Neurothekeoma of the Tongue: CT, MR, and FDG PET Imaging Findings

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**Summary:** We report CT, MR, and fluorodeoxyglucose–positron-emission tomography (FDG-PET) imaging findings of a case of cellular neurothekeoma of the tongue, a rare benign soft-tissue tumor with neural differentiation, occurring in a 15-year-old girl. CT and MR imaging showed a well-defined, well-enhancing submucosal soft-tissue mass in the midline dorsal tongue. There was high FDG uptake on PET scans. Although imaging findings are rather nonspecific, neurothekeoma may be one of diagnostic inclusions of soft-tissue masses of the tongue in a young female patient.

**Discussion**

Neurothekeoma is a rare benign soft-tissue tumor and is defined as "benign myxoma of cutaneous nerve sheath origin." It was first described in 1969 by Harkin and Reed under the name of "nerve sheath myxoma." In 1980, Gallager and Helwig coined the term "neurothekeoma" (in Greek, "theke" means sheath) after review of 53 dermal tumors showing similar histologic features. On the basis of histologic and immunohistochemical findings, there are 3 variants of neurothekeoma: myxoid (classic or hypocellular), cellular, and mixed.

Microscopically, the myxoid form is different from the cellular form by its greater degree of myxomatous change and by less cellularity, and is characterized by lobules of well-circumscribed stellate and spindle cells in a myxoid matrix, multinucleated giant cells, and fibrous connective tissue. In contrast, the cellular form shows no evidence of encapsulation, with lobules arranged in nests in which the cells are epithelioid with eosinophilic cytoplasm. A minority of cells are plump or spindle and may reveal nuclear atypia and variable mitotic figures. The mixed type of neurothekeoma usually shows areas of varied cellularity that contain focal myxoid regions.

A consistent immunohistochemical profile for neurothekeoma has not been established in both myxoid and cellular neurothekeomas. Some markers traditionally expressed by neural tumors, such as S-100 protein, neuron-specific enolase, epithelial membrane antigen, neurofilament, and glial fibrillary acidic protein, have frequently been used to clarify the cellular origin of neurothekeoma. Classically, the myxoid form produces positive results of stains for S-100, glial fibrillary acidic protein, collagen type IV, and nerve growth factor receptor and does not stain for epithelial membrane antigen or markers of histiocytic differentiation. In contrast, the cellular form typically does not stain with antibody to S-100, collagen type IV, or nerve growth factor receptor but consistently shows reactivity with NK1C3 (CD57) and the panmonocyte marker Ki-M1p. Although it is widely accepted that myxoid neurothekeoma is derived from nerve sheath, controversy still exists regarding the cell of origin in the cellular variant. Although most authors agree with a nerve...
sheath origin for both types and consider that these 2 variants represent extremes of a histologic continuum, others believe that they have separate origins with cellular neurothekeoma being derived from perineural fibroblastic or fibrohistiocytic differentiation. \cite{Kim1}

Dual immunoreactivity for neuron-specific enolase and S-100 protein supports a Schwann cell origin, whereas absence of S-100 protein and a positive detection of epithelial membrane antigen suggest a perineural-cell lineage.\cite{Kim2}

Clinically, neurothekeoma is typically a slowly growing usually asymptomatic dermal or, less frequently, mucosal or submucosal tumor of adolescents and young adults, occurring 2–4 times as often in women as in men.\cite{Kim3, Kim4, Kim5} According to a meta-analysis of 292 patients with neurothekeoma studied by Papadopoulos et al,\textsuperscript{3} ages at diagnosis ranged from 15 months to 84 years with a mean age of 28 years and an average lesion diameter of 1.2 cm. The most common location was the upper extremity (33.6%), followed by the head and neck (29.4%), the trunk (17.2%), and the lower extremity (9.7%). In 9.3% of patients, the tumor was present on mucosa or submucosa and approximately 0.8% was spinal. Literature review revealed about 20 reported cases of neurothekeoma occurring in the oral cavity.\cite{Kim6, Kim7, Kim8} The most common site affected in the oral cavity was the tongue, followed by the buccal mucosa, lower lip, palate, and retromolar area. In contrast to extraoral cases that commonly occurred in the second decade of life, the mean age of the patients with oral neurothekeoma was 33 years.\cite{Kim4}

Imaging findings of neurothekeoma have been described in only a few cases.\cite{Kim9, Kim10, Kim11} The paucity of reported imaging findings might be mainly ascribed to its small size and dermal location. There are no pathognomonic radiologic findings of neurothekeoma; thus, diagnostic confirmation should rely on histologic examination. On CT scans, the tumor is reported as a hypoattenuated-to-isoattenuated mass with variable enhancement and vascularity patterns. In 1 reported case of cellular neurothekeoma of the tongue, it was seen as an irregular hyperintense mass on T2-weighted MR images.\cite{Kim12} In cases of tumors involving the intracranial cavity or paranasal sinus, they can appear as large aggressive-looking soft-tissue masses causing erosion of the adjacent bones.\cite{Kim12, Kim13} In the present case of lingual neurothekeoma, the tumor was seen as a well-defined round soft-tissue mass in the midline dorsal tongue on CT and MR imaging. Compared with the adjacent normal intrinsic tongue muscle, the tumor was isoattenuated to T1-weighted and hyperintense on T2-weighted MR images. The mass enhanced relatively well after contrast administration. T2-weighted MR images also showed hypointense interlacing septa within the tumor, which corresponded histologically to fibrous connective tissue. Radiologic differential diagnoses of lingual neurothekeoma include neurilemmoma, neurofibroma, hemangioma, minor salivary gland tumor, soft-tissue sarcoma, and other rare soft-tissue tumors such as granular cell tumor.

To our knowledge, findings of neurothekeoma on FDG-PET have not been reported. In the present case, substantial homogeneous uptake with a maximal SUV of 5.4 was seen, which caused additional confusion about the nature of the tumor. However, the FDG uptake of schwannomas, which are neighbor tumors of neurothekeomas, can be variable; thus, FDG-PET has limited value for identifying benign-versus-malignant peripheral nerve sheath tumors.\cite{Kim15} Although the reason high FDG accumulation is found in benign tumors such as schwannoma remains unclear, the wide range of FDG uptake may be explained by the different degrees of cellularity for each lesion.\cite{Kim15}

The histopathologic differential diagnosis of lingual neurothekeoma includes the neural tumors, such as neurilemmoma and neurofibroma, and the non-neural tumors or tu-
morlike conditions, such as oral focal mucinosis, soft-tissue myxoma, malignant fibrous histiocymata, myxoid melanoma, ossifying fibromyxoid tumor, soft-tissue sarcoma with myxoid changes, chondroid choristoma, and ectomesenchymal chondromyxoid tumor.\textsuperscript{11,16} However, all these entities have distinctive microscopic features that usually give a clue to the definitive diagnosis. Although aggressive local recurrences or distant metastases have not been reported, minor recurrence limited to the primary tumor site can occur secondary to incomplete removal; thus, wide local excision is the standard treatment of neurothekeoma.\textsuperscript{3}

**Conclusion**

We report CT, MR, and FDG-PET imaging findings of a case of cellular neurothekeoma of the tongue, which is a very unusual tumor of probable nerve sheath origin. Although imaging findings are rather nonspecific, neurothekeoma may be included in the differential diagnosis of soft-tissue masses of the tongue in a young female patient.

**References**