

Solitary Circumscribed Neuroma

Dermoscopic clues to facilitate diagnosis

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Figure 1: Photograph of a pink, translucent, firm, dome-shaped papule on the left side of the chin of a 66-year-old woman.

A 66-YEAR-OLD WOMAN PRESENTED TO THE Dermatology Outpatient Clinic of the Hospital Universitario Reina Sofía, Córdoba, Spain in 2017 with a 10-year history of a painful slow-growing erythematous lesion on her chin. A clinical examination revealed a pink, translucent, firm, non-ulcerated papule of 6 mm [Figure 1]. No similar cutaneous or mucosal lesions were noted anywhere else on the body. Polarised non-contact dermoscopy of the papule (Dermlite DL3N[®], 3Gen Inc., San Juan Capistrano, California, USA) indicated arborising vessels on a pinkish-reddish background, a central white spot and peripheral pigmentation [Figure 2].

Following complete surgical excision of the lesion, a histopathological examination revealed a well-circumscribed partially-encapsulated dermal proliferation comprised of spindle cells with scanty pale cytoplasm and elongated wavy nuclei. The nuclei were grouped in distinct fascicles set in fibrillar, collagenous and occasionally myxoid stroma [Figure 3]. The patient was subsequently diagnosed with a solitary circum-

scribed neuroma (SCN). She recovered successfully following the surgery, with no recurrence observed after 10 months of follow-up.

Comment

Also known as palisaded encapsulated neuromas, SCNs usually present in adulthood as asymptomatic small nodules of between 2–6 mm, mainly located on the face. However, cases have been described in other locations, including the shoulder, hands, arms, feet and mucosal areas.^{1,2} Mucosal neuromas have been linked with multiple endocrine neoplasia 2b syndrome and phosphatase and tensin homolog deleted on chromosome 10 hamartoma tumour syndromes, including Cowden syndrome, Proteus syndrome and Bannayan-Riley-Ruvalcaba syndrome.^{3,4} Histologically, the examination of an SCN reveals a well-circumscribed partially-encapsulated intradermal nodule comprised of spindle cells grouped in distinct fascicles with a lack of nuclear

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Figure 2: Dermoscopic image showing a symmetrical non-pigmented lesion with an arborising vascular pattern and surrounded by a peripheral dark-yellowish halo.

pleomorphism and *mitosis*.⁴ Immunohistochemical studies of SCN cases have demonstrated positive cell staining for S100 proteins, collagen type IV and vimentin.²⁻⁴ In cases where the diagnosis is not clear, glial fibrillary acid protein and epithelial membrane antigen staining may also be necessary.³

From a physiopathological point of view, it has been suggested that SCNs are caused by the inexplicable hamartomatous growth of Schwann cells over axons.⁴ The clinical and dermoscopic differential diagnosis should include basal cell carcinomas (BCCs), adnexal tumours, *neurothekeomas*, vascular tumours and intradermal melanocytic *naevi*.⁴ In addition, SCNs may have similar histological findings to those of neurofibromas and schwannomas; however, while neurofibromas lack a capsule and contain a mucopolysaccharide ground substance, schwannomas are generally subcutaneous and contain Antoni types A and B tissue patterns and Verocay bodies with no axons present.³ Immunostaining with human melanoma black-45, melan-A, tyrosinase and cluster of differentiation (CD)34 can be useful to differentiate these entities.

Dermoscopy is a useful noninvasive tool that helps to identify tumours that mimic BCCs.⁴ However, the absence of BCC features—including blue-grey

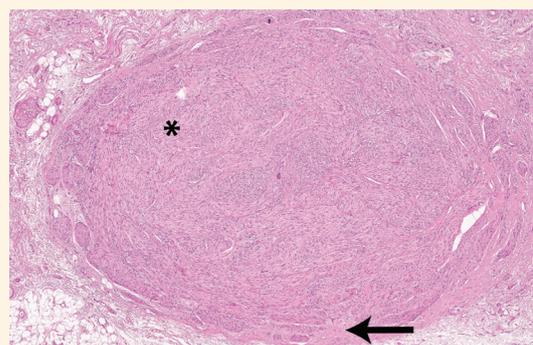


Figure 3: Haematoxylin and eosin stain at x10 magnification showing a well-circumscribed dermal nodular tumour comprised of small spindle cells (asterisk) without *atypia* or mitotic evidence of malignancy. The cells are grouped in small fascicles separated by grooves embedded in an eosinophilic fibrillar matrix (arrow).

ovoid nests, globules, dots or blotches and brown-grey leaf-like small erosions or ‘wheel-spoke’ areas—is not sufficient to warrant avoiding excision of the lesion. Clinicians should be aware of the characteristic arborising vascular patterns of SCN which appear dermoscopically as white patches on a pink-white background.^{4,5}

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