

question. CATs could summarize the evidence of one specific patient group (e.g. age group, a subgroup with a comorbidity) or a specific treatment aspect (e.g. dose, duration or frequency), comparison and a single outcome. The information from some CATs could be combined to answer a broader question. In the near future, we hope to have instructions for authors of CATs as we have for other types of articles in the *BJD*.

Perhaps we could draw your attention to the Cochrane Clinical Answers (<http://cochraneclinicalanswers.com/>). They also provide a readable, digestible, clinically focused entry point to rigorous research from Cochrane systematic reviews. They are designed to be actionable and to inform decision making at the point of care. Each Cochrane Clinical Answer contains a clinical question, a short answer and an opportunity to 'drill down' to the evidence from the relevant Cochrane review.

We are always keen for submissions of new CATs to our section. Published CATs will help to avoid duplication of work. We would particularly encourage dermatology trainees to prepare CATs on clinical scenarios they have encountered, as this can be a very useful way of developing skills in critical appraisal of the medical literature.

We too hope that Dr Sladden's dream of a 'clowder of CATs' will one day become a reality!

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A patient with disseminated strongyloidiasis with erythroderma in a nonendemic area

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DEAR EDITOR, *Strongyloides stercoralis* is an intestinal nematode endemic to tropical areas,¹ as well as south-western Japan. Patients with suppressed immunity can develop a severe disseminated strongyloidiasis involving spreading of the larvae to extraintestinal organs. A diagnosis of strongyloidiasis is challenging, particularly in nonendemic areas. Many cutaneous presentations have been reported, such as serpiginous, urticarial weals,² petechiae and purpura;³ however, erythroderma is extremely rare and has never been reported in either the English or wider dermatological literature. Herein, we report a patient with *S. stercoralis* hyperinfection with erythroderma in a nonendemic area of Japan.

A 53-year-old Japanese woman with a medical history of chronic eczema was admitted to our hospital for high fever, rash and lichenification on the whole body (erythroderma), and marked oedema (Fig. 1). She was 1.45 m in height and weighed 31 kg. The laboratory studies disclosed the following values: white blood cell count 26.9×10^9 cells L⁻¹ with 83% neutrophils and 0.9% eosinophils, C-reactive protein 89.6 mg L⁻¹, albumin 10 g L⁻¹, and negative antihuman T-cell lymphotropic virus-1 and HIV antibodies. Nephrotic syndrome was denied by urinalysis. She had no other contributory medical history or medication. Computed tomography revealed a giant uterine tumour, and no other evidence of malignancy or infection was detected. Cutaneous pathology indicated acanthosis and inflammatory infiltration of lymphocytes with no atypia in the dermis.

After admission, she manifested digestive symptoms including nausea and diarrhoea, and repeatedly became septic. Septic shock and disseminated intravascular coagulation also occurred. The blood culture grew *Streptococcus dysgalactiae* subsp., *Enterococcus faecalis* and *Providencia rettgeri*; all of these were intestinal resident bacteria. We treated the patient with the appropriate antibiotic regimen and started her on prednisolone 40 mg per day to treat her erythroderma. The eruption immediately improved and we reduced the prednisolone gradually. When the patient was on prednisolone 15 mg per day, a hysterectomy was performed. The giant uterine tumour was revealed to be a myoma; however, larvae consistent with *S. stercoralis* were isolated from the ascites taken during the operation (Fig. 2).

Because parasitic infection is quite uncommon in the region of our department and the patient did not manifest eosinophilia, we did not conduct parasite examinations, such as agar plate culture, before the operation. However, the direct detection of *S. stercoralis* in the ascites was the most indubitable evidence and a diagnosis of disseminated strongyloidiasis was made. The enzyme-linked immunosorbent assay (ELISA) of anti-*Strongyloides* antibody was negative. Although we carefully took a history of the patient and her family, she revealed not having travelled to or lived in endemic regions. Considering that fatal sepsis



Fig 1. The clinical manifestation on admission.



Fig 2. The larvae of *Strongyloides stercoralis* in the ascites.

occurred repeatedly in our patient, we administered oral ivermectin (9 mg) immediately. She received the medication twice at an interval of 2 weeks. After the introduction of ivermectin, a duodenum biopsy and agar plate culture were performed, and no larvae were detected. She recovered gradually and was discharged after 3 weeks from the start of ivermectin. However, 2 months later she died of sepsis from pneumonia. The autopsy revealed neutrophilic infiltration of the lung but no larvae.

