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Material submitted:

1 H&E slide
Protocol

Granular Cell Tumor of the Eyelid

History and clinical findings:

A 15-years old girl presented with a painless, solitary nodule on the eyelid. Unfortunately, this is all the history we have. Resection of the mass was performed.

Histopathology:

Microscopic examination shows a lesion composed of clusters of large cells with abundant granular, eosinophilic cytoplasm and uniform, round nuclei. There are no nucleoli. Clusters of tumor cells are separated by dense collagen bands. Mitoses, cytological atypia, necrosis or vascular invasion are not present. The lesion extends into adjacent soft tissue. Tumor cells form concentric rings around small nerves within the tumor. Tumor cells are PAS positive and immunopositive for SOX10 and S100.

Final pathology diagnosis: Granular cell tumor

Discussion:

Granular cell tumor is a benign tumor that occurs virtually in every organ in the body. Most commonly present in the head and neck (50%) with preponderance to the oral cavity, especially the tongue. Other common sites include soft tissue, skin, breast and lung. Granular cell tumors have been described in patients of all ages but are most common in adults, ages 30-60. Clinically, the tumors appear as painless, slow growing nodules, and are often misdiagnosed clinically in the skin and soft tissues as sebaceous cysts or fibromas. When affecting organs (breast, esophagus), may clinically mimic a malignancy.

Review of the ophthalmic literature shows reports of granular tumors in the orbit, lacrimal sac, eyelid, conjunctiva, and caruncle. In the eyelid, the tumors present as a discrete painless nodule, painless, often near eyelid margin and may mimic a chalazion. In the orbit, the tumor arises in the orbital soft tissue and may involve extraocular muscles. It is often symptomatic.

Multiple cutaneous tumors occur in 5-20% of the cases and may be familial. Rare cases are associated with tumor syndromes (for example neurofibromatosis type 1).

Histologically the tumor is composed of characteristic plump cells arranged in nests and embedded in dense collagen. Tumor cells have abundant eosinophilic granular

cytoplasm, and small, round eccentric nuclei. Mitoses are rare. The tumor is not encapsulated and often infiltrates into adjacent tissues. A characteristic feature is the formation of concentric whorls of tumor cells around myelinated nerve twigs included in the lesion. Skin lesions may have associated pseudoepitheliomatous hyperplasia of overlying epithelium.

The cytoplasmic granules are PAS positive, diastase resistant and stain red with trichrome stain. Tumor cells are immunopositive for S100, SOX10 and CD68.

On EM the granular cells surrounded by a basement membrane. The granular cells contain cytoplasmic membranous vacuoles containing electron dense material and lipid (phagolysosomes). Between tumor cells are cells with characteristics of fibroblasts and axons.

Although very rare, malignant granular tumors have been reported. The malignant granular cell tumors have frequent mitoses. Clinical behavior is usually indolent.

Non-neural (S100 negative) granular cell tumor of skin (Primitive polypoid granular cell tumor) is histologically similar but a distinct entity. First described in 1991 by LeBoit, these are cutaneous granular cell tumors that are negative for neural crest markers, but strongly and diffusely positive for CD63 (lysosomes). These are often polypoid and may present ulcerated epithelial surface. Nuclear atypia and frequent mitoses are often present and there may be associated inflammation. Clinical behavior is uncertain, rare metastasis have been reported. ALK fusions were found in a subset (60%) of these tumors.

The first description of granular cell tumor was by Abirkosoff in 1926 and it was initially believed to be of skeletal muscle origin or myoblastic differentiation, hence there are multiple names in the older literature including Abirkosoff tumor and myoblastoma. Currently, the cell of origin is believed to be Schwann cell based on diffuse expression of S100 protein and close anatomic relationship to peripheral nerve fibers.

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