Lower Neck Neurilemmoma Can Masquerade as Lymph Node Metastasis on FDG PET/CT in Patient With Nasopharyngeal Carcinoma

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Abstract: A 50-year-old man with nasopharyngeal carcinoma was suspected to have progressive malignant disease because of an unexpected FDG-avid finding that was suspected to be neck lymph node metastasis on the follow-up FDG PET/CT after the definite anticancer therapy. However, after carefully correlating with the relevant MRI findings because of the atypical depth of the neck lesion and possible connection with the spinal nerve, the FDG-avid finding was reconsidered to be a tumor that originated from the nerve. The subsequent histologic examination confirmed the diagnosis of neurilemmoma.

Key Words: neurilemmoma, FDG PET/CT, MRI, nasopharyngeal carcinoma, lymph node metastasis, head and neck

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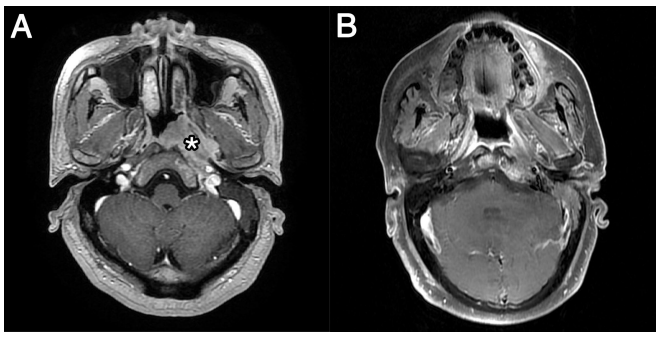

FIGURE 1. A 50-year-old man was diagnosed with nasopharyngeal carcinoma. The initial presentation on the magnetic resonance imaging (MRI) with gadolinium enhancement revealed an enhanced bulging mass in the left nasopharynx with adjacent skull-base invasion (asterisk in panel **A,** T1-weighted fast spoiled gradient echo image with gadolinium enhancement) without locoregional lymph node or distant metastasis. After completing the concurrent chemoradiation therapy, several studies were performed for restaging, including the endoscopic examination, MRI, and 2-[^{18}F]fluoro-2-deoxy-D-glucose positron emission tomography/computed tomography (FDG PET/CT). The follow-up MRI revealed disappearance of the previous tumor in the left nasopharynx (panel **B,** T1-weighted fast spin-echo image with gadolinium enhancement) in accordance with the normal endoscopic finding, suggesting good response to the anticancer therapy.

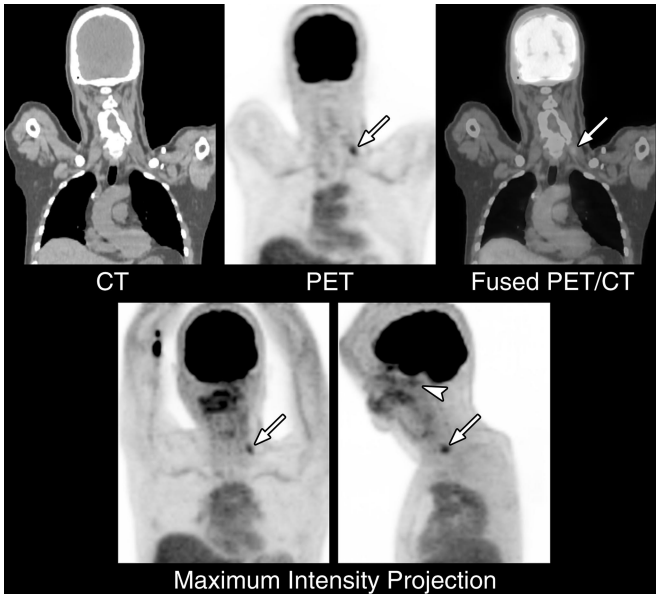

FIGURE 2. The FDG PET/CT demonstrated mildly increased radioactivity in the nasopharyngeal region (arrowhead) that was probably due to the inflammation. However, there was an unexpected focus with intense radioactivity (SUVmax: 3.6) in the left lower neck region (arrows). Initially, the finding was suspected as lymph node metastasis that would be regarded as progression of malignancy. However after carefully examining the anatomic reference on CT component of FDG PET/CT, the lesion seemed unusually deep and possibly within the left scalene muscle.

