Our Dermatology Online

Case Report

Abrikossoff's skin tumor: Report of two cases

Imane Gouzi[1], Layla Tahiri[1 2], Khaoula Abdellaoui[1], Sabrina Oujdi[3], Zakia Douhi[3], Sara Elloudi[3], Hanane Baybay[3], Meryem Soughi[3], Fatima Zahra Mernissi[3], Hinde El Fatemi[1,2], Laila Chbani[1,2], Nawal Hammas[1,2]

[1]Departement of Pathological Anatomy, CHU Hassan II Fez, Morocco, [2]Biomedical and Translational Research Laboratory, Faculty of Medicine and Pharmacy, Sidi Mohamed Ben Abdellah University, Fez, Morocco, [3]Department of Dermatology, CHU Hassan II Fez, Morocco.

Corresponding author: Imane Gouzi, MD, E-mail: imane.gouzi@gmail.com

ABSTRACT

Abrikossoff's tumor, also known as granular cell tumor, is a rare tumor first described on the tongue by Ivanovich Abrikossoff in 1926. It is mainly located in the head and neck regions with preferential mucosal involvement and may occur at any age and in both sexes, although with a female predominance. Herein, we report two cases: the case of a nodule under the left breast in a 47-year-old female with a history of breast cancer and of a subcutaneous lesion on the right thigh in a 46-year-old female. The diagnosis of granular cell tumor was reached by biopsy with immunohistochemical staining, then treatment was completed by a large surgical excision.

Key words: Abrikossoff; Skin; Granular cell tumor

INTRODUCTION

Abrikossoff's tumor, also known as granular cell tumor, is an uncommon neoplasm of unclear etiology and histogenesis. It is thought to be of neural origin, probably derived from Schwann cells [1]. It tends to affect all races and sexes, although it is most frequently diagnosed in black-skinned individuals, females, and between the second and fifth decades of life [2]. Clinically, it presents itself as a solitary, slow-growing, asymptomatic nodule on the head or neck region with preferential mucosal involvement. In this paper, we report two new cases of a skin granular cell tumor and discuss their epidemiological, clinical, histopathological, and therapeutic aspects.

CASES REPORT

Case 1

A 47-year-old female with a history of thyroidectomy under levothyroxine and lumpectomy for infiltrating carcinoma of the left breast followed by radiochemotherapy presented ten months before admission with a nodule under the left breast, erythematous, well-limited, and gradually increasing in volume (Fig. 1).

Ultrasound was performed and objectified the presence of a superficial, heterogeneous, subcutaneous nodule 2 cm in size.

A cutaneous biopsy revealed a granular cell tumor with a dermal proliferation of large, non-atypical cells presenting a granular cytoplasm (Fig. 2a). An immunohistochemical study revealed positive staining for PS100 and eliminated the metastatic origin of the lesion with negative staining for pan-cytokeratin (Fig. 2b).

Case Two

A 46-year-old diabetic female presented with a seven-month history of a subcutaneous lesion on the right thigh progressively increasing in size, painless and non-pruriginous.

How to cite this article: Gouzi I, Tahiri L, Abdellaoui K, Oujdi S, Douhi Z, Elloudi S, Baybay H, Soughi M, Mernissi FZ, EL Fatemi H, Chbani L, Hammas N. Abrikossoff's skin tumor: Report of two cases. Our Dermatol Online. 2023;14(1):77-80.

Submission: 15.07.2022; **Acceptance:** 25.08.2022

DOI: 10.7241/ourd.20231.16

A physical examination revealed a subcutaneous, firm, painless nodule, 3 cm in size. It was movable relative to the deep plane, fixed to the skin; the skin was erythematously pigmented (Fig. 3). Ultrasound revealed a suspicious tissue mass. A biopsy followed by a large surgical excision was performed.

A histopathological examination revealed a dermal and subcutaneous neoplasm composed of nests of cells with abundant eosinophilic granular cytoplasm and small nuclei (Fig. 4a). Pseudoepitheliomatous hyperplasia was also visible. No cellular pleomorphism, mitosis, or necrosis were observed. In an immunohistochemical study, immunostaining for the S100 protein and CD68 was positive (Fig. 4b).

According to these histologic and immunohistochemical features, the lesion was diagnosed as a granular cell tumor.

DISCUSSION

A granular cell tumor, called also Abrikossoff's tumor, is a benign, conjunctival proliferation compound of large eosinophilic cells with granular cytoplasm. It was first described in 1926 by Russian pathologist Alexei Ivanovich Abrikossoff, who believed it to be of muscular origin because of the resemblance of its cells to muscle cells (or fibers), therefore calling it *myoblastenmyoma*, translated as *granular myoblastoma*. The term *myoblastoma* is now abandoned because of evidence of neurectodermal origin, probably schwanian, its close relationship with the nerves, its immunophenotype, and the presence of vacuoles containing myelin structures under electron microscopy [1-3].

