Dermatological manifestations of systemic disease are now becoming well-known in the dog, although their pathogenesis is not always understood. They are very diverse clinically and are associated with various systemic illnesses. It is important to know about these manifestations as they enter into the differential diagnosis of many different conditions and can appear months or even years before the underlying illness; some are paraneoplastic syndromes.

Nodular dermatofibrosis

Aetiopathogenesis

Nodular dermatofibrosis (ND) is a syndrome involving both multiple cutaneous nodules and bilateral renal cystadenomata (adenocarcinomata). It was first reported in the German Shepherd Dog in Switzerland but has since been reported regularly throughout Europe, the United States and Australia, in the German Shepherd Dog and also the Golden Retriever. In the German Shepherd Dog, genetic studies have demonstrated an autosomal dominant mode of transmission. ND is seen mostly in adult dogs of either sex. Nodules may be present for several months or even years (3-5 years) before the condition is diagnosed.

The link between cutaneous nodules and renal cystadenomata/cystadenocarcinomata is not understood. Although in most cases, the skin nodules appear before the renal lesions, it is difficult to know whether both skin and renal lesions develop at the same time or if one precedes the other. It is possible that skin nodules and renal cystadenomata/cystadenocarcinomata occur independently but via the same hereditary mechanism.

A genetic study of a population of German Shepherd Dogs with ND has recently identified a region of chromosome that may be responsible for this condition. Another theory is that secretion of various growth factors (TGF\(_a\), TGF\(_b\)) by the renal tumours may promote collagen synthesis and consequently a paraneoplastic syndrome.

Clinical signs

**Dermatological signs** include papules and nodules on the limbs (carpi, tarsi, metacarpi, metatarsi, digits and footpads), head and lips and less commonly the trunk. Nodules, which may be dermal or subcutaneous, are 0.5-5 cm in diameter, firm, non-painful, non-moveable and sometimes alopecic, hyperpigmented and ulcerated. They vary in number between 10 and over 50. Some may coalesce to form plaques. In some locations such as digits and footpads, these nodules be painful and cause lameness.

**Systemic signs**, associated with secondary renal insufficiency, develop later, sometimes years after the nodules. Depression, weight loss, lethargy, dehydration, polyuria/polydipsia and vomiting may be seen. Abdominal palpation may reveal one or two large masses. Cystadenocarcinomata may metastasise to lymph nodes and lungs.

Diagnosis

Diagnosis is based on dermatological and possibly systemic signs, principally in the German Shepherd Dog, suggestive skin biopsies and renal ultrasound.

**Histopathological examination of intact nodules** reveals proliferation of collagen fibres in the dermis. Recent lesions contain areas of active fibroplasia and quiescent, well-differentiated fibroblasts whereas older lesions contain normal collagen fibres. These histopathological lesions are diagnostic for collagen naevus.
**Histopathological examination of renal lesions** reveals proliferation of renal tubular epithelium (leading to obstruction), proximal tubular dilatation and cyst formation. Epithelial hyperplasia becomes adenomatous and subsequently adenocarcinomatous.

**Renal ultrasound** reveals the presence of anechoic and hypoechoic structures (cysts). They are large, usually bilateral, sometimes multilobular and lead to considerable reduction in renal parenchyma.

**Chest radiography and abdominal ultrasound** must be carried out to look for possible metastases.

**Various renal parameters** should also be obtained.

**Prognosis and treatment**

**Prognosis** is guarded although ND develops slowly over several years. The prognosis depends on the time it takes for signs associated with the renal adenocarcinomata to develop.

**Treatment** is non-existent. Badly-placed, ulcerated nodules can be resected without recurrence. Benefits of treatment for adenocarcinomata are temporary, given the bilateral distribution and the risk of metastasis. Surgical removal of a kidney about to rupture may be undertaken in the absence of renal insufficiency.

