Atypical presentation of Schwannoma mimicking squamous cell carcinoma☆

Dear Editor,

Schwannomas are rare, encapsulated, benign tumors originating from the nerve sheath. Although they often occur as solitary lesions in 90% of cases, they may arise in association with central nervous system tumors in 5% of cases. They may also be a manifestation of type 2 neurofibromatosis (3%) or appear as multiple lesions (schwanomatosis).[1,2] Schwannomas can occur anywhere in the body along the course of a cranial, spinal or peripheral nerve.[3]

Cutaneous schwannomas (CS) appear as deep dermal or subcutaneous nodular lesions. More rarely, they may be located in the superficial dermis. Clinically, they are characterized as well-circumscribed, skin-colored, firm nodules which are generally asymptomatic. However, when pain or tenderness is present, it is usually associated with compression of the adjacent structures, so paresthesia is confined to the tumor site or radiating along the nerve of origin. In fact, pain, tenderness, or paresthesia may accompany up to one-third of the cutaneous manifestations.[4] CS most often occurs in the 4th and 5th decades of life, without significant evidence of gender predilection.[5]

A 50-year-old man was evaluated due to a painless lesion on the anterior aspect of the right leg evolving for 2 years, with accentuated growth in the 2 months previous to observation, with ulceration. He denied other symptoms such as pain or paraesthesia or a history of cardiac, pulmonary, or neurological pathology.

An ulcerated nodular lesion with 3 cm in diameter was observed in that location (Fig. 1). The diagnostic hypothesis of squamous cell carcinoma or keratoacanthoma was raised. Excision of the lesion was performed.

Microscopic examination showed a well-defined tumoral lesion covered by a sclero-hyaline capsule, with ulceration and necrosis of the overlying epidermis and dermis (Fig. 2A and B). The lesion consisted of two patterns: more compacted areas composed of ovoid to spindle-shaped cells with eosinophilic cytoplasm and indistinct cell boundaries (Antoni A pattern) with occasional nuclear palisading (Verocay bodies), and others areas more loosed and hypocellular consisting of cells with clear cytoplasm and well-defined

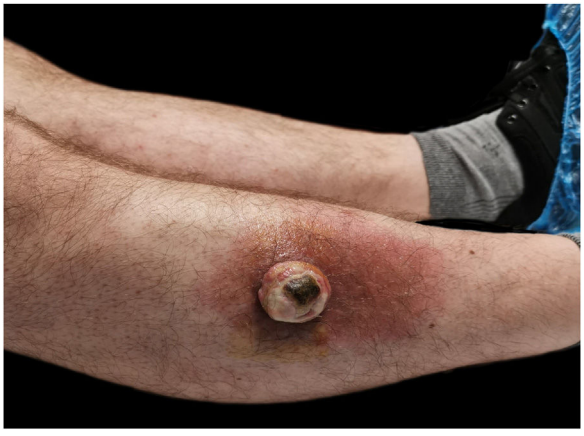

Fig. 1 A nodular ulcerated lesion on the lateral aspect of the right leg.

