Cutis verticis gyrata – A rare presentation of Primary systemic Amyloidosis

Sir,

A 35-year-old female presented in the Department of Dermatology with asymptomatic skin-colored swellings and loss of hair over the entire scalp since 3–4 years. On clinical examination, multiple, skin colored, waxy cerebriform nodules and plaques that were coalescing in nature and diffusely involve the whole scalp with alopecia. Similar lesions were present in periorbital region and nipple-areola complex [Figure 1]. Macroglossia and periorbital purpura were also present. Nails and mucosae were normal. Based on history and clinical examination, clinical diagnosis of secondary cutis verticis gyrata (CVG) was made. Macroglossia and periorbital waxy skin-colored nodules with purpura pointed toward amyloidosis. Further, the patient was evaluated thoroughly to rule out other causes of secondary CVG. On investigation, her complete blood count showed low hemoglobin and rest was normal. Renal functions test was also deranged with raised blood urea and serum creatinine. Electrocardiography, chest X-ray, X-ray skull, thyroid function tests, venereal disease research laboratory, and ultrasound abdomen were normal. Magnetic resonance imaging head showed diffuse irregular nodular thickening of scalp layers without any intracranial lesions and bony abnormalities. On histopathological examination, accumulation of homogeneous eosinophilic material in the dermis was seen in Figure 2. Congo red stain was positive and apple-green birefringence was seen on polarizing microscopy. Thus, features were suggestive of amyloidosis. Bone marrow examination, serum and urine electrophoresis were found to be normal. The patient was diagnosed as a case primary systemic amyloidosis (PSA) and was referred to medicine for further management.

CVG is a rare congenital or acquired condition of scalp, characterized by convoluted folds and deep furrows that resemble the surface of cerebral cortex. It may be primary or secondary. Secondary CVG are associated with other diseases such as acromegaly, myxedema, hamartomatous lesions, tumors, idiopathic hypertrophic osteoarthropathy, syphilis, leukemia, cretinism, tuberous sclerosis, neurofibromatosis, acanthosis nigricans, Ehlers–Danlos syndrome, and trauma. Primary forms are those which are not associated with any underlying cause.[1]

Amyloidosis is one of the causes of secondary CVG. Amyloidosis can be classified as localized or systemic. Localized amyloidosis is the deposition of amyloid in only one organ. Systemic amyloidosis can be primary or secondary. PSA can be idiopathic or myeloma associated. Secondary systemic amyloidosis is associated with chronic inflammatory disorders.[2] Ours is a case of PSA as it was not associated with any chronic inflammatory disease.

Systemic amyloidosis shows cutaneous involvement is about 50% of patients.[3] The manifestations of
Letters to Editor

International Journal of Trichology / Volume 10 / Issue 3 / May‑June 2018

Cutaneous amyloidosis depend on the site of amyloid deposition. Deposition around pilosebaceous glands leads to the destruction of hair and alopecia. Amyloid deposition in scalp skin results in enlargement of skin which gets thrown into convoluted folds and furrows resembling CVG. CVG may be a rare presenting feature for PSA as in our case. Thus, dermatologist facing this condition must have high index of suspicion for the diagnosis of such cases, as primary systemic amyloidosis can seldom present as cutis verticis gyrata at the initial stages.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Priyadarshini Sahu, Surabhi Dayal, Geetika Gera, Ashish Amrani

Department of Dermatology, Venereology and Leprology, Pt B D Sharma, University of Health Science, Rohtak, Haryana, India

Address for correspondence:
Dr. Priyadarshini Sahu, Department of Dermatology, Venereology and Leprology, Pt B. D. Sharma, University of Health Sciences, Rohtak, Haryana, India.
E-mail: priyadarshini.sahu.9@gmail.com
REFERENCES

1. Koregol S, Yatagiri RV, Wanad SR, Itagi NR. A rare association of scleromyxedema with cutis verticis gyrata. Indian Dermatol Online J 2016;7:186-9.
2. Saoji V, Chaudhari S, Gohokar D. Primary systemic amyloidosis: Three different presentations. Indian J Dermatol Venereol Leprol 2009;75:394-7.
3. Schreml S. Cutaneous Amyloidoses. In: Griffiths CEM, Barker J, Bleiker T, Chalmers R, Creamer D, editors. Rook's Textbook of Dermatology, 9th ed. UK: Wiley-blackwell Publications; 2010. p. 58.1-32.

How to cite this article: Sahu P, Dayal S, Gera G, Amrani A. Cutis verticis gyrata – A rare presentation of primary systemic amyloidosis. Int J Trichol 2018;10:141-3.
© 2018 International Journal of Trichology | Published by Wolters Kluwer - Medknow

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.