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**Necrolytic migratory erythema**

**Aetiopathogenesis**

Necrolytic migratory erythema (NME) is a rare, ulcerative, crusting disorder of mucocutaneous junctions and pressure points which precedes or accompanies the onset of chronic hepatic disease or, more rarely, a glucagonoma. The condition has been given many names including superficial necrolytic dermatitis, diabetic dermatopathy, hepaticotaneous syndrome, metabolic epidermal necrosis, metabolic necrolytic dermatitis and glucagonoma syndrome. It affects mainly dogs over 10 years old. A slight breed disposition has been reported in the Jack Russell terrier.

Unlike in man, almost all cases of NME reported in the dog have been associated with chronic hepatic disease (e.g. cirrhosis, drug-induced hepatitis (phenobarbital) and chronic active hepatitis). A few cases have been linked to a pancreatic, glucagon-secreting tumour (glucagonoma).

Aetiopathogenesis is still uncertain in the dog. As in man, it is thought that hypoaminoacidaemia may cause epidermal protein depletion and keratinocyte necrolysis. Defective zinc and essential fatty acid metabolism may also be involved.

**Clinical signs**

**Dermatological signs** usually precede systemic signs. Erythema, erosions, ulcers and crusts are seen at mucocutaneous junctions (e.g. lips, eyelids, nose, anus and genital organs) and pressure points (elbows, feet, abdomen and axillae). Footpad thickening and fissuring are strongly suggestive. Lesions are often painful and secondary bacterial and fungal infections are common.

**Systemic signs** are non-specific and occur later. Depending on the underlying condition, they include poor general condition, weight loss, lethargy, anorexia, pyrexia, jaundice, polyuria, polydipsia and polyphagia.

**Diagnosis**

Diagnosis is based on dermatological and sometimes systemic signs, skin biopsies, hepatic and pancreatic ultrasound, ultrasound-guided biopsies, and haematology and biochemistry profiles.

**Differential diagnosis** includes pemphigus foliaceus, leishmaniosis, systemic lupus erythematosus, zinc-responsive dermatosis and various nutritional deficiencies such as generic dog food dermatosis.

**Histopathological examination of skin biopsies** reveals the characteristic epidermal red, white and blue: hyperplasia of the deep epidermal layers, pallor of the superficial malpighian layers in association with interkeratinocytic and intrakeratinocytic oedema (vacuolation), and sometimes marked parakeratotic hyperkeratosis (Fig. 21:16). Keratinocyte vacuolation can cause intra-epidermal clefting. Dermal lesions are non-specific.

21.2: Same animal as that in figure 21.1: large, firm ulcerated nodule.

21.3: Nodular dermatofibrosis in a German Shepherd Dog: plaque composed of numerous coalescing ulcerated nodules on the head (courtesy of J.F. Dandrifosse).


21.5: Skin histopathology (nodular dermatofibrosis): proliferation of collagen fibre bunches (H&E stain) (courtesy of M. Mialot).

21.6: Nodular dermatofibrosis in a German Shepherd Dog: large cyst found on kidney necroscopy.

21.7: Renal histopathology (nodular dermatofibrosis): microcysts (H&E stain) (courtesy of M. Mialot).

21.8: Renal histopathology (nodular dermatofibrosis): renal tubular epithelial hyperplasia (H&E stain) (courtesy of M. Mialot).
Haematology and biochemistry profiles are often abnormal revealing non-regenerative or mildly regenerative anaemia, polykilocytosis, neutrophilia, hypoalbuminaemia, increases in beta- and gamma- globulins, and elevation of hepatic enzymes. Hypoacidaemia involving hydroxyproline, threonine, glutamine, proline, alanine, citrulline and arginine have often been observed. Hypertucagonaemia and hyperinsulinaemia are less common.

Hepatic and pancreatic ultrasound and ultrasound-guided biopsies allow diagnosis of the underlying condition. Hepatic lesions are variable, involving mainly cirrhosis. Focal chronic and subacute pancreatitis lesions are often present even in the absence of glucagon-secreting tumours.

