-sided reducible perineal hernia via rectal palpation; the right side of the pelvic diaphragm was intact.

The skin on the ventral aspect of the thorax and abdomen was pendulous. There was marked hyperelasticity of the skin in the thoracic and lumbar regions (Fig 1). An extensibility index [(vertical height of skin fold divided by body length from the occipital crest to the tail base) × 100; reference limit, <19%] of 25% was determined to be consistent with dermatosparaxis.

Surgical repair of the diaphragmatic hernia was recommended. The cat was premedicated by administration of morphine (0.11 mg/kg [0.05 mg/lb], IV), atropine (0.02 mg/kg [0.01 mg/lb], IV), and medetomidine (2.2 µg/kg [1.0 µg/lb], IV). Anesthetic induction was achieved by administration of thiopental (6.6 mg/kg [3 mg/lb], IV to effect). Isoflurane and oxygen in a circle breathing circuit were used to maintain anesthesia. Cefazolin (22 mg/kg [10 mg/lb], IV) was administered after induction of anesthesia, prior to the surgical procedure, and repeated every 90 minutes until completion of surgery.

A standard ventral midline celiotomy was performed. The falciform ligament was ligated with 3-0 polydioxanone and transected distally with Metzenbaum scissors. Observations in the abdominal cavity included a large rent in the right dorsal aspect of the diaphragm extending from the dorsal aspect of the right costal part to its mid body at the level of the caval foramen. The right lateral lobe, right medial lobe, and caudate process of the caudate lobe of the liver, as well as the stomach, duodenum, jejunum, spleen, and mesentery, were herniated into the right hemithorax. Adhesions and pleural effusion were not detected at the time of surgery. Moderate atelectasis was seen in the right lung. Reduction of the abdominal contents was achieved with digital traction. Prior to closure of the diaphragmatic rent, an open-ended, 8-F red rubber feeding tube was placed percutaneously at the level of the 12th rib and positioned in the thoracic cavity at the ninth intercostal space. After visual placement of the tube, the edges of the diaphragmatic hernia were apposed with a simple continuous pattern of 2-0 polypropylene. Biopsy specimens were obtained from the tail base [fold divided by body length from the occipital crest to the tail base] × 100; reference limit, <19%] of 25% was
the epidermis prior to closure and submitted for histopathologic and transmission electron microscopic examination. The linea alba and subcutaneous tissues were individually closed with 3-0 polydioxanone in a simple interrupted pattern. The epidermis was sutured with 3-0 nylon in a simple interrupted pattern. A 3-way stopcock with sterile medical-grade injection ports was placed on the thoracic tube, and a soft-padded bandage was applied to the thorax.

The cat was placed in intensive care for care of the thoracic tube and administration of hydromorphone (0.22 mg/kg [0.10 mg/lb], IV, q 6 h) for pain. The thoracic tube was removed 24 hours after surgery because there was minimal fluid accumulation (< 2 mL/kg/d [0.91 mL/lb/d]) and no accumulation of air in the thoracic cavity. The cat was released from the hospital 3 days after surgery. Lactulose (0.4 mL/kg [0.18 mL/lb], PO, q 12 h) and cisapride (1 mg/kg [0.45 mg/lb], PO, q 12 h) were prescribed for medical management of the perineal hernia. The surgical incision healed without complications, and sutures were removed 22 days after surgery.

Histopathologic examination of the skin revealed normal epidermis and slightly thick dermis that contained high numbers of small collagen bundles, compared with a site-matched specimen from a control cat. No collagen abnormalities were detected with Masson trichrome stain. Additionally, Verhoeff and acid orcein Giemsa stains revealed no abnormalities in the elastin fibers. Electron microscopic findings of the skin were consistent with cutaneous asthenia. The diameter of the collagen fibers was highly variable, compared with a site-matched sample from a control cat.

Four weeks later, the cat was admitted for surgical repair of the left perineal hernia. The cat was constipated despite treatment prescribed previously. The cat received premedication, anesthesia, and cefazolin as administered previously. A standard perineal herniorrhaphy was performed on the left perineum.

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Postoperatively, a grade 2/6 systolic heart murmur was detected. Abnormalities determined by use of echocardiography included systolic anterior motion of the mitral valve, a dilated aortic valve annulus in the proximal portion of the aorta (13.1 mm; reference range, 9.0 to 10.0 mm), and mild ventricular hypertrophy. Serum thyroxine concentration was unremarkable (26.8 nmol/L; reference range, 17.0 to 49.0 nmol/L), which ruled out hyperthyroidism as the cause for ventricular hypertrophy.

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Surgical repair consisted of a standard perineal herniorrhaphy with elevation of the internal obturator muscle.

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Mus were present prior to the hernia, but chronic constipation. It is possible that constipation and tenesmus were present prior to the hernia, but chronic constipation was not reported.

Although humans with EDS may have increased risk of surgical complications (fragile blood vessels and soft tissues, delayed wound healing, wound dehiscence, and wide scar formation), the cat reported here did not develop any complications during or after surgical repair of the hernias. Only a few studies have evaluated wound healing in animals with cutaneous asthenia. Freeman et al reported that there was no important difference in primary intention healing between dogs and cats with cutaneous asthenia (resembling EDS type I) and clinically normal control animals. Because there are clinical differences in skin fragility among the various syndromes of EDS, it is plausible that differences in wound healing are also associated with the various types of molecular or biochemical abnormalities. Delayed wound healing should be considered in animals with cutaneous asthenia, and appropriate decisions on suture type, suture pattern, and duration of application should be made.

Human EDS types I, II, IV, and V can be associated with cardiovascular abnormalities such as valvular prolapse, pulmonary stenosis, mitral regurgitation, aortic aneurysms, atrial septal defect, and tetralogy of Fallot. In the cat reported here, aortic dilatation may have been associated with cutaneous asthenia. Aortic dilatation is an uncommon finding in cats that is sometimes associated with hypertension. Hypertension in this cat cannot be ruled out because blood pressure was not measured prior to administration of anesthetic agents.

A presumptive diagnosis of cutaneous asthenia is often made on the basis of skin hyperextensibility and fragility. A definitive diagnosis of cutaneous asthenia requires histopathologic, electron microscopic, and biochemical studies of the skin. The cat reported here had an elasticity index of 25%, which is consistent with cutaneous asthenia. It is not unusual for animals with cutaneous asthenia to have subtle or absent light microscopic changes, as was seen in this cat. Of the 2 types of cutaneous asthenia described in cats, the electron microscopic findings in the cat reported here most closely resembled the autosomal dominant form (human EDS types I and II), which often has a bimodal distribution of small and large collagen fibers. The presence of 2 fiber sizes may represent a heterozygous state in which normal and altered collagen metabolisms exist. Differential diagnoses for cutaneous asthenia include acquired skin fragility and cutis laxa. Feline acquired skin fragility is a relatively rare disorder with several etiologies (eg, spontaneous hyperadrenocorticism, diabetes mellitus, hepatic lipidosis, and administration of corticosteroids and progestins). Acquired skin fragility was ruled out in this cat because the cat was affected at a very young age, did not receive causative medications, and a concurrent systemic illness was not identified.

Cutis laxa (previously classified as EDS type IX) has not been reported in cats. Cutis laxa was ruled out in this cat because serum copper concentration was within reference range and elastin abnormalities were not detected.
References


