Title
Chronic myelomonocytic leukemia can present with diffuse planar xanthoma

Journal
Dermatology Online Journal, 20(7)

Authors
Guidry, Jacqueline
Thompson, Wendy
Sonabend, Michael

Publication Date
2014

Volume 20 Number 7
July 2014

Photo Vignette

Chronic myelomonocytic leukemia can present with diffuse planar xanthoma

Jacqueline Guidry BS, Wendy Thompson PA-C, Michael Sonabend MD

Dermatology Online Journal 20 (7): 17

[1]Baylor College of Medicine

[2]US Dermatology

Correspondence:

Jacqueline Guidry, M.D.
Baylor College of Medicine
602 Heights Boulevard
Apartment 8
Houston, Texas 77007
832-242-4113 (phone)
jacqueline.a.guidry@gmail.com

Abstract

There is a documented association between diffuse planar xanthoma and chronic myelomonocytic leukemia. Previous accounts have hypothesized that patients with multiple lesions or extensive cutaneous disease are more likely to have an underlying abnormality of the reticuloendothelial system. However, we document a 62-year-old woman with a large pruritic yellow-orange plaque on the chest and lower anterior neck consistent with planar xanthoma that was discovered to have chronic myelomonocytic leukemia. Solitary large plaques of planar xanthoma should be considered in the same fashion as diffuse planar xanthoma and warrant a prompt hematologic evaluation.

Keywords: Plane xanthoma, planar xathomatosis, chronic myelomoncytic leukemia

Case synopsis

A 62-year-old woman with a history of hyperlipidemia controlled with medication presented with a complaint of a pruritic, burning eruption on her neck and chest for the past two years, as seen in Figure 1. On exam, she was found to have yellow-orange ill-defined non-scaly mildly indurated plaques and patches on her chest and lower anterior neck. The differential diagnoses included xanthoma, necrobiosis lipoidica diabeticorum, and pseudoxanthoma elasticum.

A 4-mm punch biopsy was performed. Histopathologic examination showed an infiltrate of large pale foamy staining histiocytes in the dermis in a nodular aggregation consistent with a xanthoma as seen in Figure 2. Based on the clinical and pathologic findings, a diagnosis of planar xanthoma (PX) was made. Based on the diagnosis, further laboratory studies were ordered: urine protein electrophoresis, serum protein electrophoresis with immunofixation, lipids, basic metabolic panel, and complete blood count with differential. The laboratory results, listed in Table 1, showed normocytic normochromic anemia, thrombocytopenia, leukocytosis with left shift and blast-like forms. Serum protein electrophoresis showed polyclonal increase in gamma globulins with multiple oligoclonal banding suggesting a non-specific chronic inflammatory pattern.

Based on these findings, the patient was referred to the hematology/oncology department and a bone marrow biopsy was performed which was consistent with chronic myelomonocytic leukemia (World Health Organization type I).