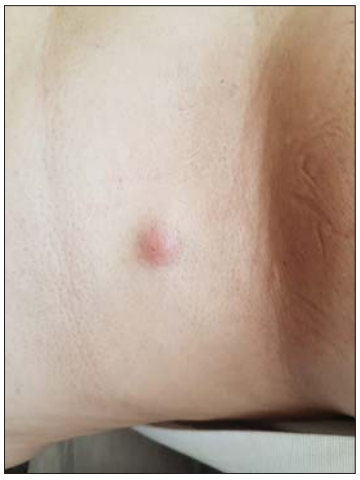

Figure 1: Subcutaneous, erythematous nodule 1.5 cm in size.

It commonly affects females between the second and fifth decades of life, most often black-skinned. Children and adolescents are rarely affected. Although it most frequently involves the head and neck regions, mainly the tongue [4,5], it has been found throughout the body: the skin, subcutaneous tissue, nerves, clitoris, vulva, glans, breast, and internal organs.

The skin lesions may present as subcutaneous nodules mimicking adnexal tumors with hyperpigmented or normal overlying skin or as a hyperkeratotic papule. It is a small lesion usually not larger than 3 cm in size, slowly-growing and firm in consistency. It is usually asymptomatic yet may be painful. Multiple granular cell tumors are not uncommon and are mainly seen in children. Solitary and multiple cutaneous granular cell tumors may be seen in association with neurofibromatosis and Noonan syndrome [6,7].

Histopathologically, the granular cell tumor reveals pseudoepitheliomatous hyperplasia, which is why care should be taken when evaluating a superficial biopsy sample to prevent the overdiagnosis of squamous cell carcinoma, because occasional tumors may be associated with mild to moderate cytological atypia in the pseudoepitheliomatous hyperplasic component. The dermis is infiltrated by cellular proliferation with syncytial, trabecular, or nested growth composed of

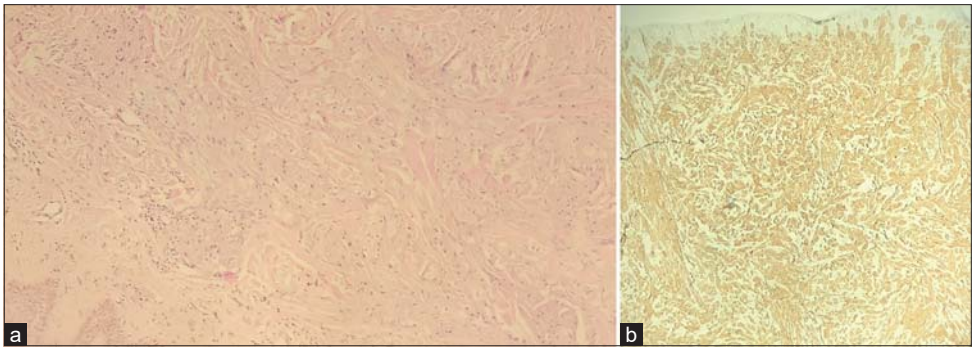

Figure 2: (a) Nets of cells with abundant granular cytoplasm (100×). (b) PS100 positive in granular cells (40×).

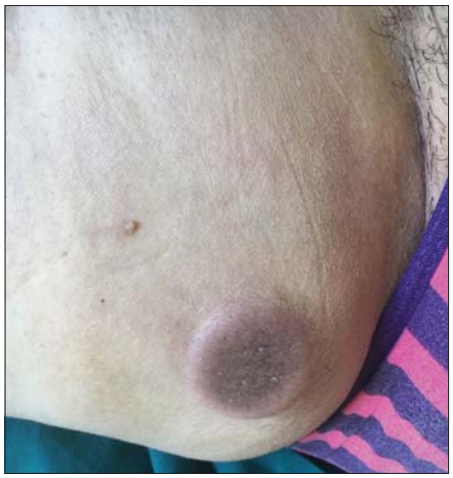

Figure 3: Well-limited, pigmented plaque with regular contours on the internal face of the right thigh.

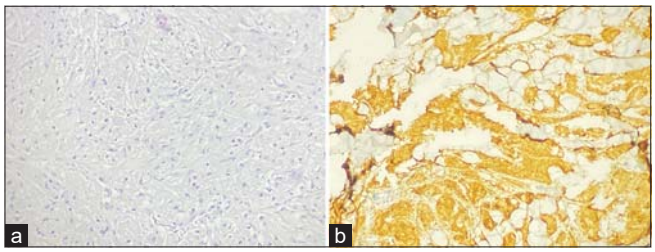

Figure 4: (a) Nets of cells with abundant granular cytoplasm (200×). (b) PS100 positive in granular cells (200×).

cells with round or oval nuclei and abundant, granular, eosinophilic cytoplasm. Vascular invasion reaching the level of subendothelial layers, without intraluminal cells, the infiltration of the erector pili muscles, and perineural extension may be seen. The latter are not criteria of malignancy, yet are just diagnostic features unrelated to prognosis. The cytoplasmic granules are positive for periodic acid–Schiff staining, the S100 protein, neuron-specific enolase, CD57, SOX10, and CD68 [8-10].