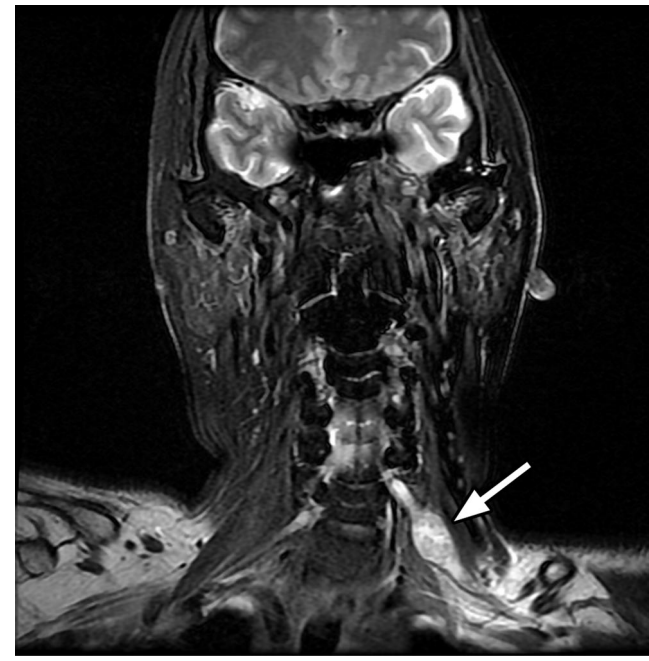

FIGURE 3. After reexamining the aforementioned follow-up MRI, we noticed a small, well-circumscribed, hyperintense lesion with relatively central hypointensity in the left lower neck region on the T2-weighted image with fat suppression (arrow). In addition, the location of this lesion seemed connecting with the root of left sixth cervical spinal nerve. The image features revealed that the lesion might originate from the spinal nerve, and most frequently, it might be a periphery nerve sheath tumor such as neurilemmoma.

