

Keratosis Follicularis Spinulosa Decalvans Associated with Leukonychia

The Editor,

Sir,

Keratosis follicularis spinulosa decalvans (KFSD) is a rare X-linked disorder (1, 2) characterized by diffuse follicular hyperkeratosis, progressive scarring alopecia of scalp, eyebrows, and eyelashes and may be accompanied by photophobia, corneal and conjunctival inflammation and dystrophy, tooth abnormalities, ichthyosiform xerosis, facial erythema, atopic dermatitis, hair disorders and nail dystrophies (3). We report here a case of KFSD with leukonychia partialis.

A 24-year old Turkish man suffering from oozing, crusting wounds on his scalp and white discoloration of his nails was referred to the Department of Dermatology, Haseki Research and Training Hospital. He presented with a progressive hair loss and periodic white discoloration (lasting for four to five months and then recovering for one to two months) of his nails twice a year for 15 years.

His dermatological examination revealed sparse hair



Fig. 1: Sparse hair on the scalp and atrophic areas as patches.

on the scalp with patchy atrophic areas (Fig. 1), sparse thin eyelashes and erythematous cheeks with multiple keratotic papules, 0.1 mm in diameter. There were follicular keratotic papules and postinflammatory hyperpigmentation on his scalp near the hair border, the back of his neck and throughout the trunk. Mild hyperkeratosis on his palms and soles, opaque white discoloration and significant cuticula on his entire finger nails were evident (Fig. 2). The patient did not have any subungual or nail bed abnormality and mycologic



Fig. 2: Opaque whitening and significant cuticula on finger nails, most notable on second, third and fifth nails.

examinations were negative.

The ophthalmological examination revealed bilateral subepithelial corneal opacities. The patient was otherwise healthy and laboratory tests showed no abnormalities. Histopathology from follicular papules of the scalp showed marked osteal dilatation, hyper and parakeratosis, complete degeneration of the inner and outer root sheath in the follicle. The follicular infundibulum showed a perifollicular fibrosis with a medium dense inflammatory cell infiltrate composed of lymphocytes, plasma cells and neutrophils and a mild perivascular lymphocytic infiltrate was present. The epidermis was not involved except for the marked acrosyringium. Skin biopsy specimens from the alopecic areas of the scalp showed complete loss and fibrosis of hair follicles.

Nail biopsy specimen showed hypergranulosis in the matrix and mild parakeratosis in the nail plate. Histopathology from the distal end of the opaque nails showed cells with keratohyalin content, suggesting the diagnosis of true leukonychia.

Scarring alopecia of the scalp and eyelashes with childhood onset, facial erythema, diffuse follicular hyperkeratosis on the trunk, bilateral corneal opacities, as well as atrophy seen in histopathologic examination of the scalp skin and follicular hyperkeratosis in the skin biopsy specimen of the papules suggested to us the diagnosis of KFSD for this case.

We report, to our knowledge, the first case of KFSD with leukonychia partialis and suggest that leukonychia may be seen in KFSD.

Keywords: Keratosis follicularis spinulosa decalvans, KFSD, leukonychia, nail biopsy, skin biopsy

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Safety Audit for Internal Emergencies and Theatre Disasters (SAFE-T): From Barbados

The Editor,

Sir,

On September 9, 2009, fire swept through a hospital on the Caribbean island of St Lucia, killing three patients (1). A definite plan to deal with major emergencies is an important element of any health institution. To our knowledge, there is scarcity of published data regarding the knowledge and preparedness of staff from the Caribbean region. The main objective of this study was to assess the level of awareness of staff working in the operating room (OR) and recovery room (RR), and to determine the level of preparedness with regard to operating theatre disasters and internal emergencies (fire, flooding, building collapse).

The Internal Research Committee of the Queen Elizabeth Hospital (QEH), Barbados, gave approval for the questionnaire. A questionnaire survey with 24 questions was formulated by a team of anaesthetists and surgeons and was administered personally to all OR and RR staff over a one-month period. The 120 questionnaires administered were

individually coded and completed anonymously. Queen Elizabeth Hospital is a 700-bed tertiary care hospital. Data were analysed using SPSS v 17 analytical software.

Of the 83 completed questionnaires (Table), 60.2% respondents have been employed at QEH for more than five

Table: Summary of the staff's response for nine (out of 24) questions, represented in percentages

Question	Response
What best describes your current job	Anaesthetist 13.3%, surgeon 14.5%, registered nurse 34.9%
Do you know what to do in the event of an IE	Knew exactly 13.3% Had no idea 15.7%
How prepared do you feel for an IE	Very well prepared 9.6% Much unprepared 9.6%
Have you ever taken part in an IE drill	Yes 22.9% (71.1% did not know when the last internal drill was held)
Have you ever noticed emergency exit signs in OR, RR and Floor	Yes 71% No 22%
Do you know how to access emergency exit	Do not know 43.4%
Are there any fire extinguishers in OR/RR/Floor	Yes we knew OR: 66.2%, RR: 31.3%, Floor: 31.3%
Is there any automatic lighting provided in the event of power failure in OR/RR/Floor	Yes 22.9%, No 20.5%, Do not know 55.6%
If a patient is under anaesthesia and a disaster occurs, what do you do	Stay in the OR with patient 2.4% Rescue the patient from danger, cover or close the surgical site immediately and raise the alarm 74.7%

IE – internal emergency, OR – operating room, RR – recovery room

years, and 44.6% have been working in OR for more than five years. The majority of staff members (95.2%) thought that they had a role to play in patient care in the event of an internal emergency; 60.2% of respondents were able to correctly match the alert code colours to their meanings and 34.9% of respondents knew the location of the staff collection point in the event of an emergency. The majority of respondents (67.5%) thought that the nursing sister in charge was responsible for taking care of the patient during an emergency. Most (71.1%) thought that the operating theatre manager was responsible for the head count during an emergency. More than 90% thought that more needs to be done to ensure that all staff members are educated and said that they would participate in yearly emergency drills.

Internal emergencies like operating room fire, flooding, building collapse and failure of electrical supply can happen at any time. The incidence of OR fires is difficult to determine, due in part to the lack of a mandatory national reporting system for OR fires (2). Case reports indicate that