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Case Report

A cutaneous malignant granular cell tumour: an uncommon entity with diagnostic challenge

Jyoti Bala[1], Ankur Sharma[2], Sachin Sharma[1]*

[1]Department of Pathology, Shri Lal Bahadur Shastri Government Medical College and Hospital, Nerchowk, Mandi, Himachal Pradesh, India
[2]Department of Dermatology, Shri Lal Bahadur Shastri Government Medical College and Hospital Nerchowk, Mandi, Himachal Pradesh, India

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***Correspondence:**
Dr. Sachin Sharma,
E-mail: drsharmasachin07@gmail.com

ABSTRACT

Granular cell tumour (GCT) is rare and accounts for approximately 0.5% of all soft tissue tumours. The malignant GCT (MGCT) especially cutaneous malignant granular cell tumour is extremely rare constituting 1-2% of all granular cell tumours and mostly found in the subcutaneous soft tissues of lower exrtremities, especially thighs. The uncommon occurrence of cutaneous MGCT and their histopathological similarities with other entities make diagnosis difficult in some cases. Here we report a case of 36 years old male patient who presented with a mass in the skin of right lower abdominal wall which has been increased gradually over the last one year without pain. The size of the mass is approximately 6.5 cm in greatest dimension, firm in consistency with surface irregularity and ulceration diagnosed as malignant GCT at the histopathological examination showing focal ulceration and lined by keratinized stratified squamous epithelium revealing acanthosis and pseudoepitheliomatous hyperplasia. The dermis show neoplastic epithelioid cells arranged in sheets and nests with vesicular chromatin, conspicuous to prominent nucleoli, and abundant amount of fine granular eosinophillic cytoplasm. Mitosis is more than 2/10HP. Immunohistochemical stains for S-100, CD 68 and vimentin were positive in the lesional cells.

Keywords: Malignant granular cell tumour, Immunohistochemistry, Pseudoepitheliomatous

INTRODUCTION

Granular cell tumour (GCT) were initially described by Arbrikossoff in 1926 and designated as myoblastoma originally believed to be of skeletal muscle origin, is now considered to be Schwann cell derivative based on ultrastructural and immunohistochemical analysis.[1,2] The name stems from the characteristic eosinophillic granular appearance of cytoplasm of the lesional tumor cells. These granules found in the GCT cells are considered to result from accumulation of lysosomes, similar to those found in Schwann cells. Also similar to that observed in nerve sheath derived tumours, GCT stain positive for S-100 antigen. Both these observations contribute to the current acceptance of GCT as being of neural origin.[3,4]

In the literature there is female preponderance and usually reported cases occurred between the third and fifth decade.[5] The tumour can be localized to skin or submucosa of various locations. In 30-40% cases, GCT affects skin, followed by area of head and neck, where the most common location is the tongue and oral cavity.[10] Other affected locations are the breast, the gastrointestinal tract, the respiratory tract, the thyroid gland, the urinary bladder, the central nervous system and the female genitalia.[11]

GCT are most common in the fourth to sixth decade of life but can occur at all the age groups and a study of 263 patients found that GCT were frequent in men (68%).[5,6] GCT can occur in various locations but most commonly found in the head and neck region (tongue and oral

mucosa), skin and subcutaneous and soft tissue. Most cases show benign behavior.

Malignant GCT (MGCT) is rare and represents 1%-2% of all GCT. In 1998, Fanburg-Smith et al proposed six histologic criteria for selection of atypical or malignant cases in their study of 73 cases of GCT. These criteria are: increased nuclear-to-cytoplasmic ratio; nuclear pleomorphism; necrosis; spindling of tumor cells; vesicular nuclei with prominent nucleoli; and a mitotic count of more than two in 10 high-power fields (200×field). Based on these criteria, they adopted a three-tier classification dividing them into benign (none of the criteria or focal pleomorphism), atypical (1–2 criteria), and malignant (3–6 criteria).[7] The distinction is relevant to the biologic course as MGCT is significantly more likely to recur locally and even metastasize resulting in death. Herein, we report a case of 36-year-old male with solitary GCT. Unusual features included were the site and presence of features concerning for malignant transformation. We present this case to increase awareness of an uncommon entity and to emphasize the pitfalls associated with incomplete removal and inadequate biopsy.

