Cutaneous inflammatory pseudotumour: a rare and multimorphic condition

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Summary

Inflammatory pseudotumour (IPT) is a benign mesenchymal tumour common in many body sites, rarely involving the skin. We present two patients with lesions of diverse gross morphologic features, with dermatopathologic patterns compatible with IPT. The first case involved a persistent facial lesion constituting a therapeutic challenge. In the second case the condition was resolved by the excisional biopsy but was associated with a systemic disease and an autoimmune disorder.

Full case histories were taken, detailed histopathological studies, marker staining was performed and follow-up ensued. We also summarized all the previous cases reports of IPT involving cutaneous structures.

KEY WORDS: cutaneous inflammatory pseudotumour; systemic disease; subcutaneous nodule.

Introduction

Inflammatory pseudotumour (IPT) is a diagnosis rarely reported in the skin (1-12). We present 2 separate cases of patients exhibiting morphological dissimilar lesions of a nonspecific clinical nature.

Case synopsis

Case I
A 33-year-old woman had longstanding mild thrombocytopenia, mild splenomegaly and IgA deficiency. Computerized tomography of the chest disclosed enlarged neck and pelvic nodes and micronodular pulmonary findings. Consequently, a firm mass was found protruding from the abdominal surface. Excision was performed and the following histopathological findings were reported (Figure 1 a-c): a firm-elastic encapsulated nodule 1.5 cm in diameter. Microscopic examination demonstrated a lymph node, nearly completely replaced by proliferation of monomorphic spindle cells with admixture of lymphocytes and plasma cells. Spindle cells were immunohistochemically positive for smooth muscle actin and negative for desmin, pancytokeratin, S100, ALK-1, CD23, CD21. No mitoses or atypia was seen.

In parallel she was subsequently diagnosed with sarcoidosis with a satisfactory response to prednisone.

Case II
A 30-year-old female presented with a swelling of the left lower eyelid without a history of trauma (Figure 2). The cutaneous surface of the involved region was erythematous. There were no previous or concomitant complaints. A biopsy of the tissue of the right orbital region revealed the following features: numerous perivascular lymphocytic aggregates with some plasmacytoid features and scattered plasma cells were apparent. The aggregates infiltrate the muscle in multiple foci. Immunostaining CD20 and CD3 shows mixed B and T cell populations in a ration B:T= 1:2. CD68 and CD138 highlight numerous histiocytes. A number of plasma cells stain by IgG. IgG4 stains were negative. Ki67 proliferation antigen was positive in a few scattered cells. CD23, CD21, CD10, BCL1, BCL6 all were negative. IGH heavy and light chains were non-contributory. PCR and FISH studies ruled out B cell clonality. The polymorphism of the cell aggregates led to conclude that the lesion belongs to a subgroup of IPT. Steroid infiltration by local injections resulted in substantial shrinking of the lesion, though later on a relapse of the lesion occurred. The process currently continues to progress slowly. The team treating her has not established the precise approach to the unsightly and disturbing condition.

These cases contribute to a modest list of previous cases of IPT involving the skin presented in Table 1.

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Inflammatory pseudotumour (IPT) is an apparently benign condition which has been described in both sexes, at all ages usually involving practically all internal organs. The differential diagnosis of nodules presenting in the reticuloendothelial system is broad and includes: infectious and granulomatous diseases, autoimmune diseases, Castleman’s disease, Kaposi’s sarcoma, Kimura’s, lymphadenitis, malignant fibrous histiocytoma and lymphomas. These entities have been excluded histopathologically. Ruling out malignant diseases including malignant lymphoma and plasmacytoma is essential. Immunostaining, myelogram and histological findings including the clinical courses all make a diagnosis of lymphoma unlikely. The extensive use of the term IPT to describe tumours that differ in their biologic potential and wide morphologic spectrum leads to much controversy. The term IPT has been considered all-inclusive of tumours lesions with a prominent plasma cell population, designated plasma cell granulomas, primarily composed of spindled myofibroblasts also known as inflammatory myofibroblastic tumours, but also includes mycobacterial spindle cell proliferations. We see eye to eye with our predecessors expressing that the few IPTs reported in the skin display such a morphologic
Cutaneous inflammatory pseudotumour

Table 1 - Previous reports of cutaneous inflammatory pseudotumour.

