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Localized abdominal idiopathic lipodystrophy

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Abstract

Localized loss of subcutaneous tissue can occur after panniculitis, injections of corticosteroids and other drugs, or associated with infectious, autoimmune or neurologic diseases. The "idiopathic lipoatrophies" are a group of poorly characterized diseases, with focal disappearance of subcutaneous fat, and usually the thighs, abdomen or the ankles are affected. Three subtypes have been described based on clinical presentation: lipoatrophia semicircularis, annular lipoatrophy of the ankles and centrifugal lipodystrophy. We describe a 52-year-old female patient who developed a localized atrophy of the abdominal areas over a period of 3 months without any inflammatory signs over the evolution of the disease. The patient denied any previous local trauma or medication of any type. The atrophy stabilized, showing no progression over the last 6 years. The histopathological examination was normal except for the absence of subcutaneous fat, although the biopsy was taken down to the fascia. There was no clinical or serologic evidence of autoimmune diseases and laboratory testing for Borrelia burgdorferi infection was negative. Other causes of localized lipoatrophies were excluded and the final diagnosis was localized idiopathic lipodystrophy. Our patient is the second report on an abdominal lipodystrophy, with no previous inflammatory signs, absence of subcutaneous fat and no associated pathogenic factor. There is no established treatment for idiopathic lipodystrophy, and the lesions do not tend to resolve spontaneously.

Introduction

Localized loss of subcutaneous tissue is rare and may occur in a number of disease processes [1]. More frequently they represent

the resolution stage of a preceding panniculitis, usually associated with previous inflammatory signs [2]. Infectious, autoimmune and neurologic diseases may also be associated [2, 3, 4, 5] and a subgroup of patients will have no identifiable cause. These patients are grouped together as "localized idiopathic lipoatrophies," which represents a heterogeneous group of poorly characterized diseases [1, 6].

Case Report

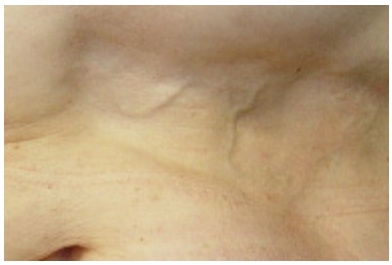

Figure 1

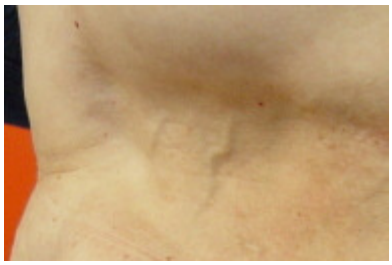

Figure 2

Figure 1. Unilateral localized abdominal lipodystrophy, without previous inflammatory signs
Figure 2. Localized lipodystrophy, with complete loss of subcutaneous fat on the affected area

We describe a 52-year-old female patient, which developed a localized atrophy of the abdominal areas over a period of 3 months (Fig. 1). The subcutaneous tissue of the upper left abdominal quadrant progressively disappeared, leaving the so-called "pseudo-athletic" aspect, with notable veins and muscular tissue (Fig. 2). There were no surface skin changes, and the area was never indurated. The patient denied any inflammatory signs over the evolution of the disease, and there were no previous local trauma or medication of any type.

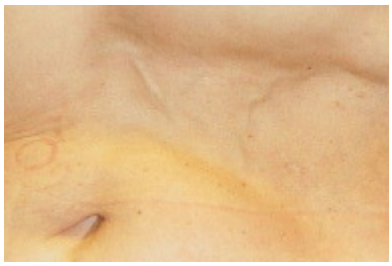

Figure 3

Figure 3. No changes in the affected area during the

evolution of the disease (pictures taken with an interval of two years)

The atrophy stabilized, showing no progression over the last 6 years (Fig. 3). The histopathologycal examination had no epidermic or dermal pathological changes, with absence of subcutaneous fat although the biopsy was taken down to the fascia. An abdominal ultrasound and magnetic resonance of the area were performed, revealing only the atrophy of the subcutaneous fat, without associated muscular or organ changes. There was no clinical or serologic evidence of autoimmune diseases and laboratory testing for *Borrelia burgdorferi* infection was negative. Other causes of localized lipodystrophy were excluded and the final diagnosis was localized idiopathic lipodystrophy. Fat transfer was suggested, but the patient refused any treatment.