**Prognosis and treatment**

**Prognosis** is very poor even when recurrent, drug-induced hepatitis is involved. There is no specific treatment for NME-associated hepatitis. Nutritional support is required, sometimes with colchicine (0.03 mg/kg/d) and/or oral S-adenosyl-methionine (20 mg/kg/d). Skin lesions may be improved by treating secondary bacterial and fungal infections and by giving amino acid supplements (e.g. 1 egg yolk/5 kg body weight/day; amino acid drip), essential fatty acids, and zinc (e.g. zinc gluconate, 10 mg/kg/d). A glucagonoma with no metastases can be successfully removed. Skin lesions start to resolve a week after surgery and disappear completely within 45 days.

**Hereditary dermatomyositis**

Aetiopathogenesis

Hereditary dermatomyositis (HD) is an uncommon disorder affecting both skin and muscle. HD has been reported only in dog and in man; the canine condition is a good model for infantile dermatomyositis. HD has been reported mainly in Collies and Shetland sheepdogs and in Europe, the Beauceron sheepdog. However, the condition has been reported sporadically in other breeds including the Australian sheepdog, Chow-chow, Labrador retriever, Kuvasz (Hungarian sheepdog), Corgi, and German Shepherd Dog. Although, it usually starts in young animals between the age of 2 and 6 months, it may occur in older dogs. No sex predisposition has been seen.

The mechanism of lesion development remains unknown. Genetic transmission involving an autosomal dominant with variable expression has been demonstrated in the Collie. A marker, FH370 on chromosome 35, has been identified in Shelties with dermatomyositis. However, additional studies are needed to determine the location of the dermatomyositis locus, to identify the responsible genes and to determine the mode of transmission. It is also possible that a viral infection (e.g. coronavirus or picornavirus?) may trigger clinical signs in the dog, as is the case for children and coxsackie B virus. Viral particles have been found in some biopsies taken from affected dogs. The correlation of circulating immune complexes with the severity of clinical signs, the presence of vasculitis in affected muscles and skin, the occurrence of follicular atrophy and follicular and epidermal degeneration, suggest a type III hypersensitivity affecting arterioles. Immune complex vasculitis is probably the cause of tissue necrosis, follicular atrophy and basal keratinocyte degeneration.

Clinical signs

Signs are variable, some animals having only skin lesions, others with only muscular problems. **Dermatological signs** usually start on the face, pinnal tips, tail and bony prominences of the limbs. Erythema, papules, vesicles, pustules, alopecia and degimentation develop simultaneously in many locations. Repeated trauma leads to secondary lesions including excoriations, erosions, ulcers, crusts and atrophic scarring. The condition is usually restricted to precise regions of the body and does not become generalised. **Muscle signs** vary in severity. They typically appear after dermatological signs. Temporal and masseter muscle atrophy, stiff gait, and eating difficulties (associated with megaesophagus) are the main signs. Problems in eating may lead to false deglutition and bronchopneumonia (Fig. 21:28). Lymphadenopathy, transient facial oedema, intermittent polyarthritis, sterility and growth retardation are seen less commonly.
Necrolytic migratory erythema in a Papillon: ulceration and crusting on the eyelids, lips and nose.

Necrolytic migratory erythema in a Malinois sheepdog: compact scaling and crusting and erosions on the dorsal surface of the nasal planum.

Necrolytic migratory erythema in a German Shepherd Dog: thick scaling and crusting and erosions on the hock.

Necrolytic migratory erythema in a Brittany spaniel: early serpiginous lesions and preputial ulceration.

Necrolytic migratory erythema in a Fox terrier: compact scaling and fissuring of the footpads.

Necrolytic migratory erythema in a German Shepherd Dog: compact scaling and ulceration of the footpads.

Necrolytic migratory erythema in a Fox terrier: ulcerative dermatitis with serpiginous borders on the medial thigh.

