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Primitive Polypoid Granular-Cell Tumor[☆]



Tumor primitivo polipoide de células granulares

In contrast to conventional granular cell tumor (Abrikossoff tumor), primitive polypoid granular cell tumor was first identified by LeBoit et al.¹ in 1991 and subsequently endorsed by Chaudhry and Calonje² as a dermal tumor of granular cells of non-neural origin. The tumor has a polypoid morphology and presents numerous mitoses, cytologic atypia, and a primitive immunophenotype. We present a new case and review the characteristics of this rare and poorly known tumor.

Our patient was a woman aged 44 years, with no past medical or family history of interest. She consulted for an asymptomatic lesion that had appeared 4 months earlier at the right nasolabial angle. Physical examination revealed a hard, polypoid lesion of 0.3 mm diameter, with a translucent surface. With a possible diagnosis of fibrous papule, milium cyst, or adnexal tumor (trichodiscoma), the lesion was excised. Histopathology revealed a circumscribed proliferation of cells in the superficial and mid dermis, surrounded by an epithelial collarette (Fig. 1). The cells, arranged in an interlinked fascicular pattern, had a polygonal morphology with abundant, granular eosinophilic cytoplasm and large vesicular nuclei (Fig. 2). Mitotic figures were present. No ulceration or necrosis was observed. Immunohistochemistry was positive for CD68 and negative for AE1-AE3, S-100, Melan A, CD34, desmin, actin, and smooth muscle. On the basis of these findings we made a diagnosis of primitive polypoid granular cell tumor.

Granular cells can be found in a varied group of tumors and reflect the intracytoplasmic accumulation of lysosomes and other components of the Golgi apparatus. Traditional and

conventional nomenclature makes reference to the cutaneous and mucosal granular cell tumor, known as Abrikossoff tumor, a benign neoplasm of neural origin derived from Schwann cells.³ However, other granular cell tumors of non-neural origin exist, including the congenital gingival granular cell tumor and the primitive polypoid granular cell tumor. In addition, numerous tumors can present granular cells, including myogenic tumors, melanocytic lesions, dermatofibroma, dermatofibrosarcoma protuberans, basal cell carcinoma, atypical fibroxanthoma, angiosarcoma, fibrous papule, ameloblastoma, and adnexal tumors with eccrine or apocrine differentiation.⁴

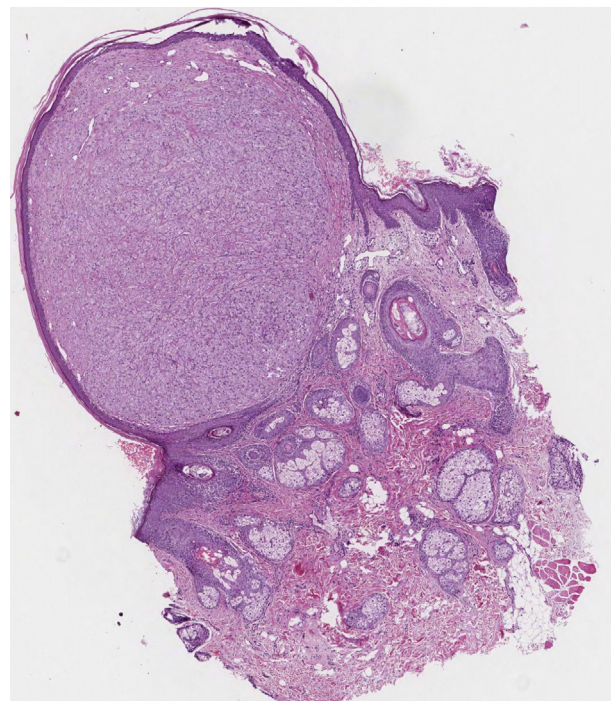


Figure 1 Tumor in the superficial and mid dermis, surrounded by an epithelial collarette. Hematoxylin-eosin, original magnification $\times 10$.

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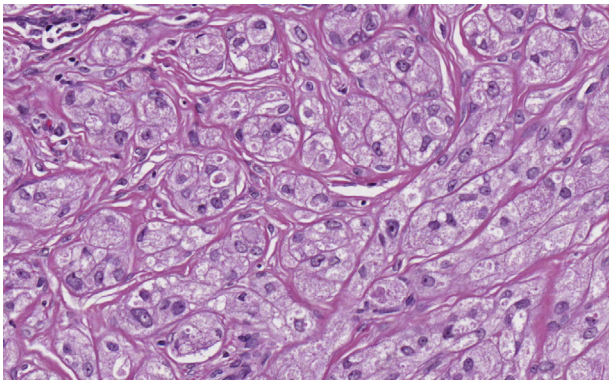


Figure 2 Cells with a polygonal morphology, abundant granular eosinophilic cytoplasm, and large vesicular nuclei. The cells are arranged in an interlinked fascicular pattern. Hematoxylin-eosin, original magnification $\times 40$.

Primitive polypoid granular cell tumor is a rare neoplasm or uncertain origin that affects middle-aged adults. It usually arises on the trunk or limbs as a polypoid or elevated lesion with a smooth surface; its size can be variable. The distinctive characteristic that differentiates it from conventional granular cell tumor is its histopathology, which shows a tumor in the mid dermis that is clearly surrounded by an epidermal collarette; the tumor is formed of large polygonal, round or spindle-shaped cells with marked nuclear pleomorphism, an elongated nucleus, abundant eosinophilic cytoplasm containing fine granules, and mitotic activity of around 1 to 3 mitoses per mm,² usually with no atypia and minimal or absent epidermal hyperplasia. These histopathology findings satisfy the criteria proposed by Enzinger and Weiss for the classification of malignant or atypical granular cell tumor, but the tumors can be differentiated by immunohistochemistry, as primitive polypoid granular cell tumor is usually negative for S-100 and positive for CD68 and neuron-specific enolase.⁵

Despite the histological characteristics, this is a tumor of low-grade malignancy. In the series reported there is only 1 case of metastasis, which arose in the cheek 25 months after excision of the lesion and did not present an epidermal collarette on histology.⁶

We have presented a new case of primitive polypoid granular cell tumor, a variant that is not clearly distinguished from the conventional tumor. Its atypical histological characteristics allow it to be classified as a new entity and distinguished from granular cell tumor of neural origin.

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Dermal Leiomyosarcoma at the End of the Left Eyebrow[☆]



Leiomiosarcoma dérmico en la cola de la ceja izquierda

Cutaneous sarcomas constitute less than 1% of superficial soft tissue neoplasms. Leiomyosarcomas (LMSs), which account for between 3% and 6.5% of cutaneous sarcomas, are

classified as dermal (derived from the hair erector muscle) or subcutaneous (derived from the smooth muscle of vessel walls).¹ This classification has prognostic relevance, as dermal LMS has a more favorable clinical course and outcome; metastasis occurs in 5% to 10% of dermal cases as compared with 30% to 40% of subcutaneous cases.²

We report the case of a 63-year-old man with a medical history of hypertension, hyperuricemia, and dilated cardiomyopathy, who presented with an asymptomatic and progressively growing nodular lesion in the ciliary region of the left eye that had appeared 6 months previously (Fig. 1). Physical examination revealed a raised, indurated, and erythematous lesion, with destruction of hair follicles, that was surrounded by a halo of firm, edematous skin.

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