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A Rare Case of Cutaneous Granular Cell Tumor Misdiagnosed as Sebaceuos Cyst

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Abstract **Case Report**

Granular cell tumor (GCT) is a rare benign skin tumor. The tumor mostly presents with a symptomatic slowly growing solitary nodule and no overlying skin changes; therefore, it is not always considered in the differential diagnosis. We report a case of cutaneous GCT in the right supra-scapular region in a 43-year-old middle eastern female, who presented with a 2-year history of a slowly growing mass, with a dimension of 3 × 2 cm, initially misdiagnosed as a sebaceous cyst and later diagnosed as a GCT at the histopathological examination, followed by second excision with wide margins. The tumor cells had centrally located nuclei and granular eosinophilic cytoplasm and stained positively for S100 and CD68 antibodies on immunohistochemistry. Even though GCTs are rare, they should be thought of when diagnosing soft tissue tumors under the skin. Malignant changes have been reported in cases after long term follow-up, so surgical removal with wide margins is considered the best line of management.

Keywords: Granular Cell Tumor; Abrikossoff Tumor; Skin Malignancy; Cutaneous GCT; non-neural granular cell tumor; Soft-tissue tumor.

INTRODUCTION

Granular cell tumors (GCTs), formerly known as granular cell myoblastoma, are rare mesenchymal tumors of Schwann cell origin consisting of cells with granular eosinophilic cytoplasm, typically presenting as an asymptomatic nodule, and affecting adults [1]. There is an eight-to-one female preponderance for congenital GCTs and a three-to-one female preponderance for non-infant GCTs, with the highest prevalence among the Blacks. The tumor can be found in nearly all types of tissue. It may be congenital or acquired, occurring between 20 and 60 years of age, with a peak incidence around age 50 [2]. Typically, these tumors manifest as a non-painful mass in the subcutaneous tissue; however, they can occasionally be multicentric at the time of diagnosis. Multiple lesions have been reported to be associated with hereditary and congenital GCT cases [3].

Due to their inconspicuous presentation, these tumors are commonly misdiagnosed clinically; a histopathological examination provides the definitive diagnosis. GCTs can develop both on the epidermis and the mucosa. They are frequently encountered in the Upper respiratory tract, pharynx, breast and in the upper extremities [4]. The majority of lesions are benign, and malignant cases are reportedly rare, occurring in only 1% or 2% of cases [5-8]. We report a case of a 43-year-old woman initially misdiagnosed as a sebaceous cyst and later confirmed to be Granular Cell Tumor on histopathological examination.

CASE REPORT

A 43-year-old female patient presented with the complaint of a slowly growing mass on her back for 1 year. The lesion was not associated with pain or tenderness. There was no discharge, and it continued to increase in size. On clinical examination, a single, firm, mobile subcutaneous cystic lesion with well-defined borders was noticed. The subcutaneous mass was large in size with dimension of 3×2 cm, and there were no skin changes on the overlying skin. Laboratory parameters were within normal limit. The lesion was diagnosed as a sebaceous cyst and was excised. The histopathological examination demonstrated diffuse infiltration of the dermis with tumoral nests. These cells had granular eosinophilic cytoplasm with centrally located nuclei (Figure 1). No signs of malignancy, such as lympho-vascular invasion, necrosis, high-rate mitotic activity, or cellular atypia, were present. The

immunohistochemistry was strongly positive for S100 and CD68 antibodies (Figure 2). Ki-67 staining showed low mitotic figures (Figure 3). The diagnosis of GCT was made, and a completion procedure was carried out with wide local excision with free margins. Postoperatively, the patient's recovery was uneventful.

