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Case Report

Abrikossoff Tumor of the Right Medial Thigh: A Rare Localization of a Very Rare Tumor

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Submitted: 11 January 2022

Accepted: 12 February 2022

Published: 14 February 2022

ISSN: 2573-1564

OPEN ACCESS

Keywords

- Abrikossoff tumor
- Inner side of the right thigh

Abstract

Abrikossoff's granular cell tumour is rare, developing between the ages of 20 and 60, with a female predominance, ubiquitous location and a predilection for the mucosa of the ENT and oral cavity. We report here the case of an Abrikossoff's tumor localized on the inner side of the right thigh.

Usually benign and single but can be malignant and multifocal. The pathogenesis has long been debated but recent studies favour a Schwannian origin confirmed by immunohistochemical study.

The treatment of granular cell tumours is surgical, allowing a diagnosis of certainty by anatomical-pathological examination of the excisional piece.

The outcome is often favourable if complete surgical resection is performed.

INTRODUCTION

First described in 1926 by Abrikossoff, granular cell tumours are benign and unique in the vast majority of cases. The main locations are the oral cavity, followed by the subcutaneous tissues of the head, neck and breasts.

CASE REPORT

A 68-year-old female patient, diabetic for 5 months on oral antidiabetics, hypertensive for 4 years on Amlodipine, consulted for an isolated painless nodular lesion, indurated on the inner side of the right thigh, progressively increasing in volume, with a purplish aspect in regard of the skin. The clinical examination revealed a painless, indurated, mobile subcutaneous nodular plaque measuring 3 cm in diameter with a central pigmented retraction on the inner side of the right thigh (FIG1).

The general examination did not reveal any palpable adenopathy or organomegaly. A biopsy was then performed showing a slightly verrucous papillomatous epidermis with sheets of cells infiltrating and dissecting the collagenous fibers, monomorphic, with abundant granular cytoplasm and monomorphic rounded nuclei without significant atypia at the dermal level. IHC study showed diffuse expression of CD68 and PS100 by the cells, thus concluding to a granular cell tumour appearance (FIG2). An MRI was performed to see the extention of the tumour and showed a superficial tissular mass of the inner thigh with irregular contours, 35mm in diameter with no infiltration of the underlying muscular planes, alongside a hyperintensity in T2 with an inflammatory appearance of the adjacent soft tissues, the deep muscular plane is respected with a good permeability of the right femoral vascular pedicle.

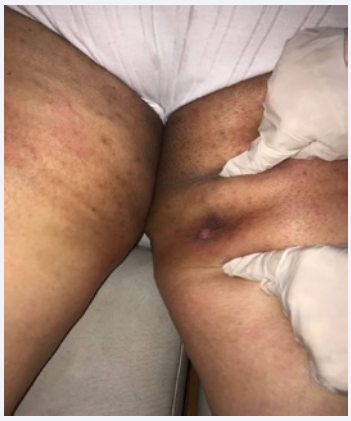

Figure 1 Nodular plate facing the inner side of the right thigh.

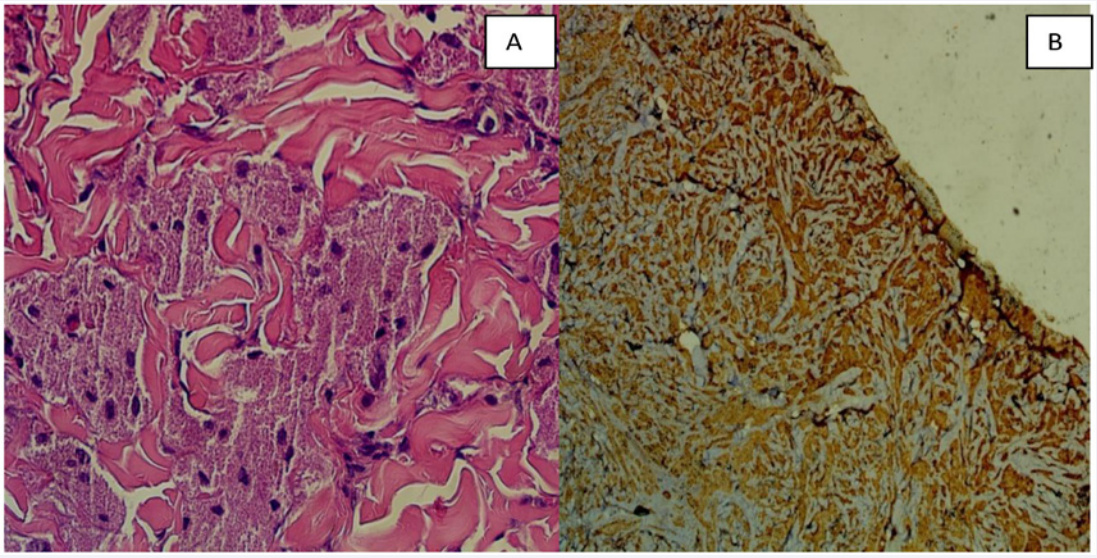

Figure 2 A : Haematoxylin eosin (HE)×400 . B: immunohistochemical -PS100 X 40: Epidermis with a slightly warty, papillomatous appearance with layers of cells which infiltrate and dissect the collagenous, monomorphic bundles, with abundant granular cytoplasm and rounded monomorphic nuclei without significant atypia at the dermal level. The immunohistochemical study showed diffuse expression of CD68 and PS100 by the cells.

