

Congenital ichthyosis in a Maltese dog: A case report

TAE-SEOK KIM¹, JUNG-A SEO², TAE-HO OH², JIN-KYU PARK³,
KYU-SHIK JEONG^{3*}, IL-HWA HONG^{1,4*}

¹Department of Veterinary Pathology, College of Veterinary Medicine,
Gyeongsang National University, Jinju, Republic of Korea

²Department of Veterinary Internal Medicine, College of Veterinary Medicine,
Kyungpook National University, Daegu, Republic of Korea

³Department of Veterinary Pathology, College of Veterinary Medicine,
Kyungpook National University, Daegu, Republic of Korea

⁴Institute of Animal Medicine, Gyeongsang National University, Jinju, Republic of Korea

*Corresponding author: jeongks@knu.ac.kr; ihhong@gnu.ac.kr

Citation: Kim TS, Seo JA, Oh TH, Park JK, Jeong KS, Hong IH (2021): Congenital ichthyosis in a Maltese dog: A case report. *Vet Med-Czech* 66, 546–549.

Abstract: This case report describes congenital ichthyosis in a Maltese dog, a condition, which has not previously been reported in this breed. The dog presented with several dry, tightly adhering scales showing a multifocal appearance. The histology revealed severe orthokeratotic hyperkeratosis and hyperplasia with an irregular granular layer without epidermolysis. Non-epidermolytic ichthyosis (NI) has strong breed predilection in dogs, and its morphological features vary by breed while the microscopic changes are similar. It is likely to be the first reported case of congenital ichthyosis in a Maltese dog showing a different morphological appearance compared to other breeds with a higher prevalence of NI.

Keywords: canine; cornification; hyperkeratosis; multi-focal

Ichthyoses in veterinary medicine are usually limited to congenital skin disorders, principally seen in dogs and calves, characterised thickening of the skin with marked hyperkeratosis resembling fish scales. Congenital ichthyoses have also been reported in humans, cats, chickens, pigs, laboratory mice, a llama (Helman et al. 1997; Scott et al. 2001; Mauldin et al. 2008; Zachary and McGravin 2017). Several forms of ichthyoses are known in humans, while, in the veterinary field, the condition is generally classified as an epidermolytic or non-epidermolytic subtype on the basis of its microscopic features. In an epidermolytic ichthyosis (EI), hyperkeratosis and hypergranulosis occur in combination with the vacuolisation and lysis of keratinocytes within the granular and spinous

cell layers (Mauldin 2013; Hoffmann et al. 2016). A non-epidermolytic ichthyosis (NI) is typically characterised by a prominent granular layer, with the presence of numerous mitotic figures in the keratinocytes, with marked lamellar orthokeratotic hyperkeratosis, and mild acanthosis in the absence of epidermal hyperplasia and dermal inflammation (Scott et al. 2001; Mauldin 2013; Hoffmann et al. 2016). Canine ichthyosis commonly appears in the non-epidermolytic form and has strong breed predilections (Mauldin 2013; Hoffmann et al. 2016; Zachary and McGravin 2017). In dogs, NI has previously been reported in purebreds, such as Golden Retrievers, American Bulldogs, West Highland White Terriers, and Jack Russell Terriers (Scott et al. 2001; Mauldin 2013; Hoffmann et al. 2016).

<https://doi.org/10.17221/143/2020-VETMED>

Most reported cases have been limited to certain breeds, some of which have established general, clinical, and microscopic features, but clinical features may vary by breeds. This case study reports NI in a female Maltese dog, a breed not previously known to be affected by this condition. The histology revealed several indications of NI, although the morphological features of this case differed from cases reported in the breeds with the higher prevalence of NI.