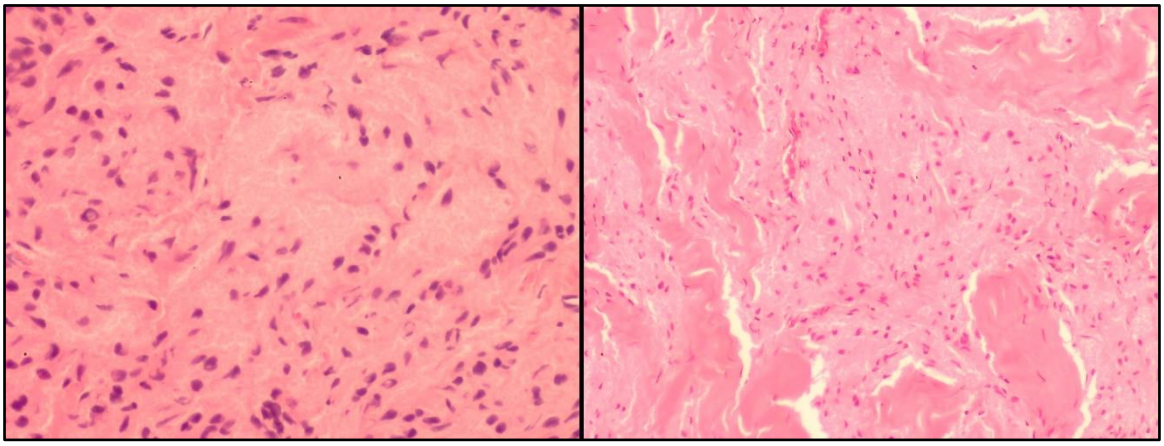

Figure 1: Nests and sheets of cells containing eosinophilic cytoplasmic granules (L) and Band of Fibrous Tissue separating the cells (R)

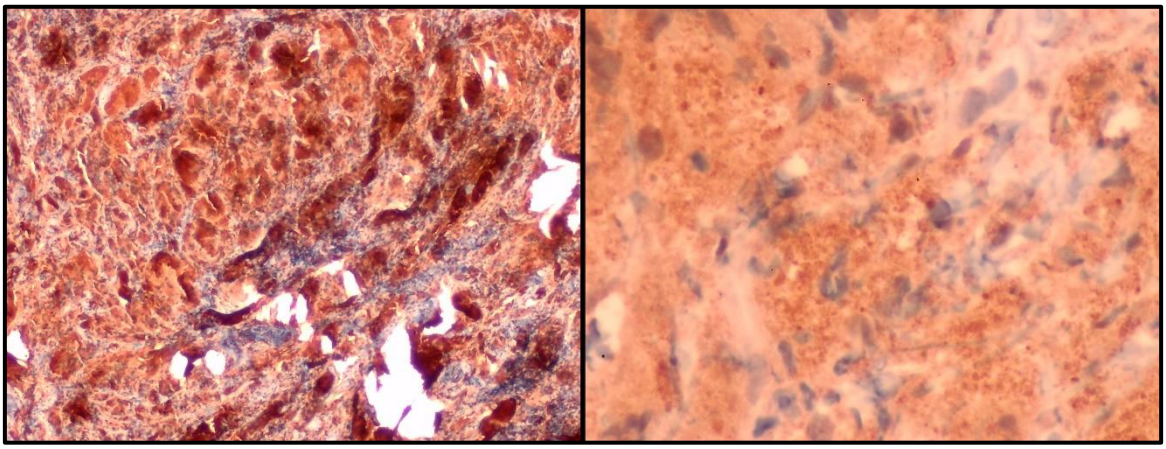

Figure 2: Positive cytoplasmic staining with CD-68 (L) and Positive staining with S100 (R)

