

Case Report

Normolipemic papular xanthoma with xanthelasma

Aman Dua*, Alka Dogra*, Neena Sood**, Saloni Sachdeva*, Atul Singla**

* Department of Dermatology, Dayanand Medical College&Hospital, Ludhiana, Punjab, India.

** Department of Pathology, Dayanand Medical College & Hospital, Ludhiana, Punjab, India.

Abstract Xanthomas are a common presentation of disorders of lipid metabolism usually associated with abnormalities of cholesterol metabolism. A 35 year old female presented to us with the lesions of xanthelasma on the upper eyelids and papular xanthomas on the rest of the body. Routine investigations and systemic examination was normal. Lipid profile was within normal range and serum protein electrophoresis showed normal pattern. Histopathology from a papular lesion showed foamy histiocytes with Touton's type of giant cells. We present a case of normolipemic papular xanthoma with xanthelasma which is a very rare occurrence.

Key words

Xanthelasma, papular xanthoma, normolipemic.

Introduction

Xanthomatosis is a widespread disorder in which localized infiltrates of the lipid containing cells appear in the skin and visceral organs. They may or may not be associated with abnormal serum lipid metabolism.¹ Xanthelasma palpebrarum is the most common cutaneous xanthoma and occurs as bilaterally symmetrical yellow plaques seen most commonly near the inner canthus of the upper eyelid. They may represent a localized cutaneous phenomenon but may signify a systemic hyperlipidaemia and may be associated with elevation of LDL as in pure hypercholesterolaemia (such

as FH) or type III hyperlipoproteinaemia.² Papular xanthoma is a rare normolipemic non Langerhan's cell histiocytosis affecting both children and adults. Clinically, it is represented by monomorphic papular eruption seen in the age group of 13 to 57 years and has a biphasic occurrence in the young adolescence and in the middle ages. It is predominately located on the trunk, extremities and rarely on the head. Histologically, it is characterized by a regular epidermis and a dense distribution of the xanthomized macrophages interspersed by numerous Touton's type giant cells.³ Papular xanthoma may be associated with a systemic disorder or may be a cutaneous manifestation of the underlying malignancy. Here we describe a female patient with xanthelasma and normolipemic papular xanthoma unassociated with any systemic disease.

Address for correspondence

Dr. Aman Dua, Asisstant Professor,
Department of Dermatology,
Dayanand Medical College & Hospital,
Ludhiana, INDIA
Email: dramandua@yahoo.co.in,
atulsingla14@rediffmail.com
Ph # 91-9872808301, 91-9872857624

Case report

A 35-year-old female presented in our outpatient department with history of yellowish papular lesions on the arm, face and upper trunk for the last 6-7 years. The lesions first appeared as small, slightly elevated plaques which were well-circumscribed on the inner canthi of the upper eyelids (**Figure 1** and **2**). After 2 years, papular lesions appeared on the rest of the face, head, back, arms, legs, posterior part of the thighs and axillae. Rest of the physical examination was negative except for the skin lesions. Routine investigations including complete hemogram and urine analysis were normal. Lipid profile was within normal limits. Serum bilirubin was normal and serum protein electrophoresis showed normal pattern.

Histopathology Biopsy of the skin lesion of the neck showed localized collection of histiocytes with foamy vacuolated cytoplasm, few lymphocytes and neutrophils in the dermis. Touton's type giant cells were also noted (**Figure 3**)

Diagnosis Based upon the clinical profile and histopathological changes, diagnosis of



Figure 1 Pretreatment xanthelasma and papular xanthoma on face.



Figure 2 Papular xanthoma over face and neck.

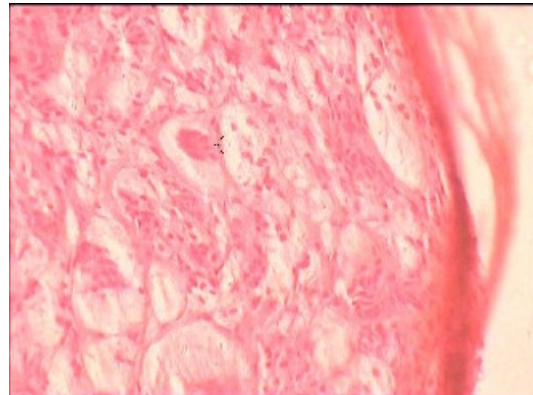


Figure 3 Microphotograph showing Touton's giant cells.



Figure 4 Posttreatment photograph of xanthelasma and papular xanthoma.

xanthelasma palpebrum with papular xanthoma was made. The patient was kept on follow up and repeat lipid profile after one month showed almost similar values.

Treatment Trichloroacetic acid was applied on xanthelasma lesions and cryotherapy with nitrous oxide was done for papular xanthoma along with oral antioxidants. Patient is responding well to the treatment. The skin lesions are healing and she is still on the follow up (**Figures 4**).

Discussion

We present a rare case of normolipemic papular xanthoma with xanthelasma in young female. Xanthomas are lesions characterized by accumulation of lipid-laden macrophages. They can be a reflection of lipid metabolism alteration or a result of local cell dysfunction. Xanthelasma palpebrum is the most common of the xanthomas and present as asymptomatic, usually bilaterally symmetric soft, velvety, yellow, flat, polygonal papules around the eyelids. Xanthelasmas are most common in the upper eyelid near the inner canthus. Usually, the lesions evolve for several months and enlarge slowly from a small papule. Xanthelasma may be associated with hyperlipidemia in which any type of primary hyperlipoproteinemia can be present. Some secondary hyperlipoproteinemias, such as cholestasis, may also be associated with xanthelasma.⁴

Papular xanthomas are normolipemic, nonconfluent, eruptive xanthomas located on the face, trunk and mucous membranes with no internal involvement. Histologically, they are characterized by foamy cells and

Touton's giant cells. Electron microscopy shows macrophages packed with free lipid vacuoles and lacking specific markers.⁵ Papular xanthoma has been reported to be associated with other systemic disorders and may be a cutaneous sign of internal malignancy. Diffuse normolipemic xanthomatosis have normal lipid levels but are often associated with serious hepatic disease or hematological dyscrasias especially multiple myeloma.⁶ Hu *et al.* reported two case of unusual normolipemic cutaneous xanthomatosis.⁷ One patient presented with widespread normolipemic papular xanthomas in which histiocytes containing Langerhan's granules were found. Vail *et al.* reported a case of chronic myelomonocytic leukemia with cutaneous xanthomas.⁸ Darwin *et al.* reported a case of generalized papular xanthoma in a patient with Sézary syndrome.⁹ The xanthomas were composed of foamy histiocytes that were shown by immunoperoxidase staining to be of the monocyte/macrophage lineage. Goerdts *et al.* described a case of normolipemic papular xanthomatosis in a patient with long standing erythrodermic atopic dermatitis.¹⁰ Papular xanthoma has also been reported to be associated with angiokeratoma of Fordyce.¹¹ Thus normolipemic xanthomatosis has been found to be associated with either a systemic disease or malignancy.

Our case showed xanthelasma with papular xanthomatosis but without any lipid disorder or associated systemic disease or malignancy. Normolipemic eruptive cutaneous xanthomatosis has been reported previously also but its occurrence is very rare.^{1,6} In our case two types of xanthomas were seen together in the same patient.

Though papular xanthomas have been reported to involute spontaneously,¹ in our case they were persistent for six years at the time when the patient first presented to us. This case is being reported because of its rare occurrence.

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