

Angiokeratoma Circumscriptum of the Oral Cavity

Sir,

The term angiokeratoma is applied to several distinct unrelated conditions with cutaneous vascular lesions, the histology of which shows superficial dermal vascular ectasia with overlying hyperkeratosis of the epidermis. Different types of angiokeratomas have been described. They are (i) the generalized systemic type – angiokeratoma corporis diffusum of Fabry; (ii) the bilateral form occurring in the dorsa of fingers and toes – angiokeratoma of Mibelli; (iii) the localized scrotal form – angiokeratoma of Fordyce; (iv) the usually solitary papular angiokeratoma; and (v) the multiple papular and plaque like – angiokeratoma circumscriptum (1). Though all types differ clinically they share similar histological features. They are characterized by hyperkeratosis, acanthosis and dilated capillaries in the papillary dermis, partly or completely enclosed by the papillomatous epidermis. Organized or organizing thrombi are occasionally observed within the dilated capillaries. Moreover, in Fabry's disease and fucosidosis, swollen and vacuolated endothelial cells are also identified in the vascular spaces. Angiokeratoma circumscriptum presents as multiple purple papules that later become verrucous and may coalesce to form plaques. The lesions are situated typically on the lower leg or foot but can occur on the thigh, buttock or occasionally elsewhere. In this report, we describe a case



Fig. 1. Ventral aspect of tongue showing grouped shiny papular lesions.

of angiokeratoma circumscriptum of the tongue that we believe is probably the first case reported in the literature.

CASE REPORT

A 16-year-old male presented with multiple pink-coloured small raised lesions on the tongue. He had had them for the past one year. The condition started as a single raised lesion on the undersurface of the tip of the tongue which gradually increased in number and extended onto the upper surface of the tongue. There was no preceding history of local trauma. He gave history of bleeding from the lesion, but denied any history of similar lesions elsewhere on the body. His past medical history was unremarkable.

On examination, there were multiple, grouped, erythematous, shiny papules, some of which had a keratotic top seen mostly over the ventral aspect of the anterior two-thirds of the tongue (Fig. 1), while some were seen on the dorsal aspect too. They were mobile, firm on palpation and did not bleed on manipulation. The rest of the cutaneous and systemic examination was normal.

A biopsy of the representative tongue lesion showed parakeratosis, acanthosis and papillomatosis with large dilated spaces lined by normally appearing endothelium and filled with erythrocytes and organizing thrombi. A few of the single cell-layered endothelium-lined spaces were clear. These histological features were consistent with that of angiokeratoma circumscriptum.

DISCUSSION

Oral mucosal involvement is most commonly a component of angiokeratoma corporis diffusum (Fabry's disease and fucosidosis). Mucosal involvement is otherwise uncommon in other types of angiokeratomas. To date, we were able to trace only a single case of solitary angiokeratoma of the oral cavity in the literature (1).

REFERENCES

1. Leung CS, Jordan RCK. Solitary angiokeratoma of the oral cavity. *Oral Surg Oral Med Oral Pathol Endod* 1997; 84: 51 – 53.

Accepted April 30, 1998.

M. Vijai Kumar¹, Devinder M. Thappa¹, Srikanth Shanmugam¹ and Chaganti Ratnakar²

Departments of , ¹Dermatology & STD and, ²Pathology and , Jawaharlal Institute of Postgraduate Medical Education and Research (JIPMER), Pondicherry 605 006, India.