# Malignant Granular Cell Tumor

# 166

## Definition

Malignant granular cell tumor (MGCT) is a malignant neoplasm classified among the nerve sheath tumors due to its origin from Schwann cells. A benign counterpart exists (→ see Chap. 114).

## Epidemiology and Presentation

MGCT is an extremely rare neoplasm which accounts for less than 2% of all granular cell tumors. It occurs in a wide age range (mean of 40 years), with a female predominance. Despite a Schwann cell phenotype, the origin from a nerve cannot be usually identified.

The most frequent sites of involvement are the soft tissue of the thigh, proximal arm, trunk, and then distal extremities. Less commonly it affects the head and neck. The lesion is 1–20 cm in diameter (median: 4 cm) and usually locates in subcutaneous or intramuscular tissues.

## Pathology

MGCT is a sarcoma composed of malignant granular cells characterized by abundant cytoplasmic lysosomes filled with amorphous granular material (hence the name). Most cases have high-grade spindle or polygonal cell morphology with eosinophilic granular cytoplasm, high mitotic rate (>2 mitoses per 10 HPF), geographic necrosis, marked pleomorphism, and high nucleus-to-cytoplasm ratio.

A minority of cases are morphologically bland and not overtly malignant in appearance, although they still behave aggressively (i.e., give rise to metastasis). Some investigators argue that metastasis remains the only definite criterion for malignancy.



S. Mocellin, *Soft Tissue Tumors*, https://doi.org/10.1007/978-3-030-58710-9_166

## Biomarkers

Like the benign counterpart, MGCT stain is strongly and diffusely positive for S100 and CD68.

## Prognosis

MGCT is highly aggressive with metastatic disease in up to 50% of patients (about 12% to lymph nodes, the remaining to distant sites). Larger primary tumor size, local disease recurrence, metastasis, and older patient age are adverse prognostic factors.

## Therapy

Surgery is the treatment of choice for localized primary tumor. Due to the rarity of the disease, no standard therapy exists for metastatic disease; besides conventional chemotherapy, target therapy (e.g., pazopanib) has been utilized in anecdotal cases.

## Suggested Readings

Bradford Bell (2020) Benign and malignant granular cell tumor of the hypopharynx: two faces of a rare entity. Head Neck Pathol. [Epub ahead of print]

Cheng (2014) A rare case of pulmonary malignant granular cell tumor detected with 18F-FDG PET/CT imaging. Clin Nucl Med 39(9):816–818

Conley (2014) Dramatic response to pazopanib in a patient with metastatic malignant granular cell tumor. J Clin Oncol 32(32):e107–e110

D'hulst (2018) 18F-FDG PET/CT and MRI of a mediastinal malignant granular cell tumor with associated recurrent pericarditis. Clin Nucl Med 43(8):589–590

Fletcher (2020) WHO classification of tumours of soft tissue and bone (5th edition)

Moten (2018) Malignant granular cell tumor: clinical features and long-term survival. J Surg Oncol 118(6):891–897

Morita (2015) Pazopanib monotherapy in a patient with a malignant granular cell tumor originating from the right orbit: a case report. Oncol Lett 10(2):972–974

Nasser (2011) Malignant granular cell tumor: a look into the diagnostic criteria. Pathol Res Pract 207(3):164–168

Pérez-González (2015) Primary cutaneous malignant granular cell tumor: an immunohistochemical study and review of the literature. Am J Dermatopathol 37(4):334–340

Quinn (2019) Malignant granular cell tumor of the bile duct. ACG Case Rep J 6(8):e00193

Wei (2015) Whole-genome sequencing of a malignant granular cell tumor with metabolic response to pazopanib. Cold Spring Harb Mol Case Stud 1(1):a000380