Letters to the Editor

Combined planar and eruptive xanthoma in a patient with type IIa hyperlipoproteinemia

Sir,

Lipoproteins are soluble compounds formed by the combination of insoluble circulating lipids (cholesterol, cholesterol esters, triglycerides and phospholipids) and proteins. Any disorder of lipoprotein metabolism (dyslipidemia) [Table 1] confers on an individual, an increased risk of cardiovascular disease, pancreatitis or xanthoma.

Xanthomas are the characteristic cutaneous presentation in hyperlipoproteinemia. Type IIa hyperlipoproteinemia or familial hypercholesterolemia usually presents with tendinous, tuberous or planar xanthomas (xanthelesma or intertriginous type). We were unable to find previous reports of eruptive xanthomas occurring in association with a planar xanthoma in Type IIa hyperlipoproteinemia.

A 4-year-old girl, adopted from distant relatives, born out of a nonconsanguinous marriage presented skin lesions for one year. The lesions appeared as skin colored papules around the ankle and natal cleft one year back. They were progressive in nature, with a sudden eruption of new lesions over the buttocks, knees and thighs for four weeks. The

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Hyperchylomicronemia</td>
</tr>
<tr>
<td>IIa</td>
<td>Elevated LDL (familial hypercholesterolemia)</td>
</tr>
<tr>
<td>IIb</td>
<td>Elevated LDL and VLDL (familial combined hypercholesterolemia)</td>
</tr>
<tr>
<td>III</td>
<td>Broad β-VLDL (Familial dysbetalipoproteinemia)</td>
</tr>
<tr>
<td>IV</td>
<td>Elevated VLDL (Familial hypertriglyceridemia)</td>
</tr>
<tr>
<td>V</td>
<td>Elevated chylomicrons and VLDL (mixed hyperlipidemia)</td>
</tr>
</tbody>
</table>

WHO: World Health Organization, LDL: Low density lipoprotein, VLDL: Very low density lipoprotein

Table 1: WHO/Fredrickson's classification of hyperlipoproteinemia/ hyperlipidemia
child was developmentally normal for her age. Cutaneous examination revealed multiple skin colored to yellowish flat topped plaques over the ankle, first metatarsophalangeal joint, natal cleft and gluteal fold. Yellowish papules arranged in a linear fashion were noted over the ankle [Figures 1 and 2]. Grouped skin colored papules were also seen over the thighs, knees and buttocks [Figure 3]. The general physical examination was normal. Hematological investigations were as follows: hemoglobin 8.8 gm/dL, total leucocyte count 10,300 cells/mm$^3$, differential count neutrophils: 51, lymphocytes: 30, eosinophils: 13, basophils: 1, monocytes: 5. The results of serum lipid assay appear in Table 2. The cardiovascular and the ophthalmological examinations were normal. Other biochemical parameters like blood sugar, renal and hepatic function tests were within normal limits. Radiography of the chest and ultrasound of abdomen and pelvis was normal. The histopathological examination revealed a diffuse dense nodular infiltrate of foamy histiocytes involving the dermis. The histiocytes were large and polygonal, at places touching each other and at other places extending interstitially into the surrounding reticular dermis [Figure 4]. A diagnosis of hypercholesterolemia (WHO type IIa) in association with planar and eruptive xanthoma was made and the child was treated with atorvastatin, 10 mg daily along with iron and folic acid supplementation. There was moderate flattening of lesions over the ankle after one month.

Xanthomas are plaques or nodules consisting of accumulation of lipid-rich macrophages known as foam cells.$^{[1,2]}$ They do not represent a disease but rather are symptoms of different lipoprotein disorders or arise without an underlying metabolic defect. Clinical presentations of xanthomas include eruptive, tuberous, tubero eruptive, tendinous, planar, verruciform and papular forms.$^{[2]}$