Case description

An 8-month-old female Maltese dog was presented at a veterinary clinic for a scaling skin disorder, which had been present since adoption as a young

puppy. No history or clinical information from the parents or litter-mates was available. Its hair was shaved for better observation. Lesions with scales showing a multi-focal appearance were present, interspersed with seemingly normal areas of skin. These lesions were especially severe on the caudal trunk, upper hind limbs, neck, and head (Figure 1A). Lesions were absent from the face and the paws. The lesions showed prominent accumulation of dry, tightly adhering, and tannish-grey scales (Figure 1B). Several layers of these scales were found on the lesions, which were non-pruritic and did not wax or wane. Representative biopsies were taken from some of the major lesions and apparently normal areas of skin. The tissues were fixed in 10% neutral-buffered formalin, routinely processed, sectioned at 4 μ m, and stained with haematoxylin and eosin

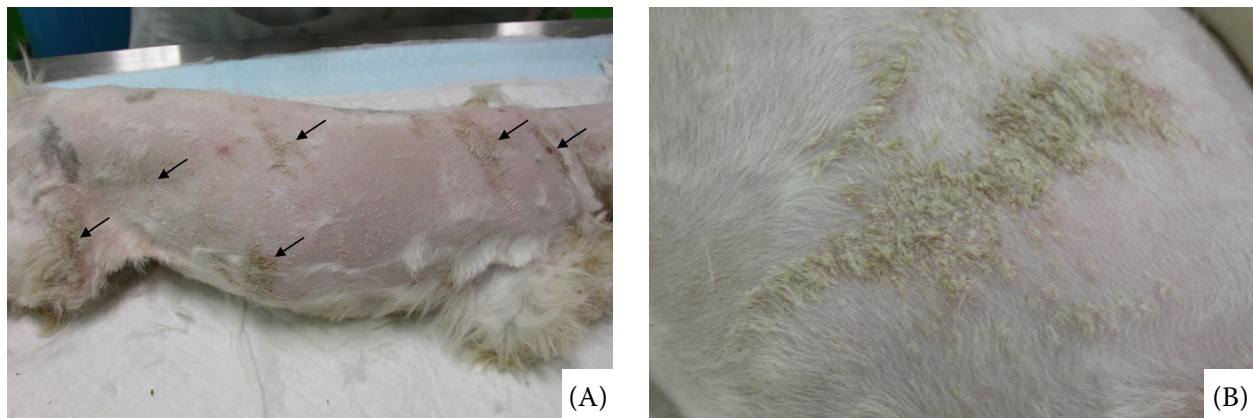


Figure 1. Skin of the Maltese dog with congenital ichthyosis (hair shaved to facilitate observation)

(A) Several scaling lesions (arrows) with a multi-focal appearance are present among the apparently normal areas of skin. Generalised erythema on the trunk is due to the shaving. (B) Prominent accumulation of dry, tightly adhering, and tannish-grey scales with several layers