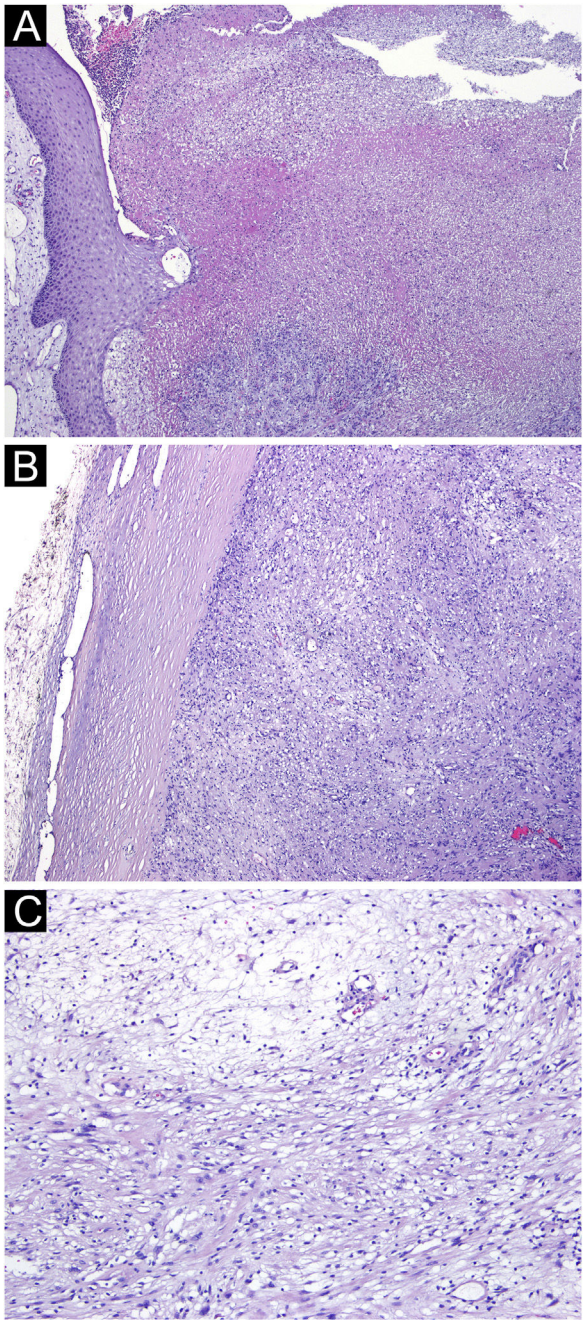

Fig. 2 (A) Ulceration and necrosis with suppuration of the adjacent epidermis and dermis (Hematoxylin & eosin, ×40). (B) The lesion was well-delimited by a sclero-hyaline capsule (Hematoxylin & eosin, ×40). (C) Tumoral lesion consisting of ovoid to spindle-shaped cells, with areas of eosinophilic cytoplasm and indistinct boundaries (Antoni A pattern) alternating with more loose and hypocellular areas of cells with clear cytoplasm and well-defined boundaries (Antoni B pattern) (Hematoxylin & eosin, ×100).

☆ Study conducted at the ULS São João, Porto, Portugal.

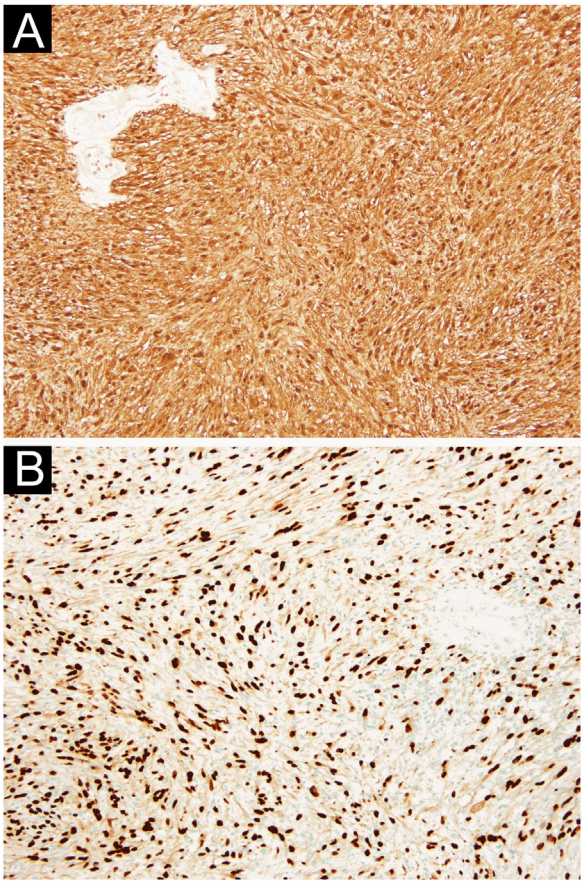

Fig. 3 (A) Diffuse expression of S100 protein (×100). (B) Diffuse expression of SOX-10 (×100).

boundaries, with collagenous stroma with myxoid areas and hyalinized wall vessels (Fig. 2C).

In the immunohistochemical study, diffuse expression of protein S100 and SOX-10 was observed, in the absence of HBM45, Melan-A, EMA, among others (Fig. 3A and B). These aspects were suggestive of schwannoma.

CS is the most common benign peripheral nerve sheath tumor, although occurrence on the lower limbs represents only about 1% of all cases.[6]

Histologically, CS is characterized by two types of histological patterns typically encapsulated by perineurium: Antoni type A and Antoni type B. Antoni A is a highly ordered cellular pattern in which spindle cells are arranged in compact fascicles and their nuclei are disposed of in palisades. Verocay bodies are a characteristic feature in this type of pattern, with collagen matrix arranged into palisading. Antoni type B tissue exhibits a looser structure of mucinous matrix and is less cellular.[5]

The differential diagnosis of CS includes proliferating pilomatricoma, lipoma, desmoid tumor, and epithelial cysts, among others. If tumors of the skin are tender or painful, nine tumors should be considered: leiomyoma, eccrine spiradenoma, neuroma, dermatofibroma, angiolipoma, neurilemmoma (schwannoma), endometrioma, glomus tumor, and granular cell tumor (LEND AN EGG - acronym).[7]

The histological differential diagnosis includes palisaded and encapsulated neuroma (PEN) and neurofibroma.[8]

It is important to appropriately distinguish superficially located schwannoma from PEN, because PEN is encapsulated, located in the upper dermis, and the patterns of interlacing fascicles can be similar to the Antoni type A pattern of schwannoma. Even tough axon-rich PEN does not show typical patterns of Antoni A and B of schwannomas, differentiating between schwannoma and PEN with low or absent axon densities can be troublesome. Neurofibromas are circumscribed but not encapsulated and are composed of spindle cells loosely spaced and wavy collagen strands.[9]

The best treatment option for CS is local excision.[10]

This case corresponds to an atypical presentation considering the rapid growth and ulceration, simulating a malignant lesion, as well as the leg location, which is infrequently described in the literature for schwannoma.