Planar xanthomas are wide-based yellowish macules or plaques found commonly on the upper eyelids (xanthelasma palpebrum/xanthelasma), palms (xanthoma striatum palmare), intertriginous regions, and diffuse planar xanthomas.$^{[3]}$ Xanthoma striatum palmare is the characteristic cutaneous feature of dysbetalipoproteinemia (WHO type III)$^{[3]}$. About 60% of patients with xanthelasma palpebrum have an association with hypercholesterolemia.$^{[1]}$ An intertriginous planar xanthoma is pathognomonic of homozygous(typeIIa) familial hypercholesterolemia.$^{[2]}$ A similar finding was seen in our case. In addition, lesions were also seen in areas of trauma (Koebner's
of increased numbers of E-selectin-positive endothelial cells and a decrease in the intracellular cell adhesion molecule (ICAM)-1 cells promotes macrophage migration into xanthoma lesions. The extravasated low density lipoprotein (LDL) can also recruit more macrophages and in association with factors like heat, movement and friction increase capillary leakage of LDL. These local factors help explain the location of tuberous xanthomas, tendinous xanthomas, and xanthelasma.[8]

Histologically, eruptive xanthoma is characterised by infiltration of the dermis with neutrophils, eosinophils and histiocytes with foam cells along with extravascular lipid deposits in the form of lace-like eosinophilic material between the collagen bundles. Planar xanthomas are composed of small groups and streaks of foam cells in the upper dermis and sometimes around pilosebaceous follicles with some inflammatory cells.[2] The differential diagnosis of eruptive xanthomas are sarcoidosis, granuloma annulare or xanthoma disseminatum.[3]

Statins are effective in the treatment of Type IIa hyperlipoproteinemia. They inhibit HMG CoA reductase, a rate-limiting enzyme in cholesterol synthesis. The other treatment modalities include lifestyle modifications like regular exercise and avoidance of smoking.[1,3]

In conclusion, though specific patterns of xanthomas are associated with different types of hyperlipoproteinemias, overlap in the clinical patterns can occur as seen in our patient.

Varadraj V. Pai, Pankaj Shukla, Mayur Bhobe
Department of Dermatology, Goa Medical College, Goa, India

Address for correspondence: Dr. Varadraj V. Pai,
Department of Dermatology, Goa Medical College,
Goa - 403 202, India.
E-mail: docpai@rediffmail.com

REFERENCES

4. Bhagwat PV, Tophakhane RS, Kudligi C, Noronha TM, Thirunavukkarasu A. Familial combined hypercholesterolemia
Unilateral asymmetrical double Becker's naevus

Sir,

A 35-year-old healthy man presented with two asymptomatic hyperpigmented lesions confined to the right half of his torso. One was a well demarcated brownish patch on the right shoulder with irregular borders, approximately 15 × 10 cm in size, with multiple small islands of hyperpigmentation in the surrounding skin (Figure 1 inset). The other lesion, extending from the right lower scapular area to the lateral chest wall, approximately 7 × 15 cm in size, showed a similar morphology with the additional feature of hypertrichosis (Figure 2). Both lesions had developed around the onset of puberty and became progressively darker; the shoulder lesion was noticed first and the other one appeared a year later. A clinical diagnosis of Becker's nevi was made.

General physical examination was within normal limits. We did not find any associated musculoskeletal changes or acneiform lesions. Genital and limb examination were normal. X-ray spine and ultrasound of the abdomen and scrotum did not reveal any abnormality. KOH examination of skin scrapings from the lesion was negative for pityriasis versicolor. Skin biopsy was performed from both lesions. Histopathology revealed epidermal acanthosis along with regular elongation of rete ridges. Increased pigmentation of the basal layer was seen without increase in the number of melanocytes. Some melanophages were seen in the dermis. Based on the clinical and histopathological findings, a diagnosis of Becker's nevi was made.

In 1949, Samuel William Becker first described a "concurrent melanosis and hypertrichosis in the distribution of nevus unius lateris" which has since been termed Becker's nevus. Becker's nevus commonly manifests in peri-pubertal males as a unilateral, solitary, acquired localized hyperpigmented patch composed of coalescing brownish macules. Hyperpigmentation usually increases for the first 2-3 years while hypertrichosis always appears after the pigmentation. However, the non-hypertrichotic variant is more common. "Progressive cribriform and zosteriform hyperpigmentation" may represent the non-hypertrichotic variant of Becker's nevus. There are a few documented cases with multiple and bilaterally symmetrical Becker's nevi; multiple and unilateral lesions are reported to occur much more rarely. Two Becker's nevi on the left side of the face in a segmental distribution with extension onto the oral mucosa have also been reported.