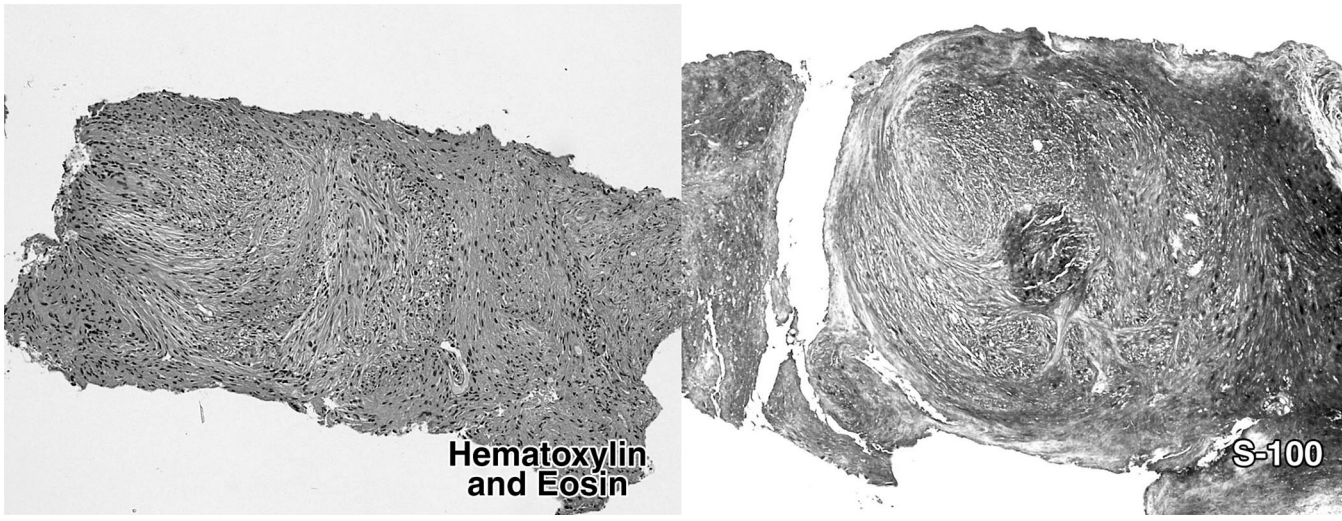

FIGURE 4. This patient then underwent ultrasonography-guided needle biopsy for pathologic confirmation. The histologic findings revealed spindle tumor cells arranged in fascicles with focal nuclear palisading (hematoxylin and eosin stain, 100×) and the immunohistochemical study showed positive staining for S-100 protein (100×), which were characteristics for neurilemmoma and confirmed the diagnosis. Neurilemmoma is a benign, slow-growing, encapsulated nerve sheath tumor derived from Schwann cells and is most commonly observed in the extremities and head and neck regions.[1,2] It is also known as schwannoma, neurinoma, neuroma, and perineural fibroblastoma and is the most common tumor of the peripheral nervous system.[1,3–5] Neurilemmoma can originate from virtually any peripheral nerve, except the olfactory and optic nerves that lack Schwann cells.[5,6] This tumor occurs at all ages, but is most commonly found in individuals between the second and fifth decades.[1] Neurilemmoma in the parapharyngeal space with the vagus nerve or the cervical sympathetic chain serving as the nerve of origin is the second most common site of head and neck region after the acoustic neuroma in the vestibular region.[5] A neurilemmoma in the neck region may be difficult to differentiate from a solitary metastatic node from an unknown or known primary cancer without periadenitis or extranodal extension of disease.[7] In addition, neurilemmoma may mimic other diseases in the head and neck region, such as thyroglossal duct cyst, branchial arch cyst, carotid body tumor, thyroid cyst/nodule, thyroid carcinoma, parotid cyst/tumor, paraganglioma, chemodectoma, aneurysm of internal carotid artery, lipoma, meningioma, and rhabdomyoma.[6,8,9] Characteristic histologic features of neurilemmomas include palisade arrangement of nuclei and Verocay bodies.[10] The encapsulation and the presence of 2 components, a highly ordered cellular component (Antoni A area) and a looser myxoid component (Antoni B area), distinguish neurilemmoma from neurofibroma.[11] Immunohistochemical studies with S-100 stain may be helpful for diagnosis of neurilemmoma.[12] High FDG uptake of neurilemmomas has been reported in those most common sites of extremities[4,13] and head and neck regions[14] as well as other uncommon sites such as the esophagus,[15] stomach,[2] bronchus,[16] liver,[3] appendix,[17] and colon.[18,19] Therefore, it may be difficult to distinguish a neurilemmoma from malignant peripheral neural sheath tumor or other malignancies based on the FDG PET/CT findings only. MRI may be helpful for providing the anatomic details and characteristic findings of neurilemmoma. Typical MRI findings of neurilemmoma manifest low-to-intermediate signal intensity on T1-weighted images, heterogeneous hyperintensity on T2-weighted images, and intense heterogeneous postgadolinium enhancement.[7,20] Similar to most neurogenic tumors of the head and neck region, the majority of neurilemmomas present as asymptomatic head and neck masses at the time of presentation.[21] The treatment of neurilemmoma is surgery, but may be left for those with growing potential and compression symptoms,[22] because there may be a considerable morbidity because of surgical treatment in some patients.[12] The current case reveals an incidental finding of focal FDG-avid lesion that masquerades neck lymph node metastasis in a patient with nasopharyngeal carcinoma. However, the unusual depth of the neck lesion on the CT component of FDG PET/CT is an important clue for differential diagnoses and may suggest etiologies other than lymph node metastasis. The concurrent MRI provides further evidences of the characteristic features and anatomic details that strongly suggest the origin from the nerve. Therefore, it is important that when interpreting an FDG PET/CT scan for a known malignancy, the clinicians should keep in mind that any FDG-avid lesion presenting with an atypical anatomic location relevant to the etiology of known malignancy should not be simply regarded as a variant presentation of