Association of Reactive angioendotheliomatosis with intraosseous hemangioma - a rare case report

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INTRODUCTION

Reactive angioendotheliomatosis (RAE) is a rare benign angioproliferative condition of the skin, which has been noted to occur in patients with a variety of underlying systemic diseases. Histopathologically, this condition is characterized by dermal vascular proliferation, and endothelial cell hyperplasia within the lumina and around dermal vessels, without significant cellular atypia. (1) The pathophysiology of RAE and the other reactive angiomatoses is unclear however the role of hypoxia and vessel occlusion has been suggested to play a role in development of these conditions (2)

There are various case reports of RAE occurring in patients with coexisting systemic diseases, including autoimmune connective tissue diseases, peripheral vascular disease, chronic infections (especially bacterial endocarditis), graft versus host disease and even sarcoidosis. We hereby describe a case of RAE arising in association with an intraosseous hemangioma.

CASE REPORT

A 42 yr old female presented with chief complaint of multiple reddish papules and nodules over right dorsum of hand for past 4-5 months back (fig.1a). Initially patient developed pain along with swelling on the dorsum of right hand following which cast application was done by a local practitioner for 1 and half month. After removal of the cast she noticed single reddish pea sized papule which was asymptomatic in nature.

She was then treated by an orthopedician with ATT for 15 days on clinical suspicion of tuberulosis but was stopped with no response. During this period patient gradually developed more papules which gradually increased in size in 1-2 month but were asymptomatic. There was no history of any other systemic comorbidity. Skin biopsy was performed which revealed dermal vascular proliferation, and endothelial cell hyperplasia within the lumina with CD34 positivity (fig.1b).

NCCT revealed a lytic lesion involving the diaphysis and metaphysis of fifth metacarpal. Excision biopsy showed an encapsulated lesion infiltrating the cortical bone and soft tissue, composed of numerous proliferating blood vessels (fig.2a) confirmed on CD-34 positivity (fig.2b). Following the hemangioma excision, cutaneous lesions resolved completely in 2 week interval.

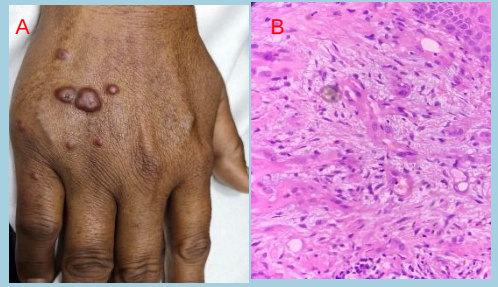

Fig-.1 A- Multiple erythematous papules as well as nodules over dorsum of hand
B- Dermal vascular proliferation with endothelial cell hyperplasia

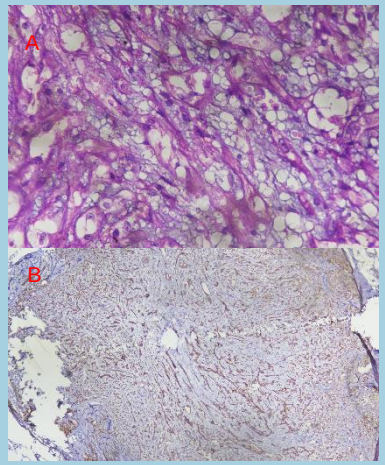

Fig-2 A- Numerous proliferating vessels. B- CD34 positivity

DISCUSSION

Reactive angioproliferations are benign vascular disorders of the skin characterized histologically by intra-vascular and extra-vascular hyperplasia of endothelial cells and pericytes.(3) They include reactive angio-endotheliomatosis, acro-angiodermatitis (pseudo-Kaposi sarcoma),diffuse dermal angiomatosis, intravascular histocytosis, glomeruloid angioendotheliomatosis and angio-pericytomatosis (angiomatosis with cryoproteins).

All lesions are characterized histologically by a proliferation of capillaries in the dermis, with variably diffuse, lobular or mixed lobular and diffuse patterns. There is marked intercase and intracase heterogeneityin histologic features. Common features include fibrin microthrombi ,reactive (fasciitis-like) dermal alterations and foci of epithelioid endothelium. (1)

CONCLUSIONS

Our impression was that RAE seemed to be poorly recognized, probably because of its rarity and also its clinical and histologic mimicry of other vascular lesions. We have therefore made an attempt to report a unique case of RAE occurring in association with intraosseous hemangioma.

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