A large excision was performed and confirmed the diagnosis of a granular tumour.

DISCUSSION

The particularity of our observation lies in the rarity of this tumour as well as its location within our patient.

Granular cell tumor (GCT) or Abrikossoff's tumor is a neurogenic tumor often unique and benign and with a cervico-facial localization, rare, represents 0.019% to 0.03% of tumors [1] which can be multifocal in 16 to 25% of cases and malignant in 1 to 3% of cases [2], it is observed essentially within adults between 20 and 60 years of age of black race with a female predominance (sex ratio 2, Pediatric and congenital forms are exceptional [1]. It occurs mainly in the cutaneous-mucosal area, essentially in the cervicocephalic region (50% of all cases), the lingual location is the most frequent (half of all cases), followed by the buccal floor and the palate. The external genitalias are affected in 5-15% of cases and the digestive tract in 1-8%. Other locations are even more rare: bladder, breast, salivary glands, nasopharynx, orbit, meninges, anus....No case on the inner side of the thigh has been reported to our knowledge.

It appears as a centimetric nodule (1-3cm), of hard consistency, raised or embedded, with a warty or ulcerated surface. It is yellow, reddish-brown, purplish or with an off-white color, well-limited and mobile within the deep planes and slow-growing, asymptomatic or can be rarely painful [3]. The main differential diagnoses are with other soft tissue tumours, in particular those affecting the nerve (neurofibroma) or with a muscle origin (rhabdomyoma). [4]

The diagnosis is easily made on histological examination showing a proliferation of large, polygonal cells, a small stroma with abundant cytoplasm loaded with fine eosinophilic granulations and hyperchromatic nuclei. On the immunohistochemistry , the neurogenic markers S100 protein (100% of cases) and Neurone Specific Enolase/NSE (90% of cases) are positive with an absence of expression of myoglobin desmin and neurofilaments.

Most cases of Abrikossoff's tumours are benign, but a malignant form exists although it is very rare and clinically suspected when having a size larger than 4 cm, the presence of necrotic or haemorrhagic areas and rapid growth. Histologically, Funburg-Smith et al. developed a more precise classification with six criterias (tumour necrosis, spindle cells, vesicular nuclei with large nucleoli, mitotic index greater than 2 per ten fields, high nucleocytoplasmic ratio and pleomorphism) [5] . The presence of at least three of these criterias qualifies the tumour as malignant.

The treatment of choice is based on complete removal of the tumour with clear margins; incomplete removal of the tumour requires a revision surgery to avoid recurrence. With our patient, the tumour was removed in its entirety. No recurrence was noted, with a current follow-up of nine months.

For malignant forms the safety margins must be up to 2- 3 cm, the risk of local recurrence is 32 to 41% of cases and metastatic recurrence 50 to 63% of cases) with an average delay of 2 years in 77% of cases. These forms require an extension assessment to look for secondary lymphatic or systemic localisations, mainly within the lung, liver or bone. Adjuvant radiotherapy is discussed by some authors, but chemotherapy has not proven its effectiveness.

REFERENCES

1. Ayadi, L, Khabir A, Fakhfakh I, Abdelmoula MH, Makni S, et al. Tumeur à cellules granuleuses. , 2008; 109: 158-162.
2. Lahmam Bennani Z, Boussofara L, Saidi W, Bayou F, Ghariani N, et al. Tumeur d'Abrikossoff purement cutanée chez 2 enfants. Archives de Pédiatrie. 2011; 18: 778-782.
3. Plantier F. La tumeur à cellules granuleuses ou tumeur d'Abrikossoff. Annales de Dermatologie et de Vénéréologie. 2013; 140: 399-402.

4. Dudrap E, Lemierre G, Auquit-Auckbur I, Courville P, Milliez PY. Tumeur d'Abrikossoff : à propos d'une localisation cutanée du sillon intermammaire et revue de la littérature. Annales de Chirurgie Plastique Esthétique. 2008; 53: 521-525.

5. Tawfiq N, Sabri S, Saiss K, ouchbika Z, Benchekroun N, et al. Tumeur à cellules granuleuses : à propos d'une localisation vulvaire compliquée de métastases pulmonaires. Cancer/Radiothérapie. 2013; 17: 671-674.

Cite this article

Khedim N, Hali F, El-Fetoiki FZ, Marnissi F, Chiheb S (2022) Abrikossoff Tumor of the Right Medial Thigh: A Rare Localization of a Very Rare Tumor. JSM Gen Surg Cases Images 4(1): 1046.