The characteristic life cycle of *S. stercoralis* is 'autoinfection', where the nematode continues to reproduce larvae within a host for a long time.⁴ Dissemination of larvae into multiple organs, as a result of an increase in the autoinfection cycle,⁵ can lead to expansion of intestinal bacteria and sepsis.⁶ The risk is extremely high in immunosuppressed patients such as those with malnutrition and autoimmune disease,⁵ and sometimes they do not manifest eosinophilia.⁷ A serology test (ELISA) is useful for diagnosis; however, the sensitivity is probably lower in patients who are already immunosuppressed.⁸ Strongyloidiasis presents various dermatological manifestations. The most characteristic lesion is larva currens, a serpiginous creeping urticarial and pruritic eruption caused by intradermal migration of the larvae. An absence of larvae in biopsy specimens is possible.² The cutaneous manifestation of erythroderma, as in our patient, is extremely rare and has never been reported in the literature. The patient had chronic eczema, but no other contributory medical history for ery-

throderma including psoriasis, medication, lymphoma and malignant tumour.

Chronic pruritic urticarial eruption is one of the cutaneous manifestations of strongyloidiasis.³ Corsini reported a patient with strongyloidiasis with generalized pruritic erythema, and suggested that an allergic mechanism might play a role in the pathophysiology.⁹ It might be possible in our patient that an allergic reaction to *S. stercoralis* resulted in deterioration of the patient's chronic eczema and led to the onset of erythroderma. Patients without a history of living or travelling in an endemic area are also very rare. It is reported that *Strongyloides* inhabited a wide area of Japan until the 1960s.¹⁰ It might be possible that the larvae had infected the patient at that period, then autoinfected up to the present. Our case demonstrates the difficulty of diagnosing the disease. It is important for dermatologists to be aware of patients with strongyloidiasis with variable skin manifestations, even in nonendemic areas and even if they have no contributory history.

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Avascular necrosis of the hip and diffuse idiopathic skeletal hyperostosis during long-term isotretinoin treatment of epidermolytic ichthyosis due to a novel deletion mutation in *KRT10*

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DEAR EDITOR, Epidermolytic ichthyosis (EI; OMIM 113800), previously termed bullous congenital ichthyosiform erythroderma or epidermolytic hyperkeratosis, is a clinically heterogeneous disorder of keratinization. It is usually characterized by severe neonatal erythroderma, blistering and fragile skin, with the subsequent development of hyperkeratosis, predominantly in flexural areas. It is caused by mutations in either the *KRT1* or *KRT10* genes encoding the suprabasal keratins K1 and K10, respectively.¹ Mutations are usually missense substitutions in the highly conserved alpha-helical rod domains of these keratins, which play a critical role in filament formation.² We report a multigeneration kindred with EI due to a novel mutation in *KRT10*.

The proband was a 32-year-old woman from Shetland. She presented with widespread fine scale and erythema of her trunk and limbs, with a history of scaling and redness since birth but no blistering, erosions or collodion membrane reported. Clinical examination revealed widespread ichthyosis and erythema affecting the trunk and all four limbs, with more significant hyperkeratosis at the elbows, knees and ankles but relative sparing of palmoplantar skin (Fig. 1). Her father, uncle and grandmother were affected, and four further generations were reported to be affected. She had been maintained on oral isotretinoin 20–40 mg daily, thus modifying the clinical appearance, from the age of 13 years, but had recently developed lower back and hip pain. Radiographs of the lumbar spine and left hip demonstrated bridging osteophytes from T11 to L1, suggestive of diffuse



Fig 1. Family with epidermolytic ichthyosis. (a) Clinical picture of the proband: hyperkeratosis of nonplantar sites modified by oral retinoid therapy; (b, c) photographs of her father.

idiopathic skeletal hyperostosis (DISH), and magnetic resonance imaging confirmed the presence of avascular necrosis of the left hip. This was successfully treated with core decompression of the left hip with improvement in the patient's pain.

A biopsy of affected skin of the upper limb was obtained from the proband and processed for light microscopy by standard methods. Structural analysis demonstrated acanthosis, marked overlying hyperkeratosis and vacuolar change of the upper epidermal cells with prominent clumping of keratohyaline granules.

Following informed consent, genomic DNA samples were obtained from blood samples from the proband and her father. Mutation analysis of the coding regions and splice sites of the *KRT1* and *KRT10* genes was performed by standard polymerase chain reaction (PCR) and Sanger sequencing methods using specific primers. Sequence analysis of *KRT10* revealed a previously unreported heterozygous deletion of 167 base pairs extending from intron 5 into exon 6 (c.1156–79_1243del), abolishing the intron 5 acceptor splice site (Fig. 2). This mutation was also present in the proband's affected father.