Pathology Quiz Case 1

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5-YEAR-OLD BOY PRESENTED WITH A SEVeral-year history of a mass in the left side of his neck. The mass had fluctuated slightly in size since its onset but had recently become more noticeable. The patient had no history of pain, fevers, or other cervical masses. His medical history was significant only for neurofibromatosis 1 (NF1). Physical examination revealed a nontender, 2×2-cm left-sided neck mass that was located in the submandibular and jugulodigastric region. There was a visible fullness in the submucosal region of the left anterior floor of the mouth. A computed tomographic scan revealed an irregularly shaped, centrally hypodense lesion in the left submandibular region.

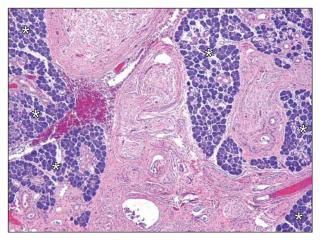


Figure 1.

An excisional biopsy was performed to establish the diagnosis. Intraoperatively, the lesion appeared to be in direct continuity with the left submandibular gland, with multiple fingerlike projections into the surrounding soft tissue. The gland and associated mass were excised. Cytologic examination of the pathology specimens revealed a lesion involving the submandibular gland, with separation of gland acini (**Figure 1**, asterisks). An alternative low-power magnification view demonstrated enlargement of multiple nerve bundles surrounding a duct (**Figure 2**, arrowhead) of the submandibular gland. Immunohistochemical stains were positive for S100 protein (**Figure 3**). However, the acinar portion of the submandibular gland was negative for S100 protein (**Figure 4**).

What is your diagnosis?

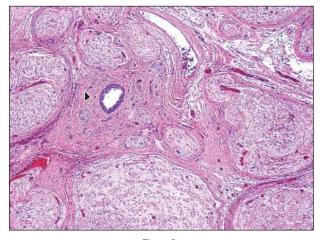


Figure 2.

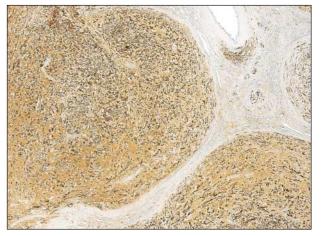


Figure 3.





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