Skin histopathology (necrolytic migratory erythema): epidermal parakeratotic hyperkeratosis, keratinocyte vacuolation in the malpighian layer, hyperplasia of the deep layers of the epidermis, dermo-epidermal clefting (H&E stain).
Diagnosis
Diagnosis is based on dermatological and muscle signs in animals of predisposed breeds, skin biopsies and possibly muscle biopsies.

Differential diagnosis includes cutaneous lupus, systemic lupus erythematosus, epidermolysis bullosa, dermatitis, dermatophytosis, vasculitis, and other ischaemic dermatopathies.

Histopathological examination of skin biopsies reveals follicular atrophy with short, straight primary follicles and often inapparent secondary follicles, along with moderate to severe dermal fibrosis. Slight mixed dermal inflammation is seen in most biopsies. Vacuolation of epidermal basal cells and follicular cells of the external root sheath leads to dermo-epidermal clefting (Fig. 21:29-31). Numerous colloid bodies can be seen in follicular infundibula and in the epidermal basal layer.

Histopathological examination of muscle biopsies reveals inflammation in affected muscles. Features include multifocal muscle necrosis; fragmented, vacuolated, atrophic, calcified myofibrillae; and an inflammatory infiltrate consisting of lymphocytes, plasma cells, macrophages and neutrophils. Arteriolar vasculitis is sometimes seen (Fig. 21:32).

Haematology often reveals a neutrophilia and a non-regenerative, normocytic, normochromic anaemia in association with chronic inflammation.

Blood biochemistry often demonstrates an increase in creatinine phosphokinase (CPK) in the acute myositis phase. A rise in circulating immune complexes (in line with skin and muscle lesion severity) precedes the appearance of clinical signs.

Electromyography of affected muscles, usually close to skin lesions, can reveal fibrillation potentials, occasional positive sharp waves and bizarre high-frequency discharges.

Prognosis and treatment
The prognosis is variable. HD can remain stable for many months, steadily worsen or even progressively resolve spontaneously. Factors such as stage of the reproductive cycle in females and exposure to the sun, are known to lead to exacerbation. If the dog is unable to eat properly because of a megaoesophagus, it may well be euthanased.

Oral glucocorticoids (prednisolone or prednisone, 1-2 mg/kg/d until remission, then on alternate days) sometimes give good results. They should be given for a short time as possible as prolonged use can aggravate muscle atrophy. Oral pentoxifylline (10 mg/kg TID) is given principally to increase muscle oxygenation but may also improve skin lesions and allow a reduction in the dose of steroid.

Neither affected animals nor other dogs from the same line should be used for breeding.

Calcinosis cutis

Aetiopathogenesis
The term calcinosis cutis covers an extremely broad spectrum of conditions with only one thing in common - pathological calcification in the skin. In dystrophic calcification, phospho-calcium metabolism is unaffected and other conditions, such as hyperadrenocorticism or diabetes mellitus, are responsible for skin calcification. In metastatic calcification, phospho-calcium metabolism is disordered, e.g. renal insufficiency with secondary hyperparathyroidism and possible elevation in serum calcium and/or phosphate. Iatrogenic calcinosis may result from local hypercalcaemia following topical application of a calcium chloride or carbonate product, an injection of calcium gluconate, or an injection of progestogens. Calcinosis may also be idiopathic (e.g. idiopathic calcinosis universalis and calcinosis circumspecta). Only metastatic calcification will be considered here.

Metastatic calcification is rare but can develop when the phospho-calcium balance is over 7000 mg/L (normal level is around 4500). It can occur alongside renal insufficiency (e.g. chronic renal insufficiency (CRI), abnormal urinary tract development (persistent urachus), and renal dysplasia). Secondary hyperparathyroidism caused by reduced active vitamin D production and hyperphosphataemia leads to hypercalcaemia and an increase in phospho-calcium balance. Ectopic calcification can be seen in various organs (e.g. lungs, kidneys and bladder) as well as in the skin.
21.17: Hereditary dermatomyositis in a Sheltie: facial erythema and cicatricial alopecia (courtesy of D.N. Carlotti).

