Granular cell tumor of the hand: A case report

Daniela Fanto, Salvatore Fanto

ABSTRACT

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Case Report: We report a case of a granular cell tumor of the hand.

Conclusion: This case is of particular interest due to the atypical location, and clinical presentation, which despite its malignant potential, mimics that of more commonly encountered benign lesions such as ganglion cysts and inclusion cysts.

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CASE REPORT

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Keywords: Hand, Soft tissue, Granular cell tumor, Subcutaneous tissue

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INTRODUCTION

Granular cell tumor (GCT) is a rare soft tissue neoplasm of presumed neuroectodermal origin [1, 2]. The majority are benign, and a malignant variant accounts for only 1–3% of cases [3, 4]. In a large series by Lack et al. of 110 patients diagnosed with GCT, none were found to be malignant. Granular cell tumor is usually located in the head and neck with hand and peripheral nerve lesions being distinctly uncommon [3]. Lesions are most commonly found in the dermis or subcutaneous tissue, thus mimicking adnexal tumors. Demographically, GCT more commonly affects females in their 40s or 50s of African-American origin [5]. However, in the Lack et al. series, reported incidence was 35% in females and 65% in males [6].

The lesion is usually asymptomatic, slow growing, solitary and measuring less than 3 cm [4, 7]. Due to this non-specific presentation, GCT is rarely clinically suspected prior to biopsy or surgical excision [6]. Hallmark features, although not pathognomonic of malignancy, include larger size, intramuscular location, invasion of adjacent structures and rapid growth [6, 8].

The case outlined is of particular interest as it presented as a more commonly encountered benign dermal subcutaneous lesion. As GCTs do have a potential for malignancy and for metastases, this tumor should be kept in mind as a differential diagnosis.

CASE REPORT

A 48-year-old African-American female presented with a nodule on the dorsum of her right hand, which she first noticed a few months earlier. She reported that the nodule had been slowly increasing in size.

Physical examination revealed a hard non-tender round nodule approximately 1.5 cm in diameter overlying the extensor tendons of the index and long fingers. The lesion was located in the subcutaneous plane dorsal to the extensor tendons without infiltration of surrounding structures. The excision of nodule was performed under local anesthesia (Figure 1).

Daniela Fanto¹, Salvatore Fanto²

Affiliations: ¹MBBS, Bachelor of Medicine, Bachelor of Surgery, NHS (National Health Service), UK; ²MD, Doctor of Medicine, Ingalls Memorial Hospital Harvey, Illinois.

Corresponding Author: Daniela Fanto; Email: danielafanto@doctors.org.uk

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Pathology revealed a granular cell tumor, specifically the lesion consisted of a population of relatively large polyhedral cells with finely granular cytoplasm due to high mitochondrial content and small, dense and central nucleus (Figures 2–4). The lesion had a cellular center with a peripheral infiltrating pattern, splitting

Figure 1: Granular cell tumor specimen after excision under local anesthesia.

Figure 2: On histopathological examination there is mild nuclear atypia in focal areas. No cellular pleomorphism, mitotic activity, or necrosis (H&E stain, x200). Immunohistochemically, the cells stain with S100 and CD56. PAS stain shows cytoplasmic staining.

Figure 3: On histopathological examination the lesion consists of relatively large polyhedral cells with finely granular cytoplasm and small, dense central nucleus (H&E stain, x400).

Figure 4: On histopathological examination the lesion has a cellular center with a peripheral infiltrating pattern, splitting the adjacent collagen fibers, with ill-defined borders (H&E stain, x100).

the adjacent collagen fibers, with ill-defined borders. There was mild atypia in focal areas with no cellular pleomorphism, mitotic activity, or necrosis (Figures 2–4). Immunohistochemically, the cells stained with S100 and CD 56 and cytoplasmic staining was PAS positive as well. Follow-up at third year revealed that the patient remained symptom free and without any signs of recurrence.

DISCUSSION

Abrikossoff first described GCT in 1926. The lesion was located in the tongue and was thought to arise from muscle. Consequently, the neoplasm was referred to as granular cell myoblastoma [9]. Fust and Cyter [10, 11] postulated that non-lingual lesions originated from peripheral nerves and coined the term granular cell

neuroblastoma, whereas Lack et al. used the currently accepted term granular cell tumor [6].

Granular cell tumor is commonly found in the dermis or subcutaneous tissue with intramuscular lesions being uncommon [4, 10]. The tumor has been documented in various sites in the body. However, the head and neck areas appear to be most commonly affected. Hand lesions are rare. In a large series involving 95 patients, Strong et al. reported that most lesions were located in the head and neck area, with only seven involving the fingers and two palms [3]. Lack et al. reported on 110 patients during a 32-year period, and found that 38 tumors arose from the oral mucosa, and 48 from the skin and subcutaneous tissue, most commonly the trunk and upper extremities [6]. Differential diagnoses for such lesions include the more commonly encountered ganglion cysts, inclusion cysts, fibromas and neuromas.

Preoperative workup varies depending on the history and the clinical findings. In some cases, X-ray and MRI scan have been used. Typically, MRI scan is most useful for the preoperative workup of larger lesions for both diagnostic purposes and preoperative planning. MRI findings indicative of GCT include heterogeneous low signal intensity on T1-weighted images and a central low signal with peripheral high signal on T2-weighted images with some lesions being darker on T1-weighted and T2-weighted images than fat [8]. Although these findings may be helpful during diagnostic workup such features are not specific or sufficient in establishing a definitive diagnosis.

Further management includes FNA and wide excision, the latter being the favored and most definitive therapy. Histologically, the tumors demonstrate polygonal sheaths of cells with distinct borders, eosinophilic granular cytoplasm and a small central nucleus [6, 8]. Cytoplasmic granules are PAS+ and diastase resistant [6]. Immunohistochemical staining is typically positive for S100 protein, neuron specific enolase, laminin and various myelin proteins [12]. S100 protein positivity is indicative of neural derivation, as this protein is associated with neural ectodermal cells. Histological differential diagnoses may include but are not limited to renal cell carcinoma, granuloma, alveolar soft part sarcoma and hibernoma. Histopathological diagnostic criteria for malignancy have been studied by Fanburg-Smith et al., who reported a case series of 73 patients with malignant soft tissue GCT. We conclude that the presence of three or more of the following histologic criteria confirms the diagnosis of a malignant lesion: necrosis, spindling, vesicular nuclei with large nucleoli, increased mitotic activity (>2 mitoses/10 high-power fields at x200 magnification), high nuclear to cytoplasmic (N:C) ratio, and pleomorphism [13]. A further adverse prognostic factor in terms of survival is a Ki67 proliferation index less than 10% [13].

Recurrence of benign GCTs following wide excision is rare and most likely associated with incomplete resection [6]. Lack et al. reported an 8% recurrence rate in patients

diagnosed with GCT who were treated surgically, and in each case, one or more the surgical margins of resection had tumor involvement [6]. Adequate follow-up is imperative as malignant lesions can recur and may metastasize, making these the most definitive features of malignancy [14].

Given that we do not currently know the possible mechanism of malignant transformation, close follow-up is essential.

CONCLUSION

It is important to be aware that granular cell tumor presentation typically mimics that of commonly occurring benign lesions, such as ganglion cysts and fibromas. Although granular cell tumors are rare, it is important to consider the possibility of this potentially malignant diagnosis.

Author Contributions

Daniela Fanto – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Final approval of the version to be published

Salvatore Fanto – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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