

Angiokeratoma Circumscriptum Neviforme

A 10 years-old boy presented with multiple small bluish-red papules on the scrotum for the past 2 years. These lesions were gradually increasing in number and had a tendency to bleed after minor trauma. On examination, multiple discrete smooth surfaced bluish-red papules were found unilaterally on scrotum. (**Fig. 1**) Palpation ruled out varicocele or inguinal hernia. Rest of the mucocutaneous examination was unremarkable. Skin biopsy from a papule showed mild hyperkeratosis with large, numerous, dilated capillaries in papillary dermis. USG pelvis and scrotum were done to rule out any vascular anomaly. Considering clinical feature and histopathology, the patient was diagnosed as angiokeratoma circumscriptum neviforme (ACN).

Angiokeratomas are characterized by asymptomatic, 2-5 mm, blue red hyperkeratotic papules. Histologically, they are composed of ectatic thin-walled vessels in the superficial dermis with overlying epidermal hyperplasia. Angiokeratoma can be localized or generalized. The generalized form, angiokeratoma corporis diffusum, is usually associated with a metabolic disorder, the most common being Fabry disease. The localized forms are solitary angiokeratoma (typically occurs on the legs and follows trauma), localized angiokeratoma of the scrotum and vulva (Fordyce type), Mibelli type (bilateral angiokeratomas on the dorsa of the fingers and toes), and



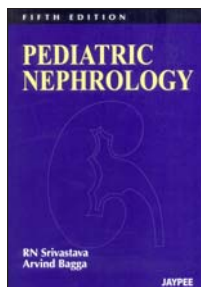
FIG. 1 Blue-red grouped papules present unilaterally on scrotum (A). Close up of lesions (B).

angiokeratoma circumscriptum neviforme (multiple, hyperkeratotic, papular and plaque like lesions, usually unilaterally on the lower leg, foot, and buttock). The major differential diagnosis is angiokeratoma Fordyce type (middle aged persons, bilateral lesions) and lymphangioma circumscriptum (yellowish grouped vesicles containing clear fluid). Herpes zoster and herpes simplex (short duration) too need to be differentiated. The principal problems are intermittent bleeding, anxiety, and over-treatment due to misdiagnosis. It is treated by various locally destructive modalities including electrocoagulation, excision, cryotherapy, or LASER therapy.

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BOOK REVIEWS



Pediatric Nephrology 5th Edition
RN SRIVASTAVA, ARVIND BAGGA
*Jaypee Brothers Medical Publishers
(P) Ltd; New Delhi: 2011.
Pages: 557; Price: 850/-*

This edition has been extensively revised and updated throughly. The chapters on electrolyte and acid base disorders and common kidney disorders of childhood like nephrotic syndrome, urinary tract infection, voiding disorders and neonatal renal disorders have been expanded. A new chapter on prevention of kidney diseases has been added. The edition is well illustrated with tables, figures and flow charts. The key points in text provide

essential features to the reader for clear take home message. The book is printed in such a way to enhance its visual appeal and make reading an enjoyable experience. The appendices have included very useful information. The book has proved its usefulness over the years. This edition has further enhanced its value in understanding and managing kidney disorders in children. The book is strongly recommended to postgraduate students, practicing pediatrician and faculty members.

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