21.18: Hereditary dermatomyositis in a Beauceron sheepdog: alopecia, depigmentation and crusting around the medial canthus.


21.22: Same animal as that in figure 21.19: erythema, alopecia, scaling and crusting on the bridge of the nose and nasal planum.

21.23: Hereditary dermatomyositis in a Beauceron sheepdog: erosions and crusts around the elbow.

Clinical signs
Dermatological signs include papules and firm, cutaneous or subcutaneous nodules, sometimes ulcerated and painful (Fig. 21:33-34). Unlike calcinosis circumscripta, where bony prominences and digits are usually affected, there is no typical distribution pattern, although footpads are perhaps more commonly involved. Several cases of footpad calcification have been reported in small breeds of dog (e.g. Shi-tzu and Pekinese) with renal dysplasia.

Usually, lesions develop after, or at the same time as, signs of renal insufficiency.

Diagnosis
Diagnosis is based on dermatological and renal signs, skin biopsies and tests (e.g. blood biochemistry, urinalysis and urinary tract ultrasound) for the cause of renal insufficiency.

Histopathological examination of skin biopsies reveals large irregular bands of basophilic granular material, staining black with Von Kossa, in the deep dermis and hypodermis. Mineralised foci are surrounded by a cellular crown of macrophages, epithelioid cells and multinucleate giant cells.

Blood biochemistry usually reveals signs of renal insufficiency (uraemia, creatinaemia, hypercalcaemia, hyper-phosphataemia, increase in phospho-calcium balance).

Urinary tract radiography and ultrasound will often demonstrate the cause of renal insufficiency (e.g. renal dysplasia and persistent urachus). Ultrasound-guided biopsies can be taken.

Prognosis and treatment
The prognosis is bleak given the severity of the associated kidney disease.

Treatment is given (in CRI) mainly to limit the development of further lesions by reducing the phospho-calcium balance. Phosphataemia is reduced by using a low phosphorus diet, phosphorus chelators (e.g. aluminium and magnesium hydroxide) and activated charcoal.

Cutaneous amyloidosis

Aetiopathogenesis
Cutaneous or mucocutaneous amyloidosis is rarely seen in the dog.

Amyloidosis is caused by the pathological, extracellular deposition of fibrillar proteins. These proteins differ according to the source of amyloid formation. Some (AL proteins) have an immunoglobulin structure and a light monoclonal chain for a serum precursor. They are associated with primary amyloidosis or amyloidosis associated with myeloma. Other fibrillar proteins (AA proteins) do not have an immunoglobulin structure and are produced by hepatocytes and fibroblasts during inflammation. They are associated with secondary amyloidosis. Amyloid is formed from a physiological serum precursor and its deposition in intercellular spaces can cause abnormalities in the breakdown of this precursor in the macrophage.

Three cases of amyloidosis have been reported. One, associated with monoclonal gammapathy in a Cocker spaniel; one in a Brittany spaniel with no apparent underlying condition but associated with a polyclonal hyperglobulinaemia; and one in a Siberian Husky with subclinical renal amyloidosis.

Clinical signs
Dermatological signs include purpura, skin fragility leading to exposure of an orange dermis on the trunk; interdigital, pressure point and footpad ulceration; and nodules on the tongue (Fig. 21:35).

Systemic signs are not always present. The possibility of renal insufficiency or plasmacytoma should be investigated.

Diagnosis
Diagnosis is based on dermatological signs, skin biopsies and presence of an underlying illness.

21.26: Same animal as that in figure 21.26: alopecia, digital erosions, multiple onychodystrophy.


21.29: Skin histopathology (hereditary dermatomyositis): hydropic degeneration of basal keratinocytes and pigmentary incontinence (H&E stain).

