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. in 1991 and subsequently endorsed by Chaudhry and Calonje 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 poligonal 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. 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 ×10.
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). 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.

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.

We have presented a new case of primitive polyloid 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.

References


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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. Hematoxylineosin, original magnification ×40.