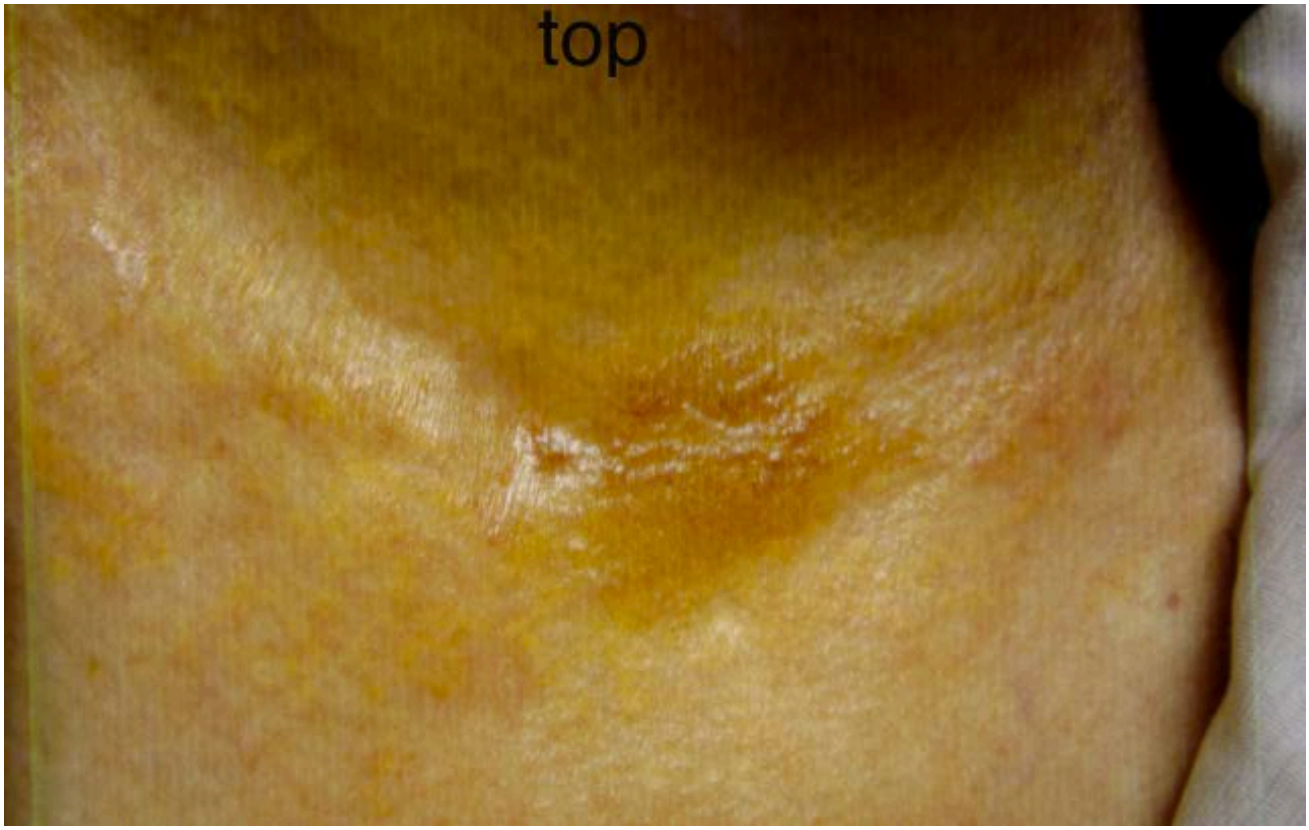

Figure 1. Yellow-orange plaques consistent with PX on patient's lower neck and chest