Authors' contributions

Pedro Rolo de Matos: Primary author, research, writing.
Miguel Costa Silva: Design, review.
Gilberto Pires Rosa: Support in writing of manuscript.
Pedro Amorso Canão: Pathological analysis and report.
Filomena Moreira Azevedo: Final review.

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Conflicts of interest

None declared.

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Facial pemphigus vegetans mimicking squamous cell carcinoma: when dermoscopy confuses the diagnosis☆

Dear Editor,

Pemphigus vegetans (PVeg) is a rare variant of pemphigus vulgaris (PV) (1%–2% of cases) characterized by flaccid bullae or pustules that erode to form hypertrophic plaques and vegetating masses.[1] Generally, the lesions are multifocal and localized on flexures, periorificial areas, and oral mucosa.[1,2] It has two clinical forms that have been described in the literature: the Neumann type and the Hallopeau type.[2] The diagnosis is made based on clinical features, but the biopsy is mandatory to confirm it. There are no dermatoscopic reports of PVeg, but the presence of pustules, micro-vesicles, and erosions can guide us in its initial stage. Histological examination shows acantholysis, epidermal hyperplasia, papillomatosis, and intraepidermal eosinophilic and neutrophilic abscesses. Direct Immunofluorescence (DIF) demonstrates intercellular deposition of IgG and C3.[1,3]

Herein, we report a challenging case presenting as a solitary facial hyperkeratotic plaque of PVeg without oral mucosal involvement initially misdiagnosed as a squamous cell carcinoma (SCC) by clinical, dermoscopy, and histological examination.

We present, a sixty-three-year-old male who was referred to our dermatology department for an asymptomatic recurrent lesion of the right frontal area which had been present for two years. Physical examination showed a 2×2.5 cm well-demarcated hyperkeratotic solitary plaque with an eroded surface (Fig. 1A). No intertriginous or oral mucosa involvement was seen. Dermoscopy revealed a predominantly white background with surface scale, multiple different-sized keratin-filled follicular ostia, and white perifollicular circles surrounded by erythema (Fig. 1B). He had an incisional 4 mm punch biopsy, performed in an exter-

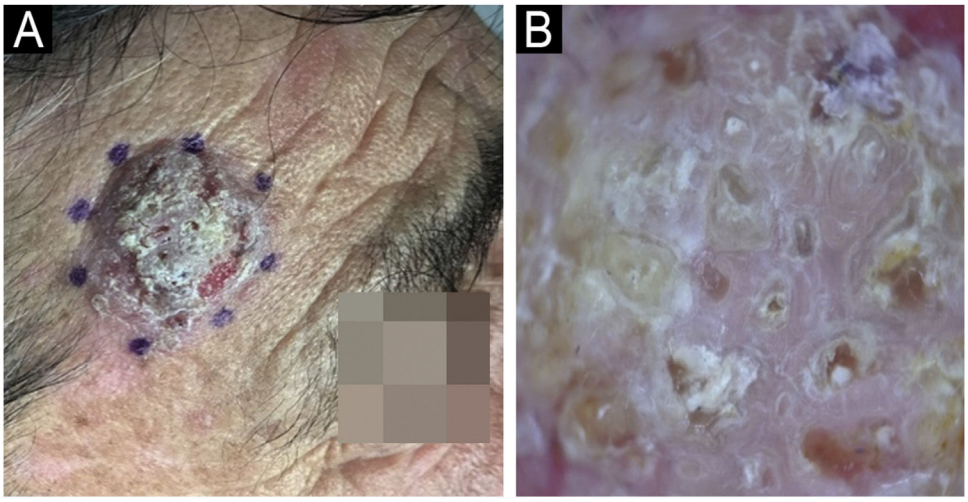

Fig. 1 (A) Well-demarcated hyperkeratotic plaque with an eroded surface. (B) Dermoscopy reveals a white background with surface scale, multiple keratin-filled follicular ostia, white perifollicular circles surrounded by erythema, and white structureless areas. Some red areas attributable to bleeding and/or dense vascularity are seen at the periphery.

☆ Study conducted at the Clínica Alemana, Santiago, Chile.