Case for diagnosis*

Caso para diagnóstico

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DOI: http://dx.doi.org/10.1590/abd1806-4841.20132772

CASE REPORT

Twenty-seven year old female patient, phototype V, presented a lesion in the right arm for one and a half years, without associated symptoms. Dermatological examination revealed a smooth-surfaced, brownish, painless nodule, movable within deep layers, measuring three centimeters and located in the right arm (Figure 1). Pathological examination of the surgical specimen showed dense proliferation of large cells with granular-appearing eosinophilic cytoplasm, distinct membranes, central or eccentric nuclei and slightly conspicuous nucleoli. The neoplastic cells formed syncytia in some areas and were inserted within the interstices of a desmoplastic stroma, with scarce focal lymphocytic infiltration. The results of immunohistochemical examination were positive for S100 protein, CD63 and CD68 (Figure 2).

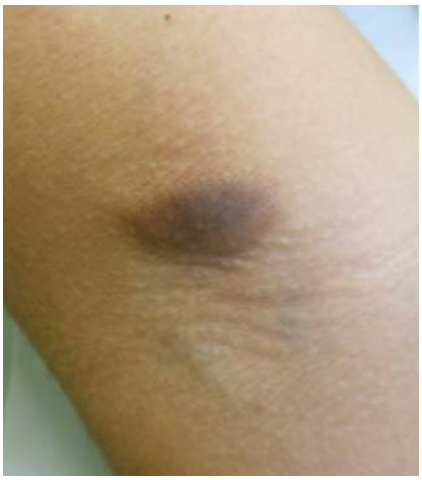

FIGURE 1: Smooth-surfaced hyperchromic nodule, movable in the deep layers, located on the right cubital fossa

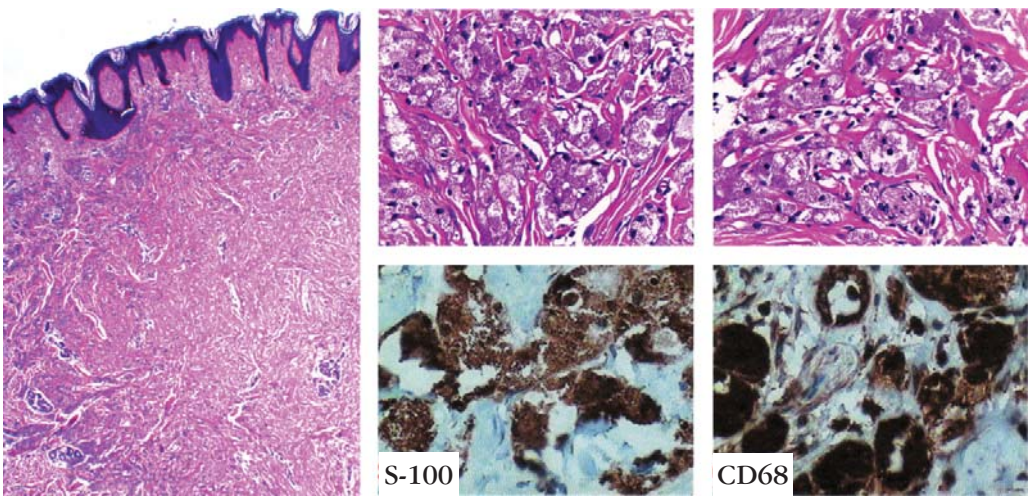

FIGURE 2: Hematoxiline-eosin stained fragment, histopathological exam showing dense proliferation of cells with a wide eosinophilic cytoplasm, central nuclei, and slightly conspicuous nucleoli. Immunohistochemistry positive for protein S100 e CD68

Received on 12.05.2013.
Approved by the Advisory Board and accepted for publication on 13.06.2013.
* Work performed at the Dermatology Service at Pará Federal University (UFPA) (UFPA) – Belém (PA), Brazil.
Conflict of interest: None
Financial funding: None

