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GRANULAR CELL TUMOUR OF THE SKIN

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Summary

Introduction. Granular cell tumours are rare, primarily benign, which can occur at various anatomical sites. We report a case of granular cell tumor of the skin, which poses no harmful prognosis for the patient upon surgical excision. Malignant subtypes of granular cell tumour have been observed in the international literature.

Case report. In this case report we present a 64-year-old female referred from a private clinic with a recurrence of a skin tumor. The histological report concluded benign granular cell tumour, with non-radical margins. Wide surgical excision of the remnant tumour was performed, and at follow up 4 months later, no recurrence was observed.

Discussion. Granular cell tumours are rare but present in the literature by mainly case series. Malignant variants have been observed, with a high tendency of recurrence and harmful prognoses.

Conclusions. Clinical as well as radiological examination of the granular cell tumours can mimic malignancy, due to an infiltrative growth pattern. Proper histological examination is of paramount.

Key words: granular cell tumour in skin, benign tumours, abrikossoff tumour, granular cell myoblastoma

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INTRODUCTION

Granular cell tumour (GCT) is a rare, but usually benign tumour found in skin and subcutaneous tissues, bronchi, oesophagus, breast, tongue, upper respiratory structures, gingiva, and the vulva [1].

First described by Abrikossoff in 1926, they are believed to originate from Schwann cells in the nervous system and can occur at any location of the body [2,3]. GCT remains a histological and diagnostic challenge, as the morphological and immunohistochemical image can overlap in benign and malignant tumours. Benign GCT represents most cases, while metastatic GCT involves less than 2% [4,5]. Predictors of malignancy include large tumour size > 5 cm, deep soft tissue location, tumour cell necrosis, increased cellularity and > 2 mitosis per 10 high power field (HPF). However, none of these findings are pathognomonic of malignancy, vascular invasion and metastasis might be the only indicator of malignant tumors [1-3]. Treatment options for benign cases include wide surgical excision, while malignant tumours include regional lymph node staging and dissection. We report a case of benign granular cell tumour of the skin.

CASE PRESENTATION

A 64-year-old woman was referred to the Department of Plastic Surgery for removal of a recurrent benign granular cell tumour on her left shoulder. According to the histology report, a benign granular cell tumour was previously removed in the same area, non-radical. Her past medical history was unremarkable. Three weeks prior to her referral, a private plastic surgeon had tried to excise the area unsuccessfully due to lack of patient cooperation (she did not handle local anaesthesia very well). On physical examination, the tumour was hard, palpable, well-defined, and measured 25 x 35 mm. Due to the benign nature, a 5 mm margin was applied, and the tumour resected. Microscopically the specimen was composed of epidermis, dermis and subcutis. In the dermis a poorly defined tumour composed of nest and ribbons of large polyhedral cells separated by thin collagenous bands was seen. The cells had abundant coarse granular eosinophilic cytoplasm, and a small nucleus with one or two distinct nucleoli. The were no cellular atypia and no tumour cell necrosis. In immunohistochemistry analysis the cells showed positive reaction for S100 protein and CD68. The proliferation index was < 1% accessed by KI-67 and there were 0-1 mitosis per 10 HPF. The tumour reached the deeper dermis, but did not infiltrate the subcutis. The distance to resection margins was 5 mm. Histological conclusion was a benign GCT.