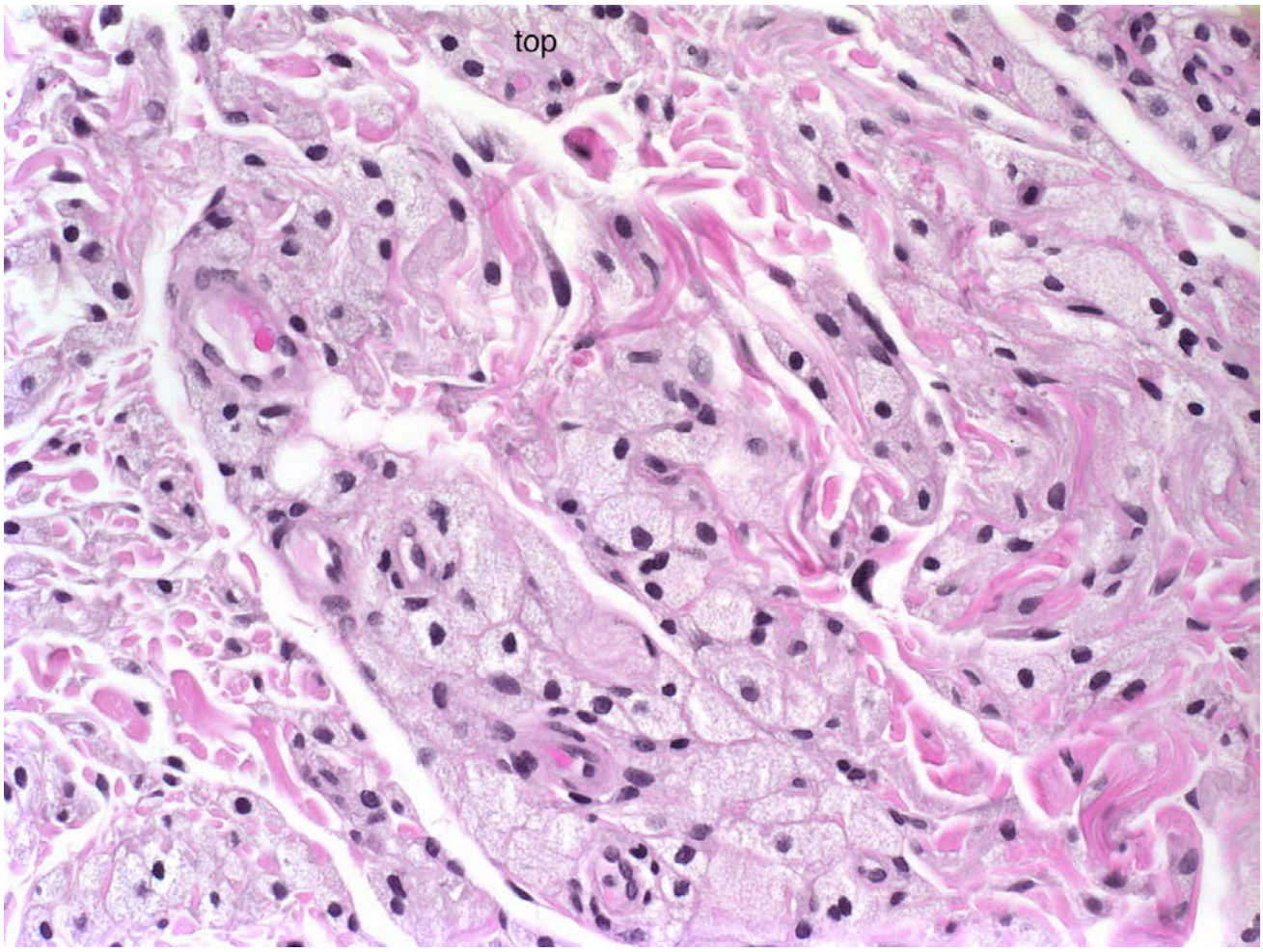

Figure 2. Hematoxylin and eosin stained biopsy results at 40x magnification showing nodular aggregates of large pale foamy staining histiocytes in the dermis consistent with xanthoma

Table 1. Laboratory findings

WBC	12,700		
	PMN		12%
	Myelocytes		2%
	Blasts		7%
	Basophils		889
	Lymphocytes		5,588
	Monocytes		3,683
Hemoglobin	9.4		
MCV	89.7		
Platelets	104,000		

Discussion

Diffuse planar xanthoma (DPX) is a documented, yet rare, manifestation of underlying monoclonal gammopathy, most commonly monoclonal gammopathy of undetermined significance or multiple myeloma [1]. In the literature, it is hypothesized that excess monoclonal antibodies bind with low density lipoproteins in the body and form complexes in the skin [2].

In some cases, DPX is the first sign or can even precede the onset of myeloproliferative disease. Therefore, presentation with DPX warrants a full hematologic investigation and continued clinical monitoring. In 1998, Marcoval et al reported 8 cases of DPX, 3 of which were associated with underlying reticuloendothelial disease [1].

Diffuse planar xanthoma is very rarely associated with chronic myelomonocytic leukemia (CML). The first description of DPX associated with CML was by Vail et al in 1985 [3]. Though it has been recognized in the literature, in 2011 Maxit & Paz described a woman with a 10 year history of severe DPX whose underlying CML was not diagnosed until autopsy [4]. Recognition of this association is extremely important for early detection of hematologic abnormalities.

In their study of 8 cases of DPX, Marcoval et al suggested that an associated hematologic abnormality is more likely in patients with extensive cutaneous involvement [1]. However, our patient only had localized involvement of her lower neck and upper chest. Therefore, we suggest that clinicians maintain a low threshold for hematologic testing in any patient presenting with localized or diffuse PX.

Historically, patients with DPX associated CML do not have abnormalities in lipid metabolism [1,3-4]. Our patient had normal lipids on laboratory evaluation, but she had a history of dyslipidemia and was being treated with a lipid-lowering agent at the time of presentation. Patients presenting with PX with historical abnormalities in their lipids should still be evaluated for underlying myeloproliferative disorders.

Conclusion

In patients presenting with PX, it is important for clinicians to evaluate for underlying myeloproliferative disease. This is true for all patients, including those with only localized cutaneous findings and those with a history of lipid abnormalities.

References

1. Marcoval J, Moreon A, Bordas X, Gallardo F, Peyri J. Diffuse plane xanthoma: clinicopathologic study of 8 cases. J Am Acad Dermatol. 1998;39(3):439-42. [PMID: 9738779]
2. Carlesimo, M, Rossi, A, La Pietra, M, Narcisi, A, Verga, E, Arcese, A, Cacchi, C, Camplone, G. Diffuse plane xanthoma and monoclonal gammopathies. Eur J Dermatol. 2009;19(6):640-1. [PMID: 19651560]
3. Vail, J, Adler, K, Rothenberg, J. Cutaneous xanthomas associated with chronic myelomonocytic leukemia. Arch Dermatol. 1985;121(10):1318-20. [PMID: 3862364]
4. Maxit, M, Paz, R. Diffuse plane xanthoma with arthritis, serositis, erythema nodosum, vasculities and myelomonocytic leukemia. Description of a case with autopsy. Medicina. 2001;61(2):187-90. [PMID: 11374143]