The histological diagnosis of malignancy is sometimes difficult and may be confirmed only by the existence of metastasis. Fanburg [11] described six criteria of malignancy: cells becoming fusiform, the presence of necrosis, large, nucleolus, vesicular nuclei, a high nucleocytoplasmic ratio, pleomorphism, mitotic activity (more than two mitoses per ten high-power fields).

The histologic differential diagnosis includes tumors with similar morphologic findings and with granular variants. Alveolar soft part sarcoma, atypical congenital granular cell epulis, epithelioid histiocytoma, hibernoma, rhabdomyoma, rhabdomyosarcoma, and tumors that may rarely have granular cell morphology, such as melanoma, ameloblastoma, benign fibrous histiocytoma, leiomyoma, leiomyosarcoma, angiosarcoma; undifferentiated pleomorphic sarcoma, and atypical fibroxanthoma are included [12-13].

The treatment consists of a large surgical excision with safe margins, which is not always easy given the poor limitation of the tumor. The few cases of recurrence are explained by incomplete excision. The Mohs surgical technique reduces excision margins, particularly in certain locations such as the external genitalia and extremities [14]. Regular and long-term overseeing is recommended to detect a possible recurrence or malignant transformation.

Our two patients benefited from the large excision of the nodules. Follow-up showed a good evolution with no recurrence or metastasis.

CONCLUSION

In this article, we have reported two new cases of Abrikossoff's skin tumor. The diagnosis of the granular cell tumor is established by a histological examination completed with immunohistochemical staining. The usual treatment is complete surgical excision.

CONSENT

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients gave their consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

REFERENCES

1. López V, Santonja N, Jordá E. Granular cell tumor on the sole of a child: A case report. Pediatr Dermatol. 2011;28:473-4.
2. Abraham T, Jackson B, Davis L, Yu J, Peterson C. Mohs surgical treatment of a granular cell tumor on the toe of a child. Pediatr Dermatol. 2007;24:235-7.
3. Muscardin LM, Paradisi M, Provini A, Cota C, Marzetti G. Multiple cutaneous granular cell tumors, joint hypermobility and mild facial dysmorphism in a child. Int J Dermatol. 2006;45:847-50.
4. Gross VL, Lynfield Y. Multiple cutaneous granular cell tumors: A case report and review of the literature. Cutis. 2002;69:343-6.
5. Dimosthenous K, Righi A. Granular cell tumor of the parotid gland: An exceptionally rare occurrence. Int J Surg Pathol. 2008;16:213-4.
6. Martin RW 3rd, Neldner KH, Boyd AS, Coates PW. Multiple cutaneous granular cell tumors and neurofibromatosis in childhood:

A case report and review of the literature. Arch Dermatol. 1990;126:1051-6.
7. Moos D, Droitcourt C, Ranchere-vince D, Marec Berard P, Skowron F. Atypical granular cell tumor occurring in an individual with Noonan syndrome treated with growth hormone. Pediatr Dermatol. 2012;29:665-6.
8. WHO Classification of Head and Neck Tumours, 4th Edition, International Agency for Research on Cancer Lyon, 2017.
9. Battistella M, Cribier B, Feugeas JP, Roux J, Le Pelletier F, Pinquier L, et al.; Cutaneous Histopathology Section of the French Society of Dermatology. Vascular invasion and other invasive features in granular cell tumours of the skin: A multicentre study of 119 cases. J Clin Pathol. 2014;67:19-25.
10. Nasser H, Ahmed Y, Szpunar SM, Kowalski PJ. Malignant granular cell tumor: A look into the diagnostic criteria. Pathol Res Pract. 2011;207:164-8.
11. Fanburg-Smith JC, Meis-Kindblom JM, Fante R, et al. Malignant granular cell tumor of soft tissue: Diagnostic criteria and clinicopathologic correlation. Am J Surg Pathol 1998;22:779–94.
12. Brenn T. Pleomorphic dermal neoplasms: A review. Adv Anat Pathol. 2014;21:108-30.
13. Neelon D, Lannan F, Childs J. Granular Cell Tumor. [Updated 2021 Nov 12]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022. Available from: https://www.ncbi.nlm.nih.gov/books/NBK563150
14. Gardner ES, Goldberg LH. Granular cell tumor treated with Mohs micrographic surgery: Report of a case and review of the literature. Dermatol Surg. 2001;27:772-4.

Source of Support: Nil, **Conflict of Interest:** None declared.