

the disease may remain stable for considerable period of time and spontaneous improvement may occur.

Before therapeutic bronchoalveolar lavage was introduced by Ramirez RJ *et al.* in 1965(6), patients with PAP were treated with corticosteroids, heparin, antibiotics, trypsin and pancreatic enzymes with little success. The treatment of PAP consists of therapeutic bronchoalveolar lavage. The lavage can be accomplished safely in children and adults and with relative ease by using double lumen endotracheal tube or catheter to isolate and lavage one lung while ventilating the other(7). In very young children the same can be accomplished by using double lumen catheter and rigid bronchoscope. The therapeutic efficiency of bronchoalveolar lavage is primarily due to the mechanical removal of intra alveolar phospholipids. Most patients who undergo whole lung lavage for PAP that is unassociated with pulmonary fibrosis experience an improvement in pulmonary function and in exercise performance. The reported complications associated with PAP are severe pulmonary fibrosis followed by respiratory failure and death, formation of emphysematous bullae followed by pneumothorax and usual complication of sudden asphyxia and death due to flooding of the airways by thick alveolar material during anesthesia(2).

Acknowledgements

The authors wish to thank Dr. (Mrs)P.M. Pai, Dean, Seth G.S. Medical College and King Edward VII Memorial Hospital for allowing them to publish this case report.

REFERENCES

1. Rosen SH, Castleman B, Libow AA. Pulmonary alveolar proteinosis. *N Engl J Med* 1958, 258: 1123-1125.
2. Prakash UBS, Barham SS, Carpenter MA, *et al.* Pulmonary alveolar proteinosis: Expe-

rience with 34 cases and a review. *Mayo Clin Proc* 1987, 62: 499-518.

3. Harris D Riley Jr. Pulmonary alveolar proteinosis. *In: Disorders of the Respiratory Tract in Children*, 5th edn. Eds Chernick V, Kendig EL Jr. Philadelphia, WB Saunders Co, 1990, pp 492-495.
4. Coleman M, Dehner LP, Sibley RK, *et al.* Pulmonary alveolar proteinosis: An uncommon cause of chronic neonatal respiratory distress. *Am Rev Respir Dis* 1980, 121: 583-586.
5. Smith LJ, Ankin MG, Katzenstein AN, Shapiro BA. Management of pulmonary alveolar proteinosis. *Chest* 1980; 68: 765-769.
6. Ramirez RJ, Kieffer RF Jr, ball WC Jr. Bronchopulmonary lavage in man. *Ann Intern Med* 1965, 63: 819-828.
7. Moazam F, Schmidt JH, Chesrwon SE, *et al.* Total lung lavage for pulmonary alveolar proteinosis in an infant without use of cardiopulmonary bypass. *J Pediatr Surg* 1985, 20: 398-401.

INFLAMMATORY LINEAR VERRUCOUS EPIDERMAL NEVUS

P.S. Umap
R.W. Bodade

Inflammatory linear verrucous epidermal nevus (ILVEN) is a rare skin disease of

From the Department of Pathology, Government Medical College Nagpur.

Reprint requests: Dr. Pradeep, S. Umap, 'Shri' 58, Dharampeth Society, 5th Lay Out, Jaiprakash Nagar, Nagpur 440 025, Maharashtra.

*Received for publication: November 20, 1992;
Accepted: January 29, 1993*

early childhood(1,2). For the first time, it was delineated by Altman and Mehregan in 1971(2). A similar case has been reported from India in 1989 by Gharpuray *et al.*(3). Prior to this, there had been no reports in the Indian literature(3). We are reporting a case of ILVEN in a 6-year-old girl with relevant literature.

Case Report

A 6-year-old girl was brought with the complaints of pruritic, raised, reddish lesions over left lower and upper extremities of three years duration. Patient was alright three years back, then lesions developed on left lower leg and during three years time appeared on the forearm and hand. She gave history of gradual increase in the size of the lesion. Local examination revealed multiple, hyperpigmented, scaly papular lesions on left side of body involving leg, extensor surface of forearm and hand. These lesions were distributed in a linear fashion.

Histopathology of the lesion on left forearm showed broad, circumscribed columns of parakeratosis originating from the epidermis and the remaining stratum corneum showed compact hyperkeratosis. It also showed papillomatosis. There was a perivascular lymphocytic infiltrate in upper dermis (*Fig. 1*).

