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Letter

Multiple cutaneous lipomatous neurofibromas

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Abstract

Cutaneous lipomatous neurofibroma is a rare variant of neurofibroma with a little known pathogenesis; its presence has never been described in Brazil. A 61-year-old woman complaining scalp papules for one year, presented with skin colored to yellowish papules on the scalp. She noted that they were sometimes itchy and occasionally bled.

Dermoscopy showed amorphous material, milky white, with a small poorly delimited yellowish area. Histopathological examination revealed spindle cell proliferation associated with mature adipocytes. Cutaneous lipomatous neurofibroma appears to be an underdiagnosed condition owing to the low index of suspicion and lack of knowledge of its existence. Few cases have been reported in the literature.

Introduction

Lipomatous neurofibroma is a rare histopathological variant of neurofibroma [1-4]. It is most often located in the head and neck region of elderly patients and the papules and nodules have generally been longstanding [2,3]. Its dermatoscopic analysis shows a yellowish amorphous material, which is characteristic and helps to differentiate the tumor from a common dermatofibroma [2]. Cutaneous lipomatous neurofibroma was first described in 2002 by ValBernal et al. and since then, few cases have been reported in the literature, none previously in Brazil.

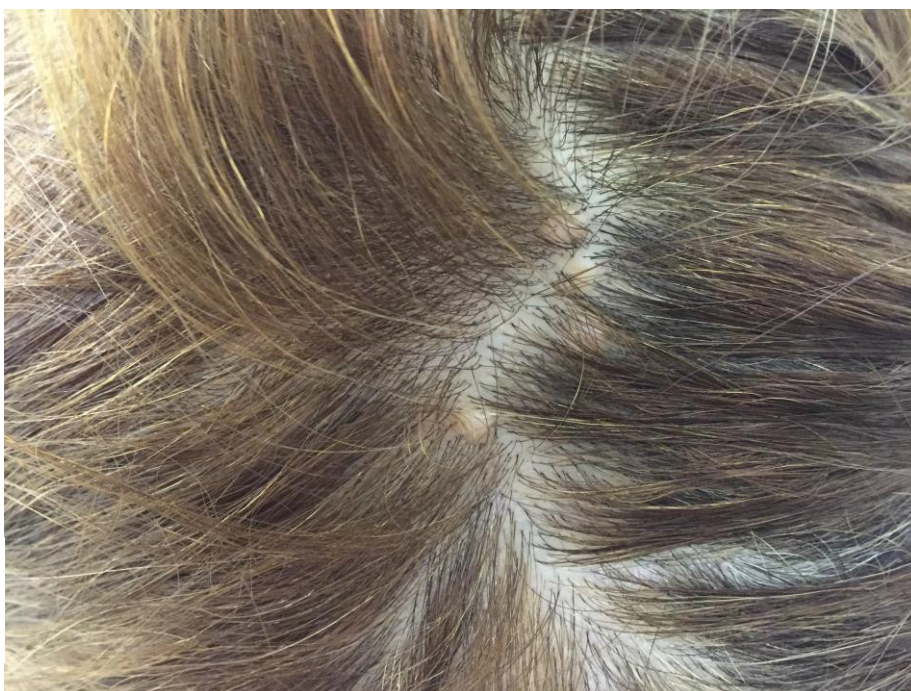


Figure 1. Multiple skin colored to yellowish papules in parietal scalp topography.

Case synopsis

A 61-year-old woman presented with several scalp lesions, sometimes itchy that occasionally bled, which emerged a year prior to presentation. On dermatological examination multiple skin colored to yellowish papules of the parietal scalp topography were observed (Figure1). Dermoscopy showed amorphous material, milky white with a small poorly delimited yellowish area (Figure2). Histopathological examination revealed spindle cell proliferation with mononuclear cells in between, associated with mature adipocytes under low power magnification (Figure3). We opted for a conservative observational approach owing to the large number and benign nature of the lesions.



Figure 2. Dermoscopy showed amorphous material, milky white with a small poorly delimited yellowish area (original magnification x 20).

