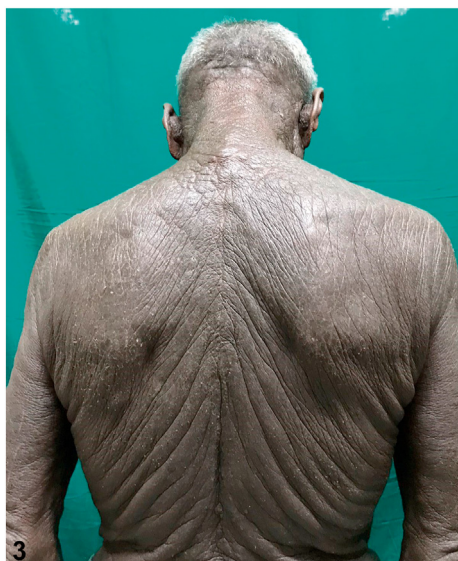


Generalized lichenification sparing the nose



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Key words: erythroderma; lichenification; nose sign.



A 76-year-old man presented with a 1-year history of generalized itching, accompanied by darkening of the skin, with significant weight loss and intermittent swelling of the legs. On examination, there was diffused, uniform, gray-brown colored pigmentation with lichenification of the skin accompanied by fine, semiadherent scales and generalized lymphadenopathy (Figs 1 to 3). The facial appearance was striking with exaggeration of the longitudinal and transverse furrows, supraciliary madarosis, and clear sparing of the nose (Fig 1). Investigations

revealed eosinophilia and peripheral blood smear examination showed no abnormality. Histopathologic examination showed hyperkeratosis; elongation of rete ridges; hypergranulosis; and dermal infiltration of the lymphocytes, plasma cells, and eosinophils.

Question 1: Which eponymous sign is depicted in the image?

- A. Hutchinson nose sign
- B. Raccoon sign
- C. Pavithran nose sign
- D. Jellinek sign
- E. Frank sign

Answers:

A. Hutchinson nose sign – Incorrect. Hutchinson nose sign refers to the papulovesicular lesions present on one side of the tip of the nose in herpes zoster and indicate the possibility of the involvement of the ipsilateral cornea because both are supplied by the nasociliary nerve, a branch of the trigeminal nerve.

B. Raccoon sign – Incorrect. Raccoon sign is seen in patients with systemic amyloidosis and is characterized by periorbital hemorrhages that occur because of a laxity of the blood vessels.

C. Pavithran nose sign – Correct. There is sparing of the nose with the absence of erythema and scaling over the nose. This was first described by Pavithran in exfoliative dermatitis.¹

D. Jellinek sign – Incorrect. Jellinek sign refers to pigmentation over upper portion of the eyelids, which is seen in hyperthyroidism.

E. Frank sign – Incorrect. Frank sign refers to the presence of a diagonal crease in the earlobes of adults and has been associated with an increased risk of atherosclerotic heart disease.

Question 2: Based on the clinical pictures, case history, and investigations, what is the most likely diagnosis?

- A. Mycosis fungoides

- B. Idiopathic erythroderma
- C. Actinic reticuloid
- D. Systemic amyloidosis
- E. Scleromyxedema

Answers:

A. Mycosis fungoides – Incorrect. The skin biopsy did not reveal the presence of any atypical lymphocytes, Pautrier microabscesses, and/or epidermotropism. Hence, this diagnosis was excluded.

B. Idiopathic erythroderma – Correct. The features of chronic erythroderma in this elderly man, with no history of a previous skin disease, generalized lymphadenopathy, and absence of histopathologic features of mycosis fungoides, support the diagnosis of erythroderma of unknown origin. The most common causes of erythroderma are atopic eczema of the elderly and drug-induced and pre-lymphomatous eruptions.²

C. Actinic reticuloid – Incorrect. Actinic reticuloid presents with infiltrated and lichenified papules and plaques over the photoexposed areas, which may progress to involve the covered parts as well.³ There will be no sparing of the nose. Histology will reveal prominent dermal fibroplasia.⁴

D. Systemic amyloidosis – Incorrect. Cutaneous manifestations in systemic amyloidosis include translucent, infiltrated papules over the face and flexural areas, a classic raccoon sign with periorbital hemorrhages, with biopsy showing extracellular amyloid deposits.

E. Scleromyxedema – Incorrect. This metabolic disorder presents with generalized, confluent papules and nodules with diffuse thickening of the underlying skin. A skin biopsy will reveal abundant dermal mucin.

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Question 3: Which of the following is a proposed mechanism for the occurrence of this sign seen on the nose?

- A.** Delayed cell-mediated hypersensitivity reaction to certain endogenous photo-induced allergens
- B.** Repeated rubbing of the nose by the patient, which is believed to help dislodge the scales from the nose
- C.** Antigen persistence leading to chronic lymphocytic stimulation
- D.** Impaired skin barrier, immune dysregulation, and allergen sensitization
- E.** Persistent vasodilatation involving the vessels supplying the nose

Answers:

- A.** Delayed cell-mediated hypersensitivity reaction to certain endogenous photo-induced allergens – Incorrect. This is the postulated mechanism for the development of chronic actinic dermatitis, possibly through photochemical modification of an endogenous antigen.
- B.** Repeated rubbing of the nose by the patient, which is believed to help dislodge the scales from the nose – Correct. Repeated rubbing of the nose and manipulation by the patient helps dislodge the scales and prevent the accumulation of physical and aero-allergens over the nose, resulting in an absence of inflammatory changes in this area.⁵
- C.** Antigen persistence leading to chronic lymphocytic stimulation – Incorrect. The nose sign is postulated to occur because of dislodging of the

allergens from the nose, resulting in the absence of scaling and pigmentation, which is evident over the rest of the face. Antigenic persistence leading to chronic lymphocytic stimulation is one of the implicated mechanisms in mycosis fungoides.

D. Impaired skin barrier, immune dysregulation, and allergen sensitization – Incorrect. These pathogenic mechanisms are implicated in atopic eczema; however, they do not connote any specific relation to sparing of the nose.

E. Persistent vasodilatation involving the vessels supplying the nose – Incorrect. The end arterial blood supply of the nose is proposed to contribute to the occurrence of nose sign by resulting in vascular insufficiency that may prevent the circulating antigens or antibodies from reaching it and getting lodged.⁵

Conflicts of interest

None disclosed.

REFERENCES

1. Kanwar AJ, Dhar S, Ghosh S. 'Nose sign' in dermatology. *Dermatology*. 1993;187(4):278.
2. Ingram JR. Eczematous disorders. In: Griffiths C, Barker J, Bleiker TO, Charmers R, Creamer D, eds. *Rook's Textbook of Dermatology*. 9th ed. Blackwell Publishing; 2016;39:39.1–39.35.
3. Toonstra J, Henquet CJ, van Weelden H, van der Putte SC, van Vloten WA. Actinic reticuloid. A clinical photobiologic, histopathologic, and follow-up study of 16 patients. *J Am Acad Dermatol*. 1989;21(2):205-214.
4. Sidiropoulos M, Deonizio J, Martinez-Escala ME, Gerami P, Guitart J. Chronic actinic dermatitis/actinic reticuloid: a clinicopathologic and immunohistochemical analysis of 37 cases. *Am J Dermatopathol*. 2014;36(11):875-881.
5. Agarwal S, Khullar R, Kalla G, Malhotra YK. Nose sign of exfoliative dermatitis: a possible mechanism. *Arch Dermatol*. 1992;128(5):704.