Clinical and Microscopic Features of Oral Cellular Neurothekeoma

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Description

Despite its name, the genesis of the rare benign tumour known as cellular neurothekeoma is unknown. Since cellular neurothekeoma stains for fibrohistiocytic markers and PRAME whereas nerve sheath myxoma stains for neural markers, there is still debate in the literature as to whether nerve sheath myxoma should be included in the morphologic spectrum of neurothekeoma. There have been some recorded hybrid forms. Gallager and Helwig first coined the term "Neurothekeoma" (NTK) in 1980 to describe a rare type of benign cutaneous tumours originating from nerve sheath. The term "nerve sheath myxoma" has been used to describe this phenomenon in the past. Later, a subtype of cellular NTK was recognised based on histopathological and immunohistological characteristics. Mucosal involvement has only occasionally been described, and it typically manifests as a cutaneous lesion. When it comes to the cellular variant, these numbers are even lower.

Neurothekeoma is histologically classified into myxoid, intermediate, and cellular kinds based on the quantity of myxoid matrix present. The myxoid neurothekeoma variant's neural ancestry is broadly accepted. However, the origin of the cellular variant is uncertain because it lacks both consistent neural immunoreactivity and ultrastructural proof of neural differentiation. Neurothekeomas often develop in the first three decades of life, with more women than men being affected. Fetsch and colleagues reported patient ages ranging from 20 months to 85 years, with a mean age of 21 years, in their analysis of 178 malignancies. The ratio of men to women was roughly 1:2. Similar to this, Hornick and Fletcher reported patient ages ranging from 1 to 65 years, with a mean age of 25 years, in a study of 133 cases of cellular neurothekeoma. The ratio of men to women was 1:1.8. Only a few specific features of neurothekeoma have been identified by limited ultrastructural studies. According to studies in immunohistochemistry and ultrastructure, the myxoid variant is thought to be schwannian.

Cellular neurothekeoma's genesis is still up for debate. According to some writers, the ability of cellular neurothekeomas to exhibit myxoid areas identical to those of classic myxoid neurothekeomas proves that these tumours share a neuronal differentiation. According to some writers, cellular neurothekeoma may differentiate into myofibroblasts or fibrohistiocytes. Additionally, it has been suggested that cellular neurothekeoma is a subtype of plexiform fibrohistiocytic tumour, atypical fibroxanthoma, or cellular dermatofibroma, however distinctions in immunohistochemical and ultrastructural features raise the possibility that neurothekeoma is a distinct entity.

Clinical Features

The head and neck are the most typical locations for neurothekeomas. The nose and scalp are the areas that are involved the most frequently. The orbital region, cheekbones, and chin come after these areas. Other often reported areas of involvement include the trunk, upper and lower extremities, and both. There have also been reports of neurothekeomas in the eyelids, paranasal sinuses, and oral mucosa. Neurothekeomas are solitary, flesh-colored nodules that are asymptomatic to mildly painful upon skin examination. Overlying erythema may have an accompanying component that is present. The nodules typically have a diameter of less than 2 cm, clinically epidermal inclusion cysts, intradermal nevi, lipomas, pilomatrixomas, and dermatofibromas can all be mistaken for neurothekeomas.

Microscopic Features

Grossly, neurothekeomas look like nondistinctive skin tumours that are typically 0.5 cm to 1 cm in size. Neurothekeomas are composed histologically of spindled epithelioid cells with an abundance of eosinophilic, finely granular cytoplasm. The cells are distributed throughout the dermis in numerous, closely spaced nodules of various sizes. The largest dimension of the removed lesion is 8 mm. Hematoxylin and Eosin (H&E) stained slides under the microscope revealed an unencapsulated neoplasm made up of spindle to epitheloid cells that were grouped in well-formed micronodules with an appearance of concentric whorls. The mucosa was heavily engaged, and considerable submucosal invasion was seen. The majority of the cancerous cells exhibited modest nuclear atypia and fibroblastic appearance. Within the whorled nodules, there were sporadic large cells of the osteoclast type. Mitotic activity was almost nonexistent, and no myxoid matrix was visible. Typically a cutaneous ailment, mucous membranes are only infrequently affected by cellular NTK. It is even less likely that cases of oral mucosal involvement will occur.

Diagnosis

The differential diagnosis of NTK frequently includes melanocytic tumour, particularly in high risk patients such those with a history of radiation treatment. Although these neoplasms share several immunological and histological characteristics, such as the presence of NKI/C3 and spindle and epitheloid cells arranged in whorls, NTK is typically negative for protein S100 and may be distinguished from melanocytic lesions. Smooth Muscle Actin (SMA) has been positively detected in 60% of cellular neurothekeomas, and these tumours are typically diffusely positive for Neuron-Specific Enolase (NSE). Although neurothekeoma is regarded as a benign tumour, broad local excision with frozen-section margin management is advised because of the risk of recurrence and local invasion.

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