CASE REPORT

A 36-year-old male patient presented with mass in the skin of the right lower abdominal wall which was first noted approximately three years ago, however the size of the mass has been increased gradually over the past one year without pain. On examination well localized firm mass measuring approximately 6.5×6.5 cm was clinically located in the skin and bulged from the right abdominal wall. The patient was otherwise well, with no other medical conditions. No lymphadenopathy was noted. Wide local excision of the growth was done.

Pathology

Gross

The mass was firm and well circumscribed with 6.5 cm diameter and approximately 5 cm bulge from the skin. The surface of the mass was rough with ulceration with rim of normal looking skin circumferentially and cut section was grey white in appearance (Figure 1a and b).

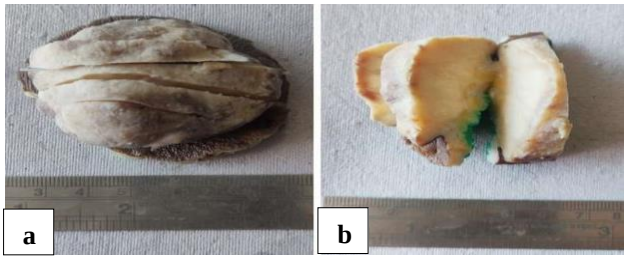

Figure 1: (a) Gross image - the surface of the mass was rough with ulceration with rim of normal looking skin circumferentially and cut section, and (b) was grey white in appearance.

Histology

Histologically surgical specimen revealed tumour covered over by skin lined by keratinized stratified squamous epithelium showing pseudoepitheliomatous hyperplasia with mild acanthosis and focal ulceration. The underlying dermis shows show sheets and irregularly arranged nests of epithelioid cells with vesicular chromatin and abundant finely granular eosinophillic cytoplasm. Moderate pleomorphism is seen. Focal areas show epidermotropism with pagetoid spread. Mitosis is >2/10HPF. Immunohistochemical stains for S-100, CD 68 and vimentin were positive in the lesional cells. Ki 67 index shows immunoreactivity in 10-15% of tumour cells.

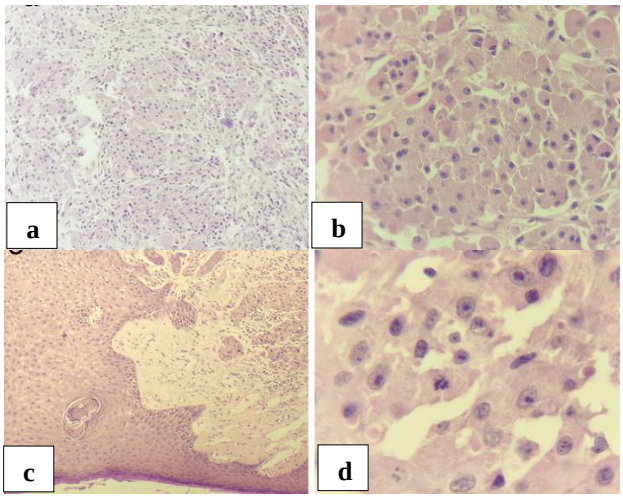

Figure 2: (a) 10x power, (b) 40x showing dermal infiltrate of epithelioid cells with abundant granular, eosinophillic cytoplasm and vesicular nuclei, (c) revealing epidermotropism with pagetoid spread, and (d) atypical mitosis.