21.30: Skin histopathology (hereditary dermatomyositis): hydropic degeneration of basal keratinocytes, apoptotic keratinocytes, lymphocytic satellitosis and pigmentary incontinence (H&E stain).

21.31: Skin histopathology (hereditary dermatomyositis): perifollicular lymphocytic infiltrate (H&E stain).

21.32: Muscle histopathology (hereditary dermatomyositis): myofibrillar degeneration and lympho-plasmocytic infiltrate (H&E stain).
Histopathological examination of skin biopsies reveals a diffuse, interfibrillar or perivascular, eosinophilic, amorphous deposit interspersed with lympho-plasmacytic cells. Congo Red staining in polarised light gives the characteristic green birefringence of amyloid (Fig. 21:36). Identical lesions can sometimes be seen in other organs (e.g. liver and kidneys).

Haematology, biochemistry and electrophoresis should be carried out.

If there is monoclonal gammapathy, a bone marrow aspirate should be performed to investigate the possibility of a plasmacytoma.

**Prognosis and treatment**

Prognosis is guarded.

There is no specific treatment available in the dog.

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**Cutaneous xanthomatosis**

**Aetiopathogenesis**

Xanthomas (xanthomatosis) are yellowish, cutaneous or subcutaneous lesions associated with an accumulation of lipid in dermal connective tissue. The few cases reported in the dog have been caused by defective lipid metabolism linked to either diabetes mellitus or acute pancreatitis. Although secondary hyperlipidaemia is seen with xanthomas associated with diabetes mellitus, it is not seen in xanthomas associated with acute pancreatitis; an effect on pancreatic lipase is suspected in this case. Of course, acute pancreatitis may result from hyperlipidaemia and, therefore, be associated with xanthomatosis.

**Clinical signs**

Dermatological signs include papules, plaques and cutaneous and subcutaneous nodules resembling candle wax (Fig. 21:37). There are no predilection sites and lesions are neither pruritic nor painful.

**Diagnosis**

Diagnosis is based on dermatological signs, skin biopsies, abnormal lipid metabolism and the presence of an underlying illness.

Histopathological examination of skin biopsies reveals spumous histiocytes, multinucleate giant cells and Touton cells (Fig. 21:38). Lipid in histiocytes takes up Congo Red stain.

Lipid metabolism should be investigated (cholesterolaemia, triglyceridaemia, lipoprotein electrophoresis, chylomicron test, pancreatic lipase).

**Prognosis and treatment**

Prognosis and treatment depend on the underlying illness.

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**Pedal self-mutilation syndrome**

**Aetiopathogenesis**

Pedal self-mutilation syndrome is a rare, hereditary, sensory neuropathy described in the English Pointer, German Pointer, Dachshund and Springer spaniel. In the English Pointer, an autosomal recessive transmission is involved. The condition develops in 3-8 month old puppies, affecting several animals in the same litter. It arises from faulty development or differentiation of primary sensory neurons. Absence of nociceptive sensation usually leads to mutilation of the paws.
21.33: Calciosis cutis in a Brittany Spaniel: erythematous papules on the bridge of the nose.

21.34: Same animal as that in figure 21.33: abdominal radiography (lateral view): persistent urachus causing chronic renal insufficiency.

21.35: Cutaneous amyloidosis in a Siberian Husky: ulcers revealing orange dermis, and purpura on the lateral thorax.

21.36: Skin histopathology (cutaneous amyloidosis): amorphous amyloid in the dermis (H&E stain).


21.38: Skin histopathology (xanthoma): spumous histiocytes (H&E stain).


21.40: Syringomyelia in a Cavalier King Charles spaniel: magnetic resonance imaging (courtesy of K. Gnirs).
Clinical signs

**Signs** include constant biting and licking of pedal extremities which are often cold and painful. Paws become swollen, and footpads and digits become ulcerated (Fig. 21:39). Puppies attack their own phalanges. In the English Pointer, carpi and tarsi can be more sensitive to pain. Skin sensitivity more proximally is preserved. Except for loss of distal deep pain sensitivity, neurological examination is normal.