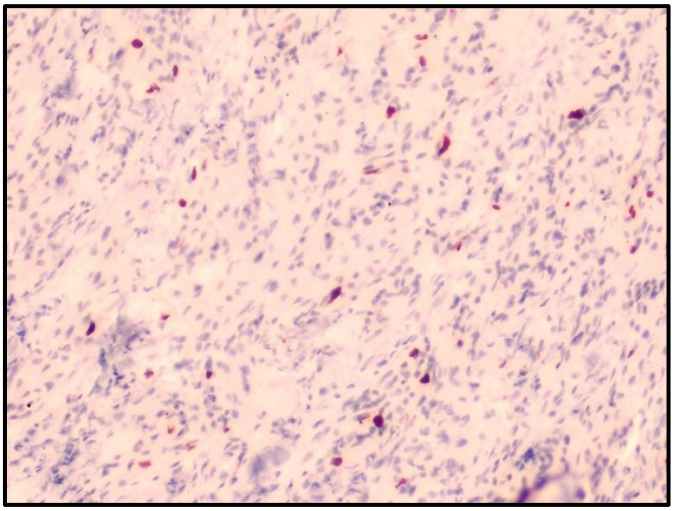

Figure 3: Low Mitotic figures in ki-67 staiuning

DISCUSSION

The Russian Pathologist, Alexei I Abrikossoff, was the first to describe a case of Benign GCT, which he found in the skeletal muscle of the tongue in 1926. He initially thought that these tumors were of skeletal muscle origin due to resemblance to skeletal muscle myocytes and named it "Myoblastoma" [9]. Although there is still debate regarding the origin of these tumors,

immunohistochemical studies have indicated that Schwann cells are primarily to blame [10, 11].

In 30%–45% of patients, the disease affects the skin, followed by the head and neck, with the most common intraoral sites being the tongue and soft and hard palate [12]. Other affected areas include the breast, the gastrointestinal tract (particularly the lower section of the esophagus), the respiratory system, the thyroid gland, the urine bladder, the central nervous system, and the female genitalia. In 5%–16% of these instances, the vulva is the major region affected, but the disease can also be seen in the cervix, uterus, and ovaries [13].

Cutaneous and subcutaneous illness is typically detected as a solitary, tiny, non-tender, slowly expanding mass, occasionally accompanied by pruritus and less frequently by discomfort. In some cases, pseudo-epitheliomatous hyperplasia of the overlying skin may be observed due to the continuous irritating effect of the tumor. This appearance can be mistaken as well-differentiated squamous cell carcinoma (SCC), whereas benign GCT is associated with other skin cancers. Differential diagnosis includes dermoid cyst, dermatofibroma, hidradenoma, prurigo nodularis, fibrosarcoma and fibroadenoma [2].

Microscopically, the cells have abundant, eosinophilic, granular, coarse cytoplasm and are polygonal or oval, less commonly spindle-shaped. The nuclei are small and dense and bands of fibrous tissue separate the cells. In 10%–15% of malignant cases, there is a peripheral infiltrative pattern with satellite nodules. In addition, the nuclei are large and vesicular containing a single or multiple nucleolus and demonstrate nuclear pleomorphism [14].

GCTs are typically immunohistochemically positive for PASD, Sudan Black B, S100 (except non-neural granular cell tumors), SOX10, and NSE. Vimentin, CD68, NKI-C3 (CD63), MITF, and CD56 can exhibit variable staining. A subset of granular cell tumors that are not neuronal express ALK. Included among the Negative Stains are HMB45, MelanA, AE1/AE3, EMA, and desmin. Although conventional GCTs frequently exhibit consistent NSE and S-100 positivity, there have been reports of unusual and inconsistent immunohistochemistry, whereas non-neural GCTs exhibit immunohistochemical diversity, likely due to their mesenchymal rather than neural or Schwannian origin [15-19]. In our case, the tumor tested positive for S-100 and CD-68.

The tumor's prognosis is determined mainly by whether it is benign or malignant. It is a widely accepted notion that benign tumors can be safely excised locally with clear margins. 1–2% of GCTs exhibit malignant features and behave like high-grade sarcomas with a significant risk of metastasis and a limited survival period [20].

CONCLUSION

Due to their inconspicuous appearance and symptomatology as a normal subcutaneous mass, GCTs are frequently misdiagnosed extemporaneously, with histological investigation eventually establishing the correct diagnosis. However, while being uncommon, the relationship of GCTs with malignancy necessitates a differential diagnosis that includes all probable benign tumors and malignancies in relation to the anatomic site of presentation.

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