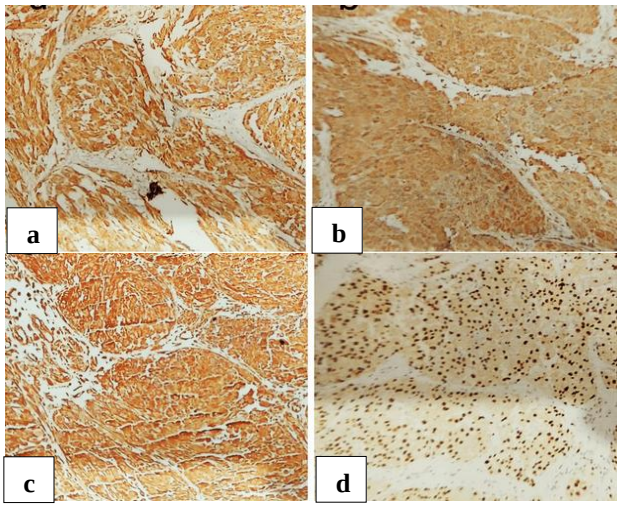

Figure 3: (a) S-100 immunostain demonstrating strong and diffuse cytoplasmic staining, (b) positive staining with CD68 immunostain, (c) immunostaining positivity for vimentin, and (d) S0X 10 immunostain nuclear positivity.

DISCUSSION

GCT remains an interesting tumour with most cases following a benign course and malignant behavior described in rare occasions. Due to increased incidence in local recurrence and metastasis, MGCT overall carry a poor prognosis. Differences in survival outcome between benign and MGCT highlight the clinical relevance of distinguishing these 2 entities. Although metastasis is the sine qua non of malignancy, the recognition of tumors with a potential for metastases is based on histologic criteria. In 1998, Fanburg-Smith et al reported a large series of GCTs, classifying them into benign, atypical, or malignant according to the presence or absence of the following 6 histologic findings: nuclear pleomorphism, neoplastic cell spindling, vesicular nuclei with large nucleoli, increased N:C ratio, necrosis, and increased mitotic rate (2 mitoses/10 high-power fields). The lesions were classified as benign if none of these findings were present or if there was only focal nuclear pleomorphism, atypical if there were only 1 or 2 findings, and malignant if 3 or more of these findings were present. Neoplasms that meet criteria of histology of malignant tumour may result in death in 40% cases because of high chance of recurrence and metastasis and those with benign potential have no recurrence vor local metastasis after adequate resection. When the tumor grows near an epithelial surface, in sites such as skin, vulva, or larynx, pseudoepitheliomatous hyperplasia occurs frequently, which may be misinterpreted as squamous carcinoma more often if the biopsy taken is superficial. Elastosis is often present in the stroma.[9]

MGCTs are aggressive neoplasms. The most common sites for metastatic spread in MGCTs are the regional lymph nodes, lung, liver, and bones.

All management of MGCT is not clearly defined and may be hampered in part both by the rarity of their occurrence in the literature and the lack of adequate follow-up. Treatments options for MGCT range from surgical excision with/without lymph node dissection, radiation, and chemo-therapy.[8] Despite the availability of multiple treatment options, all authors recommend at least surgical excision with adequate margins, and given reports of success with the addition of radiation, it is reasonable to offer such therapy to patients diagnosed with MGCT.

CONCLUSION

In summary MGCT's are uncommon neoplasms and those arising in the skin are exceptional. The skin and subcutaneous forms are easily mistaken for epidermal inclusion cyst and lipomas and if the biopsy is superficial than it can be misdiagnosed as squamous cell carcinoma if pseudoepitheliomatous hyperplasia is present. Immunohistochemistry helps in confirming the diagnosis and helps to distinguish from granular cell variants of other tumours like granular cell melanoma. Malignant GCT occurs in about 2% of cases but poor outcome establishes the importance of correct preoperative diagnosis. Surgical excision with clear margins is the best treatment however because of high metastatic potential patients should be carefully followed.

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