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Number of cases</th>
<th>Age</th>
<th>Gender</th>
<th>Number of lesions each patient</th>
<th>Morphology and special characteristics</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hurt MA, Santa Cruz DJ (1)</td>
<td>1990</td>
<td>4</td>
<td>?</td>
<td>Male</td>
<td>Arm X 2, Calf X1, Posterior neck X 1</td>
<td>One was located in the superficial subcutis and three in the reticular dermis</td>
</tr>
<tr>
<td>Yang M (2)</td>
<td>1993</td>
<td>1</td>
<td>44</td>
<td>male</td>
<td>hand</td>
<td></td>
</tr>
<tr>
<td>Vadmal MS, et al. (3)</td>
<td>1999</td>
<td>1</td>
<td>47</td>
<td>Female</td>
<td>Elbow</td>
<td>Painless erythematous indurated nodule. Concurrent Wegener's granulomatosis.</td>
</tr>
<tr>
<td>Nakajima T, Sano S, Itami S, Yoshikawa K (4)</td>
<td>2001</td>
<td>1</td>
<td>25</td>
<td>Female</td>
<td>Thigh</td>
<td>Broad superficial pigmentation</td>
</tr>
<tr>
<td>El Shabrawi-Caelen L, Kerl K, Cerroni L, Soyer HP, Kerl H. (5)</td>
<td>2004</td>
<td>5</td>
<td>89</td>
<td>Male</td>
<td>Shoulder</td>
<td>Two distinct patterns; one type (n = 3) displayed dense, lymphoplasmacytoid infiltrates containing lymphoplasmacytoid cells and plasma cells with occasional germinal centers and hyalinized collagen bundles but was devoid of a myofibroblastic component</td>
</tr>
<tr>
<td>Yung et al. (6)</td>
<td>2005</td>
<td>1</td>
<td>33</td>
<td>Male</td>
<td>Shoulder</td>
<td>Large firm indurated reddish plaque</td>
</tr>
<tr>
<td>Saricaoglu H, Akin S, Adim SB, Karadogan SK (7)</td>
<td>2006</td>
<td>1</td>
<td>57</td>
<td>Male</td>
<td>Right cheek</td>
<td>Purple red superficial pigmentation</td>
</tr>
<tr>
<td>Frey J, Huerter C, Shehan J (8)</td>
<td>2006</td>
<td>1</td>
<td>70</td>
<td>Male</td>
<td>Thigh</td>
<td>Pigmentation and overlying scaling</td>
</tr>
<tr>
<td>Pagni F (9)</td>
<td>2007</td>
<td>1</td>
<td>63</td>
<td>Male</td>
<td>Arm</td>
<td>Subcutaneous with normal epidermis</td>
</tr>
<tr>
<td>Gonzalez-Vela MC et al. (10)</td>
<td>2007</td>
<td>1</td>
<td>10</td>
<td>Male</td>
<td>Arm</td>
<td>----</td>
</tr>
<tr>
<td>Chen YF et al. (11)</td>
<td>2009</td>
<td>1</td>
<td>50</td>
<td>Male</td>
<td>Left lower leg</td>
<td>Rubbery dark brown noduloplaque</td>
</tr>
<tr>
<td>Son SB et al. (12)</td>
<td>2010</td>
<td>1</td>
<td>26</td>
<td>Male</td>
<td>Back</td>
<td>Slight red-brown nodular lesion</td>
</tr>
<tr>
<td>Current report</td>
<td>2016</td>
<td>2</td>
<td>30</td>
<td>Female</td>
<td>Face</td>
<td>Cutaneous component infiltrative</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>33</td>
<td>Female</td>
<td>Abdomen</td>
<td>No superficial component, nodular morphology</td>
</tr>
</tbody>
</table>

Heterogeneity, that we are led to the speculation that these tumors probably constitute more than one single-disease entity (5). In effect, the final histopathologic diagnosis is based on the contribution of each of the diagnostic features, namely, general appearance, cell constitution and markers unfortunately without a specific pathognomonic picture.

All in all, reports of predominant cutaneous appearance of IPT are rare. The literature reveals very few, while from Israel actually no previous reports whatsoever. To date, we have located 19 reports of these lesions found in skin and soft tissues (1-12) and one occurrence in the oral mucosa. The pathogenesis leading to such lesions remains unclear. Hurt et al. suggested that these lesions are in fact findings of cutaneous lymphoid hyperplasia secondary to insect bites (3). In our cases no such history existed. In one report an association was found with a viral infection (10). No focal or systemic infection was disclosed in the time of lesion eruption here.

Past cases found in the literature are summarized in Table 1. In line with all the previously reported cases, as in our case I, the diagnostic procedure of excisional biopsy was curative. In this patient both gross and microscopic examination could not reveal involvement of the epidermal or dermal structures. Morphologically, in the majority of reports the lesion in fact does involve the epidermal component of the skin with resulting pigmentation and/or scaling and ulceration. Only in one report by Pagni has such a manifestation...
been observed in the skin (9). Our cases demonstrate novel clinical presentations of hitherto rarely recognized form. The specific association of the coinciding phenomena, on the one hand with a generalized granulomatous disease (sarcoidosis) and on the other hand a solitary lesion of IPT is yet to be clarified, since this has never been reported. In previous reports of cutaneous inflammatory pseudotumour, there were no reports of a systemic condition aside from one case there was a concurrent autoimmune condition of vasculitis (2). Visceral IPT has an established association with autoimmune conditions and we often observe regression of the masses together with gain of control over the underlying systemic condition.

Unique features of case I include the involvement of the abdominal wall region, association with systemic autoimmune condition and recognized immune system abnormality i.e. IgA deficiency, chronic thrombocytopenia, sarcoidosis responsive to steroids with regression of lymphadenopathy and no skin lesions. Unique features of case II include locally diffuse involvement of the orbital and periocular region with epidermal involvement, as well as extension to the orbital tissue itself. We observed intermittent spontaneous resolution with residual postinflammatory response and a fluctuating course with incomplete response to repeated therapeutic interventions. Therefore, in contrast, to the previous case, this case constitutes a predominantly clinical dilemma owing to its aesthetic repercussions. This is frustrating since the paucity of experience and knowledge with this condition disallows accepting a clear cut decision between an active invasive therapeutic intervention versus a safer more conservative observational approach. With accumulation of a broader database, in the future, we may hopefully better define the nature of this neoplasm and the optimal clinical approach.

Conclusion

The main purposes of this report is to disseminate knowledge of this uncommon finding and perhaps lead to accumulated sufficient data to shed light on the appearance and perhaps behavior or even etiological circumstances contributing to the transformation of this interesting though ambiguous clinical finding.

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Conflict of interest

None declared.

References