Diagnosis

Diagnosis is based on consistent clinical signs in a predisposed breed.

**Skin biopsies** are unhelpful.

**Histopathological examination of spinal ganglia** (taken post-mortem) reveals a reduced number of neurons. Degeneration of myelinated and non-myelinated fibres is observed in dorsal roots and peripheral nerves.

Prognosis and treatment

**Prognosis** is very guarded.

*There is no treatment;* animals are rapidly euthanased. Parents and siblings of affected animals should not be used for breeding.

Syringomyelia syndrome

Aetiopathogenesis

Syringo-hydromyelia is caused by an abnormality in the central nervous system. It involves one or more cavitations in the cerebral trunk (syringobulbia) or, more commonly, in the spinal cord. Hydromyelia results from a localised fluid dilatation in the pre-existing central medullary canal and is, therefore, confined by ependymal cells. Syringomyelia results from a cavitation in medullary parenchyma creating cavitations which should not be present. As the difference between these two lesions is very difficult to appreciate ante-mortem, the term syringohydromelia is often preferred. At a late stage, poor closing of the spinal cord is sometimes seen dorsally. The abnormality may result from faulty neural tube development or other congenital or acquired neurological conditions.

This abnormality, associated with other central nervous system malformations (notably of the caudal fossa), is transmitted via an autosomal recessive gene in the Cavalier King Charles spaniel. Incidence is raised in the United Kingdom.

Clinical signs

**Signs** are very varied. Initial signs usually appear between 6 months and 3 years. Typically, they start with torticollis and forelimb weakness associated with muscle wasting. After the age of one year, cervico-scapular pruritus (hyperaesthesia) is apparent following involvement of the spino-thalamic tracts; affected dogs often present with neck pain, torticollis and spontaneous crying. In the most serious cases, severe peripheral motor neuronal paralysis (due to involvement of the cranial cervical region), mental retardation (caused by hydrocephalus), vestibular and cerebellar signs can be seen. There is no correlation between the size of gross lesions and the severity of clinical signs.

Diagnosis

Diagnosis is based on the presence of various neurological signs in the breed, and various diagnostic procedures (e.g. myelography, CT scanning and magnetic resonance imaging). **Myelography** is probably the most difficult procedure to interpret. The spinal cord is abnormally large in the cervical region, and dorsal and ventral arachnoid spaces are collapsed. An exaggerated dilatation of the central canal fills with contrast media and sometimes even the dorsal and lateral funiculi, resulting from syrinx formation, are apparent.
CT scanning reveals the presence of an abnormal cavity in the spinal cord, and sometimes meningoceles. It can also reveal engagement of the cerebellum resulting from a reduction in free space in the caudal fossa, and hydrocephalus.

Magnetic resonance imaging is the procedure of choice. Many bony cervico-occipital abnormalities can be demonstrated including tentorial platybasia, occipital stenosis and reduction of the sub-tentorial space. Various parenchymal abnormalities (e.g. cerebellar engagement in the foramen magnum, syrinx and meningoceles) can also be appreciated, possibly associated with hydrocephalus (Fig. 21:40).

Treatment
Various therapeutic options are available.

Medical treatment involves oral glucocorticoids (prednisolone, 1-2 mg/kg BID), and oral acetylazolamide (1-4 mg/kg TID) aimed at reducing cerebrospinal fluid production. Treatment needs to be adapted for each case. The condition can be stabilised long-term with medical treatment.

Surgical treatment may be indicated if medical treatment is unsuccessful or if there are abnormalities in the cervico-occipital and bulbo-medullary junctions. Various surgical procedures (e.g. decompression of the posterior fossa by dorsal occipital craniotomy, durotomy, syrinx marsupialisation and drainage of fluid from the sub-arachnoid space and peritoneal cavity) have been proposed.

References