Discussion

In localized idiopathic lipoatrophy there is focal disappearance of subcutaneous fat, usually affecting the thighs, abdomen, or the ankles [2, 6]. Three subtypes have been described based on clinical presentation, with lipoatrophia semicircularis presenting as a band-like semicircular depression on the anterolateral aspects of the thighs probably due to repetitive trauma [6, 7, 8]. The presence of circumferential atrophic lesions of the ankles has also been described as annular lipoatrophy of the ankles, frequently associated with auto-immune diseases [9, 10]. Centrifugal lipodystrophy was initially described in Japanese children, with usually unilateral centrifugally spreading lipoatrophy, showing a bluish centre and erythema of the edges [11, 12]. There is only one previous report on an adult female with a clinical evolution similar to our patient, and was named unilateral localized idiopathic lipoatrophy [13]. In both patients localized unilateral loss of subcutaneous abdominal fat occurred, with no pre-existing inflammation or trauma. The overlying skin was normal and the lesions remained stable. In a review of panniculitis, Requena et al. defends that the designation of lipoatrophy should be used when there is a preceding inflammatory process involving the subcutis, corresponding to the end-stage of many types of panniculitis [2]. In contrast, lipodystrophy should be used when there is an absence of previous inflammatory stage and is more commonly associated with endocrinologic, metabolic, or autoimmune diseases [2]. Nevertheless, in the literature the 2 terms are often used interchangeably, adding further confusion to the nomenclature. As in both cases no previous clinical inflammation was described and histology could not identify any remaining subcutaneous tissue, the correct designation probably should be unilateral abdominal idiopathic lipodystrophy.

In the differential diagnosis of localized atrophies of the subcutaneous fat it must also be considered borreliosis and local injections of insulin, corticosteroids or other medication [3, 14, 15, 16, 17]. Panniculitis should also be excluded, where the presence of a preceding inflammatory stage is important [2]. The lack of epidermal or dermal changes differentiates localized idiophatic lipodystrophy from local panatrophy, which is a rare disorder with partial or total loss of subcutaneous fat, with associated atrophy of the overlying skin [18]. In panatrophy of Gowers, one of the two subtypes, single or multiple sharply defined areas of atrophy develop over a period of few weeks and in the affected area the subcutaneous tissue disappears [19]. There is no previous inflammatory process or sclerotic skin changes, and occurs mostly in young women [20]. Sclerotic panatrophy is another subtype, with sclerotic linear bands and occasional involvement of muscle or bone, occuring more frequently in children [21]. In both idiopathic lipoatrophy and panatrophy, after a rapid evolution, the lesions tend to remain unchanged indefinitely [13, 19]. Partial lipodystrophy is a more complex systemic disease, where there is disappearance of the fat from the face and upper half of the body, usually associated with progressive mesangiocapillary glomerulonephritis and hypocomplementemia [22]. There are rare cases of unilateral involvement, but the differential diagnosis is usually simple [22,23].

Two histopathological patterns have been described in localized lipoatrophies, with the involutional subtype being the most common, showing a pathological pattern with similarities to embryonic fat globules [24, 25]. In the inflammatory subtype there are normal lypocytes, mixed inflammatory infiltrate and the presence of immunoreactants [26]. The biopsy in our patient was performed after the disease process had stabilized, and no subcutaneous fat was seen. The same happened in the other published case of unilateral localized idiopathic lipoatrophy and probably a biopsy taken in the early stages might have provided more information [13].

Although the pathogenesis is not known, there are associations of lipodystrophy to immunologically related diseases such as systemic lupus erythematosus, systemic sclerosis, autoimmune thyroid and renal disease [6, 23]. Repetitive trauma is considered the pathological factor involved in lipoatrophia semicircularis [7, 8]. It has also been suggested that there may be a primary or secondary abnormality of the sympathetic nervous system associated with localized disorders of the subcutaneous fat [26]. Lipodystrophy associated with antiretroviral treatment for the human immunodeficiency virus infection, has been related to changes of autonomic balance, resulting in redistribution of adipose tissue [27, 28]. The facial hemiatrophy of Parry-Romberg syndrome also seems to be related to a disorder of the sympathetic

nervous system [29].

There is no established treatment for idiopathic lipodystrophy and the lesions do not tend to resolve spontaneously [6, 13]. Autologous fat transfer to the atrophic site and the use of temporary or definitive fillers are therapeutical options [30, 31, 32, 33].

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