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DISCUSSION

Granular cell tumor (GCT), also known as Abrikossoff tumor, is a relatively rare benign condition that originates in Schwann cells and presents clinically as a slow-growing, painless, solitary nodule, that in half the cases is located in the head and neck areas, including 23% of these in the tongue.[1,2] GCT can affect patients of all ages, races and genders, but is more frequent between the third and fifth decades of life in women and people of African-American ethnicity.[2] This tumor was first described in 1926 by Abrikossoff; however, its pathogenesis has been the object of research and debate for many years. In 1935, Feyrter suggested a neural differentiation. In the 1960s, clinical and pathologic origins from Schwann cells were reported.[1,2] Clinically, it appears as an asymptomatic dermal or subcutaneous nodule, skin-coloured or brown-red, ranging from 0.5 to 3 centimeters in diameter, with slow growth and generally benign behavior.[3-5] Some patients may have warty, itchy or painful lesions.[3] Half of these tumors develop in the head and neck regions, with the tongue as the most common site.[1-3] However, any organ or tissue can be affected, such as the digestive and respiratory tracts, pituitary and parotid glands, skeletal muscles and eyes.[3,5] Multiple lesions occur in 5-25% of cases and reports in children are rare.[3] Histologically, the dermis shows polygonal cells with abundant granular eosinophilic cytoplasm, central or eccentric vesicular nucleus containing diastase resistant, and PAS positive granules. Cellular boundaries are usually inaccurate, resembling a syncytium.[4] Under electron microscope, degenerated myelinated axons are seen in the cytoplasm of these tumor cells.[3] Positive expression of S-100, CD 68, CD57, neuron-specific enolase and vimentin confirms the neural origin, although the classic form of this tumor does not carry any resemblance to other tumors of the peripheral nerve sheath.[3,4,6,7] Epidermis may present pseudoepitheliomatous hyperplasia.[7] Malignant cases represent 1-2% and usually have a poor prognosis. Differential diagnoses include melanocytic nevus, dermatofibromas and adnexal tumors. Treatment involves complete excision, because if incompletely removed the local recurrence rate is high.[4,5] ❑

Abstract: Granular cell tumor is a rare benign neoplasm of neural origin. We report the case of a female patient, 27 years old presenting a brown-red nodule in the right arm, which pathological examination showed to be formed by polygonal cells with eosinophilic granular cytoplasm and immunohistochemistry positive for S100 protein and CD68. Granular cell tumor is usually solitary and in half the cases located in the head and neck areas, 30% of these in the tongue. It is most frequent between the third and fifth decades of life in women and people of African-American ethnicity. Its origination is controversial, including the possible origins in muscle, fibroblasts, neural crest, neural sheath or histiocytes. The positivity for S-100 and CD68 suggest the neural origin.
Keywords: Granular cell tumor; S100 Proteins; Schwann cells

Resumo: O tumor de células granulares é uma neoplasia benigna rara, de origem neural. Relatamos caso de paciente feminina, 27 anos, com nódulo de superfície acastanhada no braço direito, cujo exame anatomopatológico evidenciou densa proliferação de células, com amplo citoplasma contendo grânulos eosinofílicos, e imuno-histoquímica positiva para proteínas S100 e CD68. O tumor de células granulares é geralmente solitário e, em metade dos casos, localiza-se em cabeça e pescoço, dos quais 23% na língua. É mais frequente entre a terceira e a quinta décadas de vida, em mulheres e pessoas de etnia negra. A positividade para S-100 e CD68 favorece origem neural.
Palavras-chave: Células de Schwann; Proteínas S100; Tumor de células granulares

REFERENCES

1. Ordóñez NG, Mackay B. Granular cell tumor: a review of the pathology and histogenesis. Ultrastruct Pathol. 1999;23:207-22.
2. Levavi H, Sabah G, Kaplan B, Tytiun Y, Braslavsky D, Gutman H. Granular cell tumor of the vulva: six new cases. Arch Gynecol Obstet. 2006;273:246-9.
3. Gross VL, Lynfield Y. Multiple cutaneous granular cell tumors: a case report and review of the literature. Cutis. 2002;69:343-6.
4. Sehgal VN, Verma P, Sharma S. An asymptomatic, progressive, ulcerative nodule over the anterolateral surface of the left upper arm. Clin Exp Dermatol. 2012;37:695-7.
5. Brandão M, Domenech J, Noya M, Sampaio C, Almeida MVC, Guimarães MS, *et al*.Tumor de células granulares no pé (tumor de Abrikossoff): localização infreqüente de tumor relativamente raro (Foot granular cell tumor (Abrikossoff's tumor): unusual location of a relatively uncommon tumor). An Bras Dermatol. 2001;76:215-222.
6. Rekhi B, Jambhekar NA. Morphologic spectrum, immunohistochemical analysis, and clinical features of a series of granular cell tumors of soft tissues: a study from a tertiary referral cancer center. Ann Diagn Pathol. 2010;14:162-7
7. Torrijos-Aguilar A, Alegre-de Miquel V, Pitarch-Bort G, Mercader-García P, Fortea-Baixauli JM.. Cutaneous granular cell tumor: a clinical and pathologic analysis of 34 cases. Actas Dermosifiliogr. 2009;100:126-32.

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How to cite this article: Lage TL, Miranda MFR, Bittencourt MJS, Dias CM, Parijós AM, Raiol TKA. Case for diagnosis. Cutaneous granular cell tumor: case report. An Bras Dermatol. 2013;88(6):1005-7.