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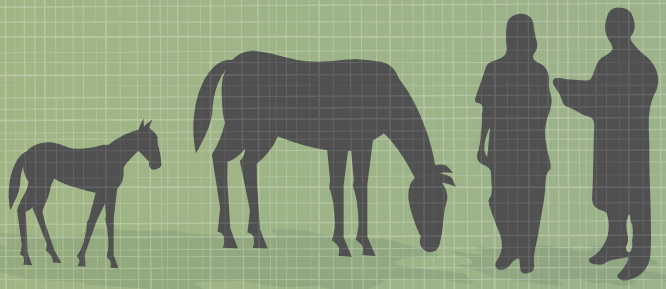

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Sarcoidosis

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Idiopathic systemic granulomatous disease or equine sarcoidosis is a severe systemic disorder, which is characterised by granulomatous inflammation of multiple organs including the skin. Exfoliative dermatitis with generalised scaling and crusting combined with variable alopecia dominates the clinical picture. An even rarer dermatological 'nodular' form also includes nodules and tumour-like masses.

The cause is unknown, although different infectious but also noninfectious aetiological agents have been suggested, like EHV-2, *Borrelia burgdorferi*, *Mycobacteria* spp., hairy vetch (*Vicia villosa*) toxicosis. Like in human sarcoidosis the pathogenesis remains not fully understood. Currently, an interaction of one or multiple environmental agents causing a Th-1-driven inflammatory process that results in granuloma formation is thought to play a role.

Clinical signs depend on the organ system involved, which most commonly include the lungs and lymph nodes, but the gastrointestinal tract, nervous system, etc. can also be affected. In contrast to this very rare systemic disease, two other forms have been described: the partially generalised form with multifocal skin lesions and sometimes also peripheral lymphadenopathy, and the localised form, which is restricted to one skin area (like one leg or the face).

Localised form of sarcoidosis

In a study with 22 cases with sarcoidosis, the majority (68%) suffered from the localised form. The localised form typically affects one lower limb with crusting, scaling, and hair loss of varying degrees. Usually, horses are systemically healthy and show no signs other than the affected lower limb with the typical hyperkeratotic, crusted, and alopecic areas, which are sometimes painful and warm and can result in lameness. The coronet and hoof wall can also be abnormal, which makes the localised sarcoidosis a differential diagnosis for coronary band disorders.

There seems to be no sex predilection, but in some reports, geldings seem to be over-represented, for example Loewenstein *et al.* (2004) reported six of seven horses to be geldings. In contrast, Sloet and Grinwis (2013) only had 27% geldings in their case series of 22 patients with sarcoidosis. They draw a comparison to human medicine, where women are mainly affected by sarcoidosis.

The disease is described to occur in different breeds, but some authors report a predisposition in Thoroughbreds; for example five of nine horses described by Spiegel *et al.* (2006) were Thoroughbreds.

Most horses are older than 3 years and the majority of reported cases seem to be between 5 and 21 years of age.

The author has seen 17 cases of localised sarcoidosis: six mares, nine geldings and one stallion (in one horse sex was not reported). Eight horses were Warmbloods, two Thoroughbreds, one Thoroughbred cross, two Quarter Horses, one Standardbred and one Lusitano (in two horses breed was not reported). In all but one horse, which had two legs affected (one forelimb and one hindlimb), only one leg was affected: in eight horses one hindlimb and in seven horses one forelimb.

Diagnosis of the localised form of sarcoidosis

Diagnosis is based on history, clinical appearance and histopathology, with exclusion of infectious agents and other granulomatous diseases and distinctive histopathological alterations of the skin (lymphohistiocytic infiltration and multinucleated giant cells). In some cases, vasculitis may also be present. Some horses will also show haematological and blood biochemistry changes consistent with chronic inflammatory disease, like hyperproteinaemia or anaemia, and may be also hypercalcaemic.

Treatment and prognosis

Treatment of sarcoidosis consists of regulation of the exaggerated immune response and suppression of the granulomatous inflammation. In humans, corticosteroids are the first-line agents for treating sarcoidosis with tapering of steroids over a minimum of 1 year. If there is a lack of response to therapy or occurrence of corticosteroid side effects, second-line agents, like methotrexate or azathioprine, are used. Third-line agents in human sarcoidosis are infliximab and adalimumab, so called cytokine-specific biological agents.

In horses, clinical management seems to be problematic. Although a good response to long-term systemic corticosteroids (dexamethasone, prednisolone) is reported for the localised form, anecdotally not all horses seem to respond that well. Few horses have shown spontaneous improvement or remission and some cases seem to remain unaltered, with or without treatment, but more commonly a slow but unstoppable progression is observed. Administration of corticosteroids is still the treatment of choice and Sloet (2001) reported a satisfying result in treating local sarcoidosis with 1 mg/kg prednisolone orally in the morning. 'Satisfying' in this publication was defined as preventing progression rather than complete resolution. Sloet and Grinwis (2013) recommended an initial high dose of systemic corticosteroids (1–2 mg/kg prednisolone orally once a day) for several weeks or longer. No local treatment was advised because the skin of affected areas is usually very fragile and sensitive. Sometimes tumour necrosis factor-alpha inhibitors like pentoxifylline are used in addition to steroids. But so far, the benefits of additional treatments, like omega-3 fatty acids, are not known.

Seven of the 17 horses seen by the author had been treated with prednisolone orally, all with either only slight improvement or no improvement. The lack of response to treatment with corticosteroids might be explained by the fact that most horses are treated with prednisolone doses not exceeding 1 mg/kg. Some authors think that aggressive immunosuppression, with a prednisolone dose of 2–4 mg/kg orally once a day, is warranted. Another reason could be that horses are not treated for long enough and therapy is discontinued after only a few weeks.

Prognosis seems to be highly variable but is considered poor in generalised and partially generalised cases. Prognosis for the localised form is good for survival but guarded with respect to the skin problem. In one study 26.7% recovered fully with or without treatment, but 53% needed continuous low maintenance doses of prednisolone (0.2–0.5 mg/kg/day). In another study, two of nine horses with localised sarcoidosis showed a full recovery and seven showed only some improvement in clinical signs. Unfortunately, these remaining skin lesions are viewed as problematic by most owners.

Two of the 17 horses had major involvement of the coronary band and were finally euthanased due to laminitic lesions on the affected limbs. To the author's knowledge, this has not been reported previously.

Localised sarcoidosis seems to be not as rare as the generalised form. It is not clear if there is a real increase in prevalence or if the increase in case numbers is due to earlier recognition of the disease. Like in human medicine, drugs other than corticosteroids might be more efficient. Research is needed to not only find more successful ways to treat the disease but also answer questions regarding the aetiopathogenesis and why there is only one leg affected, if environmental antigens or an infectious process leading to chronic antigenic stimulation are involved.

References

Available on request from the author.