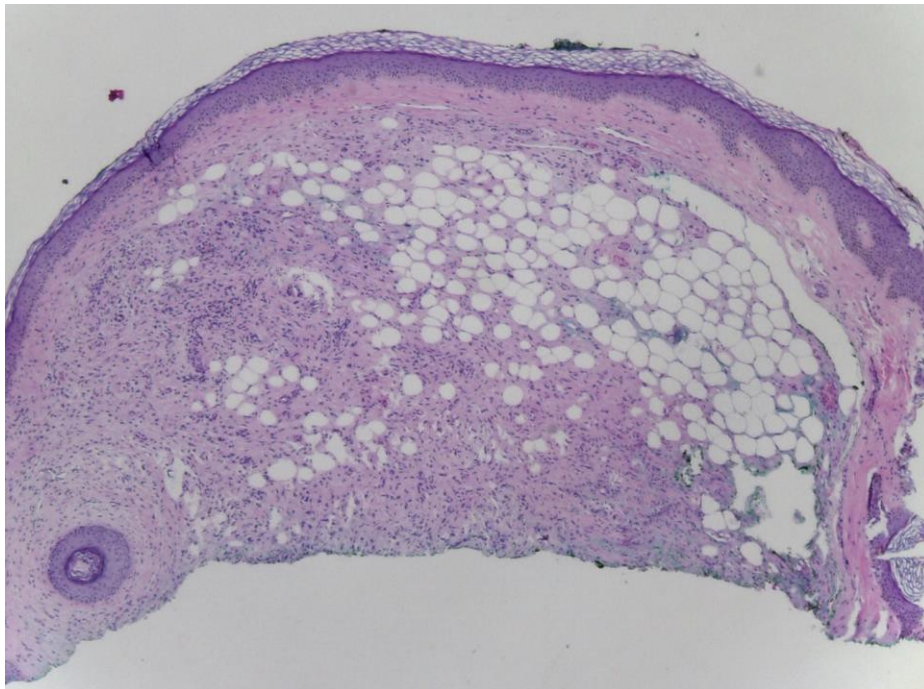


Figure 3. Spindle cells proliferation with mononuclear cells in between, associated with mature adipocytes (haematoxylin and eosin, original magnification $\times 40$).

The origin of the fat presence in this type of neurofibroma is unknown and some hypotheses have been suggested to explain its pathogenesis. Val Bernal et al initially proposed that the fat was an integral part of the tumor, not being derived by metaplasia or degeneration. They postulated that fat cells would be intrinsic to the tumor, arising from local stem cells [1]. In contrast, Ahn et al. asserted that the presence of fat would be the result of senescent change or chronic injury in older patients and in longstanding lesions. Repetitive injury, such as shaving, combing, or exposure to ultraviolet radiation could account for these focal changes [2-4]. However, in 2005, ValBernal et al conducted a systematic study based on 320 specimens with a diagnosis of cutaneous neurofibroma, using conventional microscopy, immunohistochemistry, and statistical methods to determine the presence of fat cells, their quantity, distribution, and frequency. This study demonstrated a relatively high

incidence of mature adipocytes in cutaneous neurofibroma: intratumoral fat was observed in 22 (6.9%) of neurofibromas. All these were dermal neurofibromas. Intraneoplastic fat was divided into two groups: focal and diffuse (regularly interspersed). Eighteen tumors (5.6%) presented with adipocytes focally intermingled with the spindle cells. There were four (1.3%) neurofibromas showing spindle cell proliferation with regularly scattered adipocytes. The conclusion was that there are two patterns of fat distribution within these neurofibromas: the presence of localized fat infiltration, which would be a consequence of tumor metaplasia, and the diffuse presence of fat (a less frequent subtype), that characterizes an intrinsic tumor characteristic [2-4]. Although little reported in the literature, the lipomatous neurofibromas appear to be an underdiagnosed condition owing to the low index of suspicion and the paucity of knowledge of its existence, since few cases have been reported in the literature to date.

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