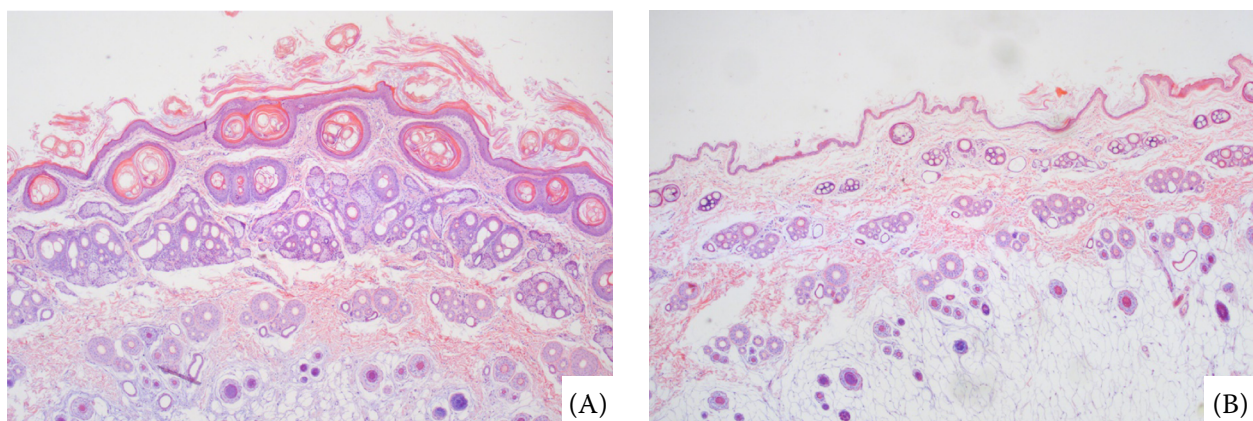


Figure 2. Microscopic examination of the ichthyosis in the Maltese dog

(A) Major lesions showing epithelial hyperplasia in combination with marked, lamellar, orthokeratotic, and follicular hyperkeratosis with plugging of follicles in the stratum corneum. (B) The skin areas without scales have no signs of hyperkeratosis or hyperplasia. Haematoxylin and eosin (H&E), $\times 40$

<https://doi.org/10.17221/143/2020-VETMED>

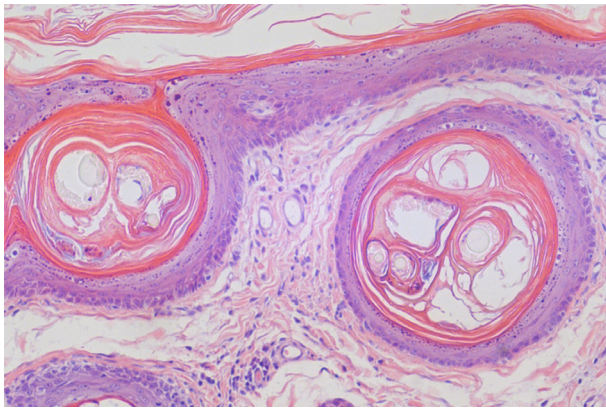


Figure 3. Epidermis of the skin with severe scales

A small number of vacuolated keratinocytes are present, and the granular layer with abundant keratohyalin granules is prominent. Marked orthokeratotic hyperkeratosis of dilated hair follicle infundibula with frequent keratotic plugging can be observed. No significant dermal inflammation is present. Haematoxylin and eosin (H&E), $\times 200$

(H&E). The affected skin showed epithelial hyperplasia in combination with marked, laminar, orthokeratotic, and follicular hyperkeratosis with plugging of the follicles in the stratum corneum (Figure 2A), whereas the skin areas without scales showed no signs of hyperkeratosis or hyperplasia (Figure 2B).

The superficial epidermis in the affected skin contained small numbers of vacuolated keratinocytes, and the granular layer, which contained abundant keratohyalin granules, was irregular, but prominent (Figure 3).

Marked orthokeratotic hyperkeratosis of dilated hair follicle infundibula with frequent keratotic plugging and no significant dermal inflammation was found. No notable non-cutaneous signs were identified.

DISCUSSION AND CONCLUSIONS

The clinical features of this case, which showed multi-focal non-pruritic scales since birth, indicate the primary disorders of cornification, especially ichthyosis (Scott et al. 2001; Mauldin 2013). The microscopic investigation demonstrated that the affected skin of the present case was characterised by NI, including marked, laminar, orthokeratotic, and follicular hyperkeratosis with vacuolisation of the keratinocytes, an irregular, but prominent, granular layer, and epithelial hyperplasia. No der-