Discussion

ILVEN usually starts to develop at birth or shortly thereafter in the form of persistent, reddish, slightly verrucous, scaly papular lesions(1,2,4). Exceptionally, it can be seen in adults(5). It is distributed in a linear or zosteriform pattern and is unilateral so labelled as *nevus unius lateris*(1).

Clinically, the disease shows female predominance with the ratio of 4 : 1(2). The most common location of the lesion is on the



Fig. 1. ILVEN showing broad, circumscribed column of parakeratosis and compact hyperkeratosis of remaining stratum corneum. Upper dermis shows perivascular lymphocytic infiltrate.

lower extremities especially on left side of the body(1,2,4). The condition must be differentiated from other examples of *nevus unius lateris* and from linear psoriasis, linear lichen planus and lichen striatus(1). The lesions of ILVEN are mostly pruritic and persistent which differentiates it from lichen striatus(2,4). Histologically, lichen striatus tends towards a lichenoid pattern and ILVEN towards a psoriasiform pattern(4). The Parakeratosis and ILVEN are diseases that involve the epidermis predominantly but also the papillary dermis to some extent. The various forms of parakeratosis and the ILVEN have in common well circumscribed zones of parakeratosis beneath which the granular layer is absent and a lymphohistiocytic infiltrate in upper dermis(1).

The microscopic picture of ILVEN is essentially that of a non-specific chronic dermatitis(6). It is characterized by moderately well circumscribed, short and broad columns of parakeratosis, arising from epidermis and are perpendicular to the skin surface usually at the summits of epidermal papillation. Between the areas of parakeratosis the stratum corneum is hyperkeratotic in compact patterns. Beneath the zones of parakeratosis, granular layer and dyskeratotic or vacuolated are absent and beneath the areas of orthokeratosis, granular layer is thickened. The epidermis is hyperplastic, and usually papillated and psoriasiform. Superficial perivascular lympho-histiocytic infiltrate is seen in the dermis(1,3,4,7-9). In a few lesions, extensive parakeratosis is seen(2,5).

Older way of treatment in the form of topical corticosteroids, surgical excision or cryosurgery was not satisfactory. Considerable improvement in the lesion and delayed recurrence of inflammatory episodes were observed by local application of potent corticosteroids like betamethasone dipropionate(3).

Acknowledgements

The authors thank the Dean and Head of the Department of Pathology, Government Medical College, Nagpur for permitting them to publish this case.

REFERENCES

1. Ackerman AB, Niven J, Grant-Kels JM: Differential Diagnosis in Dermatopathology. Philadelphia, Lea and Febiger, 1982, pp 94-97.
2. Altman J, Mehregan AH. Inflammatory linear verrucous epidermal nevus. Arch Dermatol 1971, 104: 385-389.
3. Gharapuray MB, Kulkarni V, Sule RR. Inflammatory linear verrucous epidermal nevus,

Indian J Dermatol Venereol Leproal 1989, 55: 64-65.

4. Lever WF, Schaumburg-Lever G. Histopathology of the Skin, 7th edn. Philadelphia, JB Lippincott, 1990, p 176.
5. Hodge SJ, Barr JM, Owen LG. Inflammatory linear verrucous epidermal nevus. Arch Dermatol 1978, 114: 436-438.
6. Kaidbay KH, Kurban AK. Dermatitic epidermal nevus. Arch Dermatol 1971, 104: 166-171.
7. Dupre A, Christol B. Inflammatory linear verrucous epidermal nevus. Arch Dermatol 1977, 113: 767-769.
8. Landwehr AJ, Starink TM. Inflammatory linear verrucous epidermal nevus. Dermatologica 1983, 166: 107-109.
9. Toribio J, Quinones PA. Inflammatory linear verrucous epidermal nevus. Dermatologica 1975, 150: 65-69.

CYSTIC KIDNEYS IN TUBEROUS SCLEROSIS

S.R. Ranade
P.A. Amanapure
D.V. Raichur
P.G. Patil

Tuberous sclerosis is a neurocutaneous disorder which in addition to the skin and brain affects the kidneys, heart, eyes, lungs and bones. Nearly 50 to 80% of the patients with tuberous sclerosis have renal angio-

From the Department of Pediatrics, Wanless Hospital, Miraj.

Reprint requests: Dr. (Mrs) Shashikala R. Ranade, Head of Department of Pediatrics, Wanless Hospital, Miraj 416 410.

Received for publication: July 20, 1992;

Accepted: February 3, 1993