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## Multiple Cutaneous Granular Cell Tumors

### Tumores cutâneos múltiplos de células granulares

To the Editor:

Granular cell tumor (GCT), described by Abrikossov in 1926, is a rare and usually benign neoplasm considered to be of neural origin.<sup>1</sup> It usually presents as a solitary papule or nodule. Multiple cutaneous lesions such as those found in the case we present<sup>1,2</sup> are very rarely reported in the literature.<sup>3</sup>

Our patient was a 41-year-old white man with no relevant past history who, for 3 to 7 years, had had 4 slow-growing subcutaneous tumors of between 1.5 and 4 cm in diameter located on the left hip, the left iliac fossa, the right thigh (Figure 1), and the right scapular region. They were painful on palpation and not adherent to deep tissues. Magnetic resonance imaging showed that the lesions were located in the subcutaneous tissue and were independent of the fascia and underlying muscle (Figure 2). The skin biopsy showed a diffuse proliferation of polygonal cells with abundant granular eosinophilic cytoplasm, no necrosis or mitoses, and positive staining for S-100, vimentin, and CD68 (Figure 3), allowing us to confirm the diagnosis of granular cell tumor for all the lesions. The plastic surgery department performed a wide excision of the lesions. After 19 months of follow-up, there were no signs of recurrence, metastatic disease, or new lesions.

GCT is a rare tumor occurring in 0.017% to 0.025% of surgical specimens.<sup>2</sup> In patient series reported in English-language journals it most often affects black adult women (30-60 years), but in a Spanish series men aged 10 to 50 were predominant.<sup>3</sup> Its most common location is the tongue (40% of cases), followed by the skin and the subcutaneous tissue of the chest and limbs, although it has been reported in many organs.<sup>1,2,4</sup> Congenital<sup>5</sup> and familial<sup>6</sup> cases have also been reported. In 4.5% to 13% of cases it has been reported in association with other types of neoplasm.<sup>1,6,7</sup>

Clinically, it is a firm, solitary, circumscribed, asymptomatic nodule that is usually less than 3 cm in diameter; it may be pruritic or painful. The diagnosis is not usually suspected clinically<sup>1-4</sup> but depends on histology.

Histology shows it to be an ill-defined, nonencapsulated tumor composed of sheets, nests or cords of rounded or polygonal cells with a small central nucleus and abundant eosinophilic cytoplasm packed with coarse, diastase-resistant, periodic acid-Schiff-positive granules that represent phagolysosomes.<sup>1,3</sup> The overlying epidermis may be normal or show pseudoepitheliomatous hyperplasia.<sup>1-4</sup>

Immunohistochemistry reveals that a high percentage of granular cells are positive for S-100 (98-100%), neuron-specific enolase (98-100%), and vimentin (100%) and a lower percentage for CD57 (69%) and CD68 (65%).<sup>8</sup>

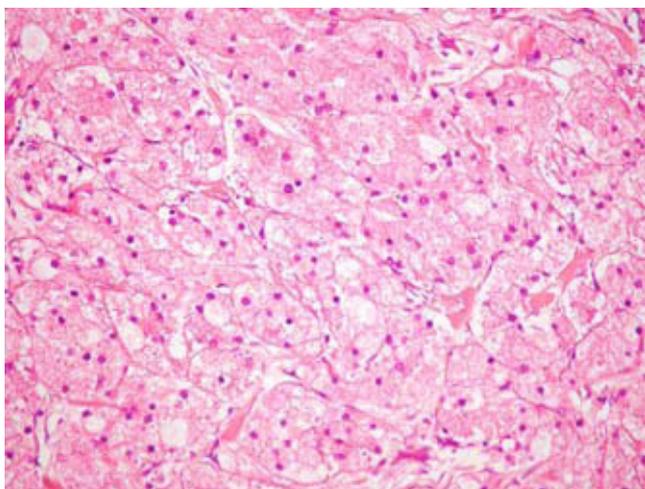
The treatment of choice is simple excision of the entire lesion with adequate surgical margins. Radiotherapy and chemotherapy are not recommended.<sup>1-4</sup>



**Figure 1** Detail of the subcutaneous lesion located on the right thigh.



**Figure 2** Magnetic resonance imaging shows a lesion in the subcutaneous tissue that does not infiltrate deep structures.



**Figure 3** Tumor composed of polygonal cells with abundant granular, eosinophilic, diastase-resistant, periodic acid-Schiff-positive cytoplasm. Hematoxylin-eosin (original magnification  $\times 40$ ).

The prognosis of GCTs is excellent. The recurrence rate is 8%<sup>1,6</sup> and is usually associated with inadequate surgical margins, though recurrence is not found in all cases with affected margins. Malignant cases (1-2%) behave like high-grade sarcomas<sup>8</sup> and the diagnosis of malignancy is based on the clinical behavior of the tumor, although attempts have been made to relate it to histologic features such as necrosis, mitotic index, pleomorphism, atypia, and ulceration.<sup>8</sup> Adverse clinical prognostic factors are local recurrence, the presence of metastases, old age, and size greater than 4 cm.<sup>8</sup>

Multiple cases like ours account for 0% to 30% of patients depending on the series.<sup>1-6</sup> This surprisingly large range is related to the difference in the follow-up period, although it is probably also related to racial differences, because multiple lesions are more common in black patients.<sup>2,6</sup>