mal inflammation or epidermolysis were found. Similar histopathological features have been described earlier in other purebred dog breeds with non-epidermolytic ichthyosis (NI) such as Golden Retrievers, American Bulldogs, Highland White Terriers, Cavalier King Charles Spaniels, Doberman Pinschers, Jack Russell Terriers, Norfolk Terriers, and Yorkshire Terriers (Scott et al. 2001; Mauldin 2013; Hoffman et al. 2016; Zachary and McGravin 2017). Changes in the folliculosebaceous unit containing accumulations of an amorphous material has been reported in Great Danes, but it has not been described in any other breeds yet (Hoffman et al. 2016). Although the histological features of NI are similar in most breeds, its clinical presentation, including the characteristics of the scales, distribution of the lesions, erythroderma, and abnormalities in other parts of the skin and other organs, varies (Mauldin et al. 2008). NI in Golden Retrievers is presented with a relatively “mild” form of scaling, which is prominent on the trunk and without epidermal hyperplasia (Mauldin 2013). Their scales are large, loosely adherent, soft, and white-to-grey, but the paw pads and nasal planum are not affected, and the condition may be associated with ventral hyperpigmentation (Mauldin et al. 2008; Mauldin 2013). Unlike Golden Retrievers, American Bulldogs consistently develop clinical signs before weaning, and while the skin lesions do not wax or wane, they are generally more severe (Mauldin 2013). Large white to light tan scales are distributed across the entire haircoat and paw pad hyperkeratosis is occasionally present (Mauldin 2013). Jack Russell Terriers with NI have generalised and loosely attached parchment paper-like scales, and secondary infections with bacteria and the fungus *Malassezia* causing pruritis are common (Mauldin 2013; Zachary and McGravin 2017). Affected Norfolk Terriers have superficial epidermis sloughing off after a mild mechanical trauma and its intertriginous area develop pigmented scales (Zachary and McGravin 2017). Cavalier King Charles Spaniels have also been diagnosed with congenital ichthyosis with keratoconjunctivitis sicca, scaling, abdominal hyperpigmentation, footpad hyperkeratosis, and nail dystrophy (Barnett 2006; Mauldin 2013). Although the clinical features of ichthyosis vary among breeds, most of the cases are characterised by loosely attached scales that are distributed over the entire skin area. In contrast, this case showed multi-focal scaling interspersed with normal areas of skin, and the scales were dry

<https://doi.org/10.17221/143/2020-VETMED>

and tightly adhering. Lesions with scales were especially severe on the caudal trunk, upper hind limbs, neck, and head, except on the face and the paws. Interestingly, genetic mutations giving rise to ichthyosis also vary among breeds, such as the Norfolk Terrier (variant in *KRT10* gene), Golden Retriever (variant in *PNPLA1* gene), American Bulldog (variant in *ICHTHYN* and similar to the *PNPLA-2* in Golden Retrievers), and Jack Russell Terrier (variant in *TGM1* gene) (Mauldin 2013). Therefore, the Maltese dog in this case study may have a different genetic mutation from other breeds with previously reported predilections for ichthyosis. Thus, even if a case shows different morphological appearances from the known ichthyosis, or occurs in a dog breed that has not been known to previously have ichthyosis-form dermatitis, congenital ichthyosis should be highly considered in the differential diagnosis of a scaling skin disease, which is present from birth. In conclusion, we present the first report on a congenital, non-epidermolytic ichthyosis in a Maltese dog, which is characterised by multi-focal lesions with scales interspersed with normal skin.

Conflict of interest

The authors declare no conflict of interest.

REFERENCES

- Barnett KC. Congenital keratoconjunctivitis sicca and ichthyosiform dermatosis in the Cavalier King Charles Spaniel. *J Small Anim Pract.* 2006 Sep;47(9):524-8.
- Helman RG, Rames DS, Chester DK. Ichthyosiform dermatosis in a Soft Coated Wheaten Terrier. *Vet Dermatol.* 1997;8(1):53-8.
- Hoffmann A, Metzger J, Wohlke A, Peters M, Junginger J, Mischke R, Distl O, Hewicker-Trautwein M. Congenital ichthyosis in 14 Great Dane puppies with a new presentation. *Vet Pathol.* 2016 May;53(3):614-20.
- Mauldin EA. Canine ichthyosis and related disorders of cornification. *Vet Clin North Am Small Anim Pract.* 2013 Jan;43(1):89-97.
- Mauldin EA, Credille KM, Dunstan RW, Casal ML. The clinical and morphologic features of nonepidermolytic ichthyosis in the Golden Retriever. *Vet Pathol.* 2008 Mar; 45(2):174-80.
- Scott DW, Miller WH, Griffin CE. *Muller and Kirk's small animal dermatology.* 6th ed. Philadelphia, PA, USA: W. B. Saunders; 2001. p. 922-4.
- Zachary JF, McGravin MD. *Pathologic basis of veterinary disease.* 6th ed. St Louis, MO, USA: Elsevier Mosby; 2017. p. 1001-2.

Received: July 8, 2020

Accepted: July 30, 2021