A Rare Case of Persistent Xanthoma Disseminatum without Any Systemic Involvement

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Sir,

Xanthoma disseminatum (XD) is a rare nonfamilial, histiocytic proliferative disorder involving skin and mucous membrane and frequently associated with diabetes insipidus. It is clinically characterized by asymptomatic, discrete, erythematous to yellow-brown papules, and nodules distributed symmetrically over the face, trunk, eyelids, flexural areas such as axillae, inguinal folds, antecubital, and popliteal fossae. Mucous membrane is involved in about 30% of cases. Systemic involvements such as the upper respiratory tract, skeletal, and the central nervous system are not uncommon. Based on the evolution and prognosis there are three clinical variants of XD, namely, (a) a self-healing form, (b) a persistent form (most common), and (c) a progressive form.\(^1\)

A 38-year-old male patient from nonconsanguineous parentage presented with multiple, asymptomatic, discrete, yellowish-brown, smooth-surfaced papules, and nodules distributed mostly around the mouth [Figure 1]. Similar lesions were also present over the nose, eyelids, axillae, inguinal areas, antecubital and popliteal fossae, and palms [Figure 2]. The lesions started around the mouth 7 months back and then gradually appeared over the other parts. Mucosal examination revealed multiple yellowish papules over the soft palate. There was no history of polydipsia, polyuria, visual impairment, bone pain, dyspnea, or dysphagia. Family history was noncontributory. Ophthalmologic and otorhinolaryngological workup were normal. Differential diagnoses, such as XD, eruptive xanthoma, histoid leprosy, post kala-zar dermal leishmaniasis, generalized eruptive histiocytoma, and multicentric reticulohistiocytosis were kept in mind before investigation.

Laboratory investigations - hemogram, erythrocyte sedimentation rate, blood sugar, liver function test, renal function test, lipid profile, urine analysis, and thyroid function test were normal. Ultrasonography, electrocardiography, and chest X-ray were also normal. Skin biopsy showed diffuse dermal infiltration of histiocytes, many foamy cells, Touton giant cells, and chronic inflammatory cells [Figure 3]. Immunohistochemistry (IHC) was positive for CD68 [Figure 4]. On the basis of clinical, histopathological, and IHC, the diagnosis of XD was reached.

The patient was treated with oral prednisolone 20 mg once daily and azathioprine 50 mg twice daily. There was no improvement in lesions after 3 months of treatment. He was lost to follow-up after 3 months of treatment.

XD is a rare normolipidemic, non-Langerhans cell histiocytosis occurring mostly in children and young adults. Involvement of the upper respiratory tract may manifest as dyspnea and stridor.\(^2\) Involvement of the conjunctiva and cornea may lead to blindness. Meningeal involvement is common, and infiltration to hypothalamus and pituitary may lead to diabetes insipidus in up to 40% of cases.
Histopathology showed a mixture of scalloped histiocytes, foamy cells, chronic inflammatory cells, and Touton giant cells. IHC showed CD68 and factor XIIIa positivity. XD patients have normal lipid levels, but secondary accumulation of cholesterol develop after primary proliferation of histiocytes. Reducing the proliferation of histiocytes and inhibiting subsequent lipid accumulation form the therapeutic goal in XD.

Azathioprine, prednisolone, and cyclophosphamide have shown good results. Combination of lipid lowering agents such as rosiglitazone, simvastatin, and fenofibrate has been reported to be effective in a normolipidemic individual with XD. Many therapeutic options such as surgical excision, laser ablation, electrodesiccation, radiotherapy, and systemic medications have been tried, but the treatment is challenging.

There was no response to azathioprine and prednisolone in our case, but disease was not progressive after 3 months of follow-up. Hence, we considered this to be persistent variety of XD.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.
Conflicts of interest
There are no conflicts of interest.

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