In almost 50% of children with multiple lesions, there are associated systemic disturbances: pigmentary, cardiovascular, musculoskeletal, and central nervous system disorders, and neurofibromatosis.<sup>9</sup>

However, multiple GCTs in adults, such as those found in the case we present, have not been consistently associated with visceral involvement, other abnormalities, or malignancy. We would like to highlight 2 points:

*Multivisceral involvement:* Cases of GCTs affecting several organs simultaneously have been reported. More specifically there have been reports of multiple cutaneous GCTs associated with gastric GCTs<sup>10</sup> and pulmonary GCTs.<sup>6</sup>

*Malignant change:* Khawsur et al<sup>6</sup> describe 2 cases of malignant lesions that were found in the context of multiple benign skin lesions that had been appearing for years in a black father and son.

In conclusion, we report a case of multiple GCTs that is exceptional in Spain. Though these tumors are benign, on the basis of the literature reviewed we recommend a systems review, a complete physical examination, imaging tests aimed at detecting associated visceral lesions, and regular monitoring in such cases.

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## Smooth Muscle Hamartoma Associated with Acquired Blaschkoid Nevus Spilus

### Hamartoma del músculo liso asociado a nevo *spilus* blaschkoide adquirido

*To the Editor:*

Smooth muscle hamartoma (SMH), described by Stokes in 1923, is a proliferation of smooth muscle bundles. It can be single or multiple and may be congenital and acquired.<sup>1</sup> The most frequent presentation is the localized congenital form, which is characterized by a plaque with a variable degree of hyperpigmentation, hypertrichosis, and induration. It is usually less than 10 cm in diameter and appears on the trunk or proximal areas of the limbs. Rubbing the affected area can lead to pseudo-Darier sign, which consists of elevation, induration, or transient piloerection.<sup>2-4</sup> We report the case of a patient who recently attended our hospital with localized SMH within an acquired blaschkoid nevus spilus.

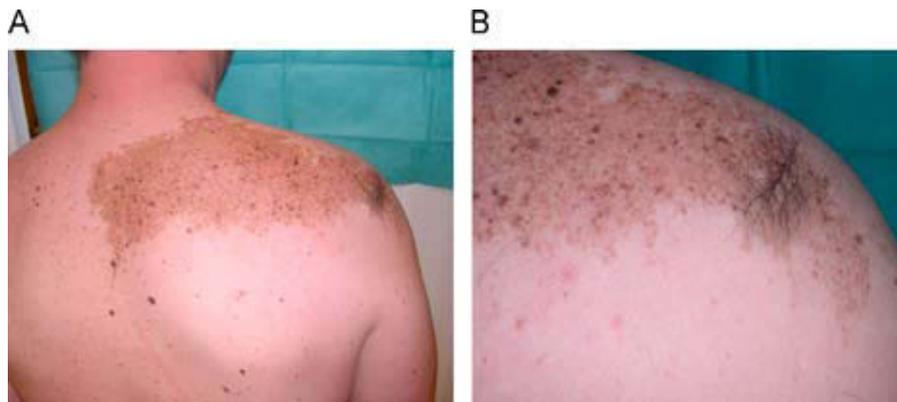
The patient was a 32-year-old man with no past history of interest who was seen for an asymptomatic pigmented lesion that appeared on the right shoulder when he was 6 years old. The patient and his mother described the gradual onset of darker spots within the lesion and, aged 13, the appearance of a hairy and slightly pruritic area.

Physical examination showed a homogenous, light brown, macular lesion with well-circumscribed irregular margins. It

had a unilateral, segmental distribution that followed the lines of Blaschko in a wide band across the upper area of the back and right shoulder and was sharply interrupted at the midline. Multiple blackish or dark brown spots were scattered over the lesion, most of which were elevated and less than 1 cm in diameter (Figure 1A). Near to the right external border there was a light brown plaque, 2 cm in diameter, firmer to the touch, and with long thick dark hair (Figure 1B); rubbing the lesion elicited transient piloerection.

Histopathology of the biopsy taken from the hairy area showed lentiginous melanocytic hyperplasia in the epidermis and disorganized, irregularly shaped smooth muscle fascicles in the dermis that were not associated with the pilosebaceous unit. These fascicles were surrounded by a clear space that separated them from the surrounding collagen (Figure 2). Immunohistochemical staining for muscle-specific actin highlighted the irregular distribution and organization of the smooth muscle fascicles in the dermis (Figure 3). A diagnosis was made of blaschkoid nevus spilus associated with SMH.

Smooth muscle hamartoma is sometimes associated with other skin conditions. Becker nevus is an abnormality that usually appears at the onset of puberty, presenting as a hyperpigmented area that can develop hypertrichosis. Histopathology shows a degree of acanthosis, elongated rete ridges, and hyperpigmentation of the basal layer. It is not unusual to observe smooth muscle hyperplasia in the dermis. For this reason, some authors suggest that SMH and Becker nevus represent opposite ends of a spectrum



**Figure 1** A, Upper back and right shoulder with a homogeneous clear brown macular lesion containing multiple blackish lesions and a hypertrichotic area. B, The area near the right external edge of the nevus spilus shown in greater detail. Light brown plaque, 2×2 cm diameter, containing thick, long, dark hair.