CASE REPORT

Abrikossoff's tumour on the upper limb: a rare presentation

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SUMMARY

Abrikossoff's tumour or granular cell tumour is a rare entity. Most common locations are the head and neck, with only a few cases reported on the upper limbs. A 55-year-old man with a nodular lesion on the left arm resorted to surgery consultation. Nodule was firm, mobile, painless and non-ulcerated. Total excision using a Limberg flap procedure was performed. Following 3 months of follow-up, the patient is fine. Abrikossoff's tumour is frequently presented in the second to sixth decade of life as an ulcerated nodule with progressive growth. Malignant form is rare, with metastases occurring in up to 3% of patients. Excision must be accomplished with free margins. Recurrence is rare. Abrikossoff's tumour on the upper limbs is rare. Although benignity is the rule, doctors must be aware of the possibility of harbouring a cancer. Surgery is the treatment of choice.

BACKGROUND

Abrikossoff's tumour or granular cell tumour is a very rare entity. It was first described in 1926 by the Russian pathologist Abrikossoff, and although benignity is the rule, there are case reports of malignancy.^{1–3} The incidence of granular cell tumour is reported to be 0.017% to 0.029%.⁴ It may arise from several soft tissue and visceral sites, but the most common locations are the head and neck (45%–65%), from which more than half are located in the oral cavity, mainly in the tongue (23%–28%).^{1–6} However, other locations can be affected and Abrikossoff's tumour has been described in the oesophagus, larynx, trachea, parotid gland, thyroid, lungs, breast, chest wall and genitalia.^{1 2 6} Since upper limbs are rarely affected, the authors present a rare case of a granular cell tumour of the left arm.

CASE PRESENTATION

A 55-year-old male patient resorted to General Surgery consultation because of a nodule on the lateral side of the left arm. The nodule was firm, mobile (fixed to the skin but apparently freed from deeper tissues), pinkish, painless, non-ulcerated and without inflammatory signs (figure 1). It appeared spontaneously several years before, gradually growing up to 3 cm. He had no systemic symptoms. A core biopsy was performed, and an Abrikossoff's tumour was the suspected diagnosis.

TREATMENT

Patient was submitted to a total excision using a Limberg flap procedure with local anaesthesia

(figure 2). There were no postoperative complications. Due to local skin tension, stitches were removed 15 days after surgery. Cicatrisation was perfect. Pathology confirmed the diagnosis of benign Abrikossoff's tumour or granular cell tumour of the left arm, with round cells of Periodic Acid-Schiff (PAS)+ cytoplasm. No immunohistochemistry was made. Lesion was totally excised.

OUTCOME AND FOLLOW-UP

Following 3 months of follow-up, the patient is fine, without signs of recurrence.

DISCUSSION

Abrikossoff's tumour or granular cell tumour is rare, frequently presented in the second to sixth decade of life as a single ulcerated well-defined nodule with progressive growth (usually smaller than 3 cm), mainly in the subcutaneous tissue of the head, neck and oral cavity, and affecting women two to three times more than men.^{1–4} Children are rarely affected.³ Additionally, black people are three times more affected than white people.^{3 4} Colour varies from pinkish to grey or brown, and there can be multiple lesions (10%–25%) that rarely reach bigger dimension.^{2 3} Multiple granular cell tumours are uncommon and represent 7%–29% of all adult cases.⁴ Although head and neck are the most common locations of granular cell tumour, particularly the tongue and oral cavity (30%–50%) which are areas with high density of peripheral nerves, there are several case reports with tumour presenting in the breast, chest wall, respiratory track, oesophagus and less frequently in vulva, bladder, perianal area, stomach, appendix, and parotid gland. Abrikossoff's tumour on the extremities is even rarer.^{1 2 6 7} Upper limbs are so rarely involved that we could only find 21 case reports in a PubMed research (table 1).

According to some authors, granular cell tumour not presenting in the oral cavity is probably more prevalent than expected.⁶ In the case reported, the tumour presented as a 3 cm pinkish painless nodule without ulceration and growing slowly. Although its characteristics match other study data, it was located on the left arm, which is a rare location with only a few cases reported in the worldwide literature.

The aetiology of this rare tumour is unknown. The majority of studies present Schwann cells as the most probable origin, followed by histiocytes, fibroblasts or pericytes. Nowadays, the consensus

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Figure 1 Pinkish nodule on the left arm.

is that Abrikossoff's tumour has a neural or nerve sheath origin.^{1-4 6 8 9}

Because symptoms are non-specific, histological findings are essential for diagnosis.⁶ Large cells with a granular eosinophilic cytoplasm with PAS-positive and diastase-resistant granules (granularity is due to accumulation of lysosomes) and small nuclei are arranged in groups.^{1-4 6} It also shows connective tissue separated from the surrounding structures by dense fibrous proliferative tissue with pseudoepitheliomatous surface.³ In fact, granular cell tumour can be confused with a squamous cell tumour because of the pseudoepitheliomatous hyperplasia present in the overlying epithelium.¹⁻³ When epidermis is invaded, differentiation with melanoma may not be easy.² Making the diagnosis is difficult because the cytoplasm granularity is non-specific, and can be observed in some non-neural tumours.¹ Abrikossoff's tumour can be misdiagnosed with leiomyoma, dermatofibrosarcoma or angiosarcoma if only light microscopy is used.⁶ In this setting, immunohistochemistry can increase pathology accuracy. Abrikossoff's tumour cells are negative for myoglobin, Human Melanoma Black-45 (HMB-45) and Glial Fibrillary Acidic Protein (GFAP), while they are positive for protein S-100, CD-68, nerve growth factor receptor 5, neuron-specific enolase, inhibin α , microphthalmia-associated

Figure 2 Tumour was totally excised using a Limberg flap procedure. Final aspect.

Table 1 Published cases of Abrikossoff's tumour on the upper limbs

Author	n	Age (years)	Gender	Location
Pushpa <i>et al</i> ¹	1	53	Female	Right arm
Dell'Horto <i>et al</i> ²	1	23	Female	Left arm
Elkousy <i>et al</i> ⁵	3	?	?	Hand
		?	?	Hand
		?	?	Hand
Kim <i>et al</i> ⁶	3	9	Female	Right forearm
		26	Female	Right palm
		69	Female	Right middle fingernail
Sehgal <i>et al</i> ⁸	1	36	Male	Left arm
Youssef <i>et al</i> ¹⁰	1	7	Female	Right arm
Ghosh ¹¹	1	35	Male	Right arm
Usui <i>et al</i> ¹²	1	34	Male	Left arm
Benson <i>et al</i> ¹³	1	44	Male	Right arm
Lage <i>et al</i> ¹⁴	1	27	Female	Right arm
Devil <i>et al</i> ¹⁵	1	28	Male	Left arm
R DeWall <i>et al</i> ¹⁶	1 (multiple)	15	Female	Right wrist/left hand
Hazan <i>et al</i> ¹⁷	1 (multiple)	54	Female	Left forearm
Newman <i>et al</i> ¹⁸	1	8	Female	Left hand
Al-Qattan <i>et al</i> ¹⁹	1	12	Female	Left hand
Fanto <i>et al</i> ²⁰	1	48	Female	Right hand
Nasit <i>et al</i> ²¹	1	21	Female	Left hand
Total	21			

transcription factor and protein gene product 9,5.^{1 6} The positivity for S-100, CD-68 and neuron-specific enolase supports the neural origin.^{8 10} In the case presented, pathology revealed large round cells of granular eosinophilic cytoplasm with small nuclei, and focal pseudoepitheliomatous hyperplasia in the epidermis. These characteristics made the diagnosis of granular cell tumour. No immunohistochemistry was necessary.

Standard treatment is total excision with free margins, which is usually curative.^{1-3 6 11} Recurrence is unusual in benign tumours.¹³ In the case reported, a total removal by the mean of a Limberg procedure to allow primary closure was performed. No recurrence was noted after 3 months of follow-up, but surveillance is still going on.

Malignant form is rare, with metastases occurring in 1%–3% of all cases of granular cell tumour.^{1 2 8 9 11} Malignant tumours tend to metastasise and relapse within 1 year after surgical treatment, emphasising the importance of long-time follow-up.³ Although there are no consistent criteria for malignancy, size >4–5 cm, located deep in the subcutaneous tissue, ulceration of the overlying epidermis, rapid growth, vascular and/or adjacent tissue invasion, atypias and mitosis figures (>2 mitosis per 10 high-power fields at x200 magnification), necrosis, vesicular nuclei with prominent nucleoli, cell spindling, high nuclear to cytoplasmic ratio and pleomorphism increase the suspicion of harbouring a malignant tumour.^{12 4 6 8 11} Additionally, only the presence of metastases is an accepted criterion for malignancy.¹⁻³ There are reports of metastases to lymph nodes and lungs.¹² No data exist about the efficacy of radiotherapy and chemotherapy to treat Abrikossoff's tumour.³ According to these criteria, the tumour treated by the authors was benign, but a long-time follow-up is needed to exclude malignancy.

Since multiple lesions can occur with several years of interval, and recurrence is a possibility, follow-up must be on a regular

basis for a long period of time. Whole patient examination is advised in order to make an early diagnosis.⁴

Learning points

- ▶ Abrikossoff's tumour or granular cell tumour is rare, and is even more rare when presented in the upper limbs.
- ▶ Although benignity is the rule, doctors must be aware of the possibility of harbouring a cancer.
- ▶ Total excision is crucial for cure, and long-time follow-up is important.
- ▶ Recurrence is rare.

Contributors CCA: study design, data collection, manuscript writing, manuscript review. MNA: data collection, manuscript review. TC: manuscript writing, review. MS: manuscript review.

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