DISCUSSION

GCT is usually a benign, presumably neural tumour with a characteristic abundance of granular eosinophilic cytoplasm. It was first described by Abrikossoff in 1926 [2,3] as *granular cell myoblastoma*, originating from the muscle. It can occur as a small, hard nodule in the dermis, subcutis or mucosal surfaces. Common sites are intraoral areas such as the tongue, palate, and oral mucosa, where deep soft tissues and visceral organs are less common. Most common in women, the tumour can occur at any age, though mainly in the fourth and sixth decades of life. The tumour usually presents as a single, firm, painless nodule in the subcutaneous tissue. From a histological perspective, the tumour cells are arranged in agglomerated strips of isolated connective tissue, which is common in fibrous tissues and gives the nodule a hard surface. The nuclei of GCT cells are surrounded by eosinophilic cytoplasm which resembles a granular appearance, hence the name. GCTs have poorly defined margins and an infiltrative growth pattern, which explains why initial clinical examination and radiological examination of deep soft tissue GCTs mimic carcinoma. GCTs are positive for S-100 protein which are common for Schwann cells, as well as CD68 which is found in a range of different blood cells and myocytes [6,7]. Majority of GCT cases are benign and cured by a simple, radical resection. However, malignant GCT have been observed in the literature and account for less than 2 percent of known cases in the literature. Unfortunately, they tend to present recurrence and metastasize within one year after excision. Treatment options for malignant cases include regional lymph node staging and dissection, while the effects of chemotherapy and radiotherapy are questionable [4,5,8].
In 1998, Fanburg-Smith et al. [5] published an extensive series of GCT cases, dividing tumours into benign, atypical, and malignant based on six histological features: (1) nuclei pleomorphism; (2) tumour cell spindling; (3) vesicular nuclei with prominent nucleoli; (4) increased N:C ratio; (5) necrosis and (6) increased mitotic rate (> 2 mitoses/10 HPF). Benign GCT were classified as those with none of the mentioned features, atypical had 1-2 features whereas malignant variants had 3 or more

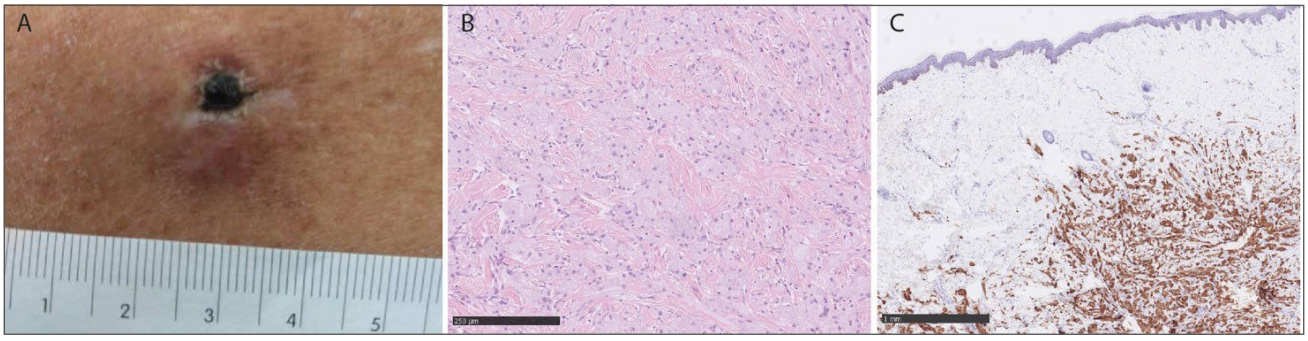

Figure 1. A) clinical picture of the granular cell tumour after initial biopsy; **B)** the tumour was composed of ribbons of large polyhedral cells separated by collagenous bands. The cells had coarse granular eosinophilic cytoplasm, and a small nucleus with one or two distinct nucleoli (H&E, 10x objective); **C)** immunohistochemical stain for S100 showed strong positive reaction in tumour cells. This also illustrates the poorly defined borders of a granular cell tumour (2.5x objective).

features. In addition, mitoses and/or necrosis in combination with a ki-67 index > 10% were also associated with malignant behavior [5].
In our case, the patient did not have an initial radical excision, which is why recurrence occurred.

CONCLUSIONS

In conclusion, GCT are primarily benign tumours found in the skin, subcutaneous tissues, intraorally and in deeper structures. The clinical presentation, presence of necrosis, as well as radiological findings with an infiltrative growth pattern, can mimic malignancy. Even though GCT are rare clinically, clinicians should be aware of their existence. Follow-up is not necessary in benign cases.

Conflict of interest statement

The Authors declare no conflict of interest.

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Author contributions

All authors have contributed equally to the publication.

Ethical consideration

Written informed consent was obtained from the patient for study participation and data publication.

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