Dermatology Practical & Conceptual

Dermoscopic and Cytological Findings in Scleromyxedema

Tuğba Tehçi[1], Elif Burcu Şenyurt[2], Murat Durdu[3], Enzo Errichetti[4]

1 Department of Dermatology, University of Health Sciences -Adana Health Practice and Research Center, Adana, Turkey
2 Department of Pathology, University of Health Sciences -Adana Health Practice and Research Center, Adana, Turkey
3 Department of Dermatology, Başkent University Faculty of Medicine, Adana Dr. Turgut Noyan Application and Research Center, Adana, Turkey
4 Department of Dermatology, University Hospital "Santa Maria della Misericordia", Udine, Italy

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Corresponding author: Murat Durdu, Department of Dermatology, Başkent University Faculty of Medicine, Adana Dr. Turgut Noyan Application and Research Center, Adana, Turkey. Tel: +90 2422406710 Fax: +90 2422496040 Email: sivandr@hotmail.com

Case Presentation

A 63-year-old male presented with pruritic, firm, dome-shaped, skin-colored/whitish papules mainly located over the forehead, neck, elbows, hands and feet (Figure 1A) that had progressively increased in number over the last year; skin induration of the trunk without papular lesions was also evident on palpation. Dermoscopic examination of the papules showed round/oval, homogenous, white-ivory structureless areas similar to "rice grains" with no vessels (Figure 1B) [1], while cytological assessment of slit-skin smear taken from the lesions revealed round fibrotic collagen structures and mucinous materials (Figures 1, C and D). Based on clinical, dermoscopic and cytological findings, a possible diagnosis of scleromyxedema was made, and a biopsy was taken for histological examination, that confirmed this hypothesis by revealing fibroblast proliferation, collagen deposition, perivascular lymphoplasmocytic infiltration, and mucin deposition in the dermis (Figures 1, E and F). Laboratory tests showed monoclonal gammopathy, while no systemic involvement was detected on further examination. Intravenous immunoglobulin therapy (2 g/kg dose for 5 consecutive days per month) was started, with significant improvement after three cycles of treatment.

Teaching Point

Scleromyxedema is a form skin mucinosis with possible extra-cutaneous involvement, including neurological, renal, hematological, and rheumatological, that may carry a poor prognosis if not treated timely [2]. Diagnosis is generally clinical, yet in initial phases/incomplete instances it may be challenging to differentiate from similar conditions (eg lichen planus, lichen amyloidosis, papular lichen simplex chronicus, papular granuloma annulare, multiple follicular adnexal tumors) that, however, show a different

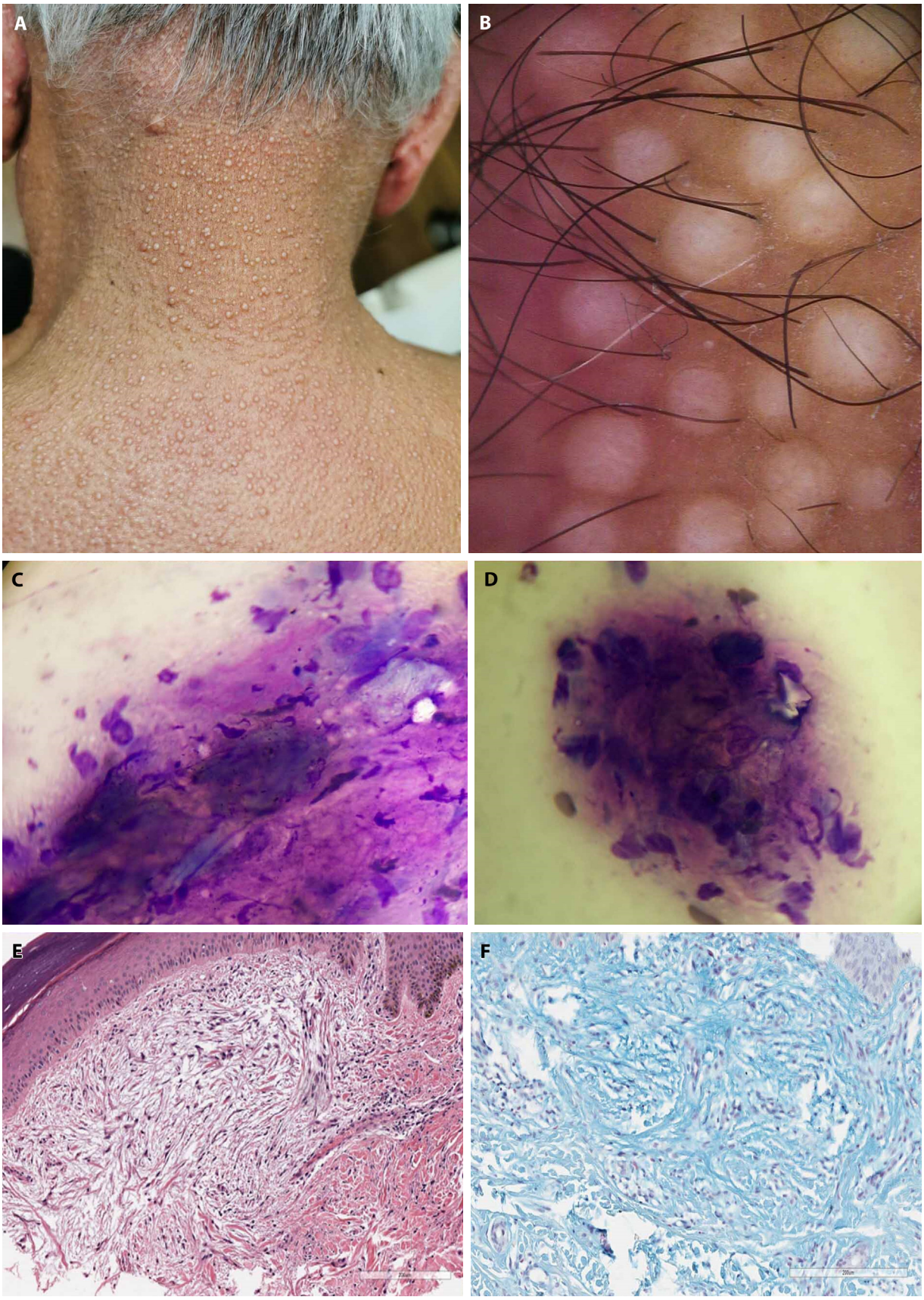

Figure 1. (A) Dome-shaped, firm, small papules on the nape are similar as seen on clinical examination. (B) Dermoscopy reveals round and oval-shaped white-ivory homogenous areas similar to rice grains (magnification x10) [1]. (C,D) Cytology shows round fibrotic collagen structures and mucinous materials (May-Grünwald Giemsa x1000). (E) Histopathological examination displays increased mucin deposition in the superficial dermis and stellated fibroblasts between collagen fibers (H&E x200). (F) Fragmented elastic fibers are also evident (Alcian blue stain x200).

dermoscopic and cytological patterns [3]. Therefore, the use of such techniques may increase the index of suspicion for scleromyxedema with consequent prompt treatment.

References

1. Mendes Bastos P, Borges AS, Cardoso JC, Oliveira A. Dermoscopy and reflectance confocal microscopy for the diagnosis of scleromyxedema. *JAAD Case Rep*. 2019;5(5):451-453. Published 2019 May 8. doi:10.1016/j.jdcr.2019.03.005
2. Ferreli C, Gasparini G, Parodi A, Cozzani E, Rongioletti F, Atzori L. Cutaneous Manifestations of Scleroderma and Scleroderma-Like Disorders: a Comprehensive Review. *Clin Rev Allergy Immunol*. 2017;53(3):306-336. DOI: 10.1007/s12016-017-8625-4. PMID: 28712039.
3. Lallas A, Errichetti E, Ioannides D. *Dermoscopy in General Dermatology*. CRC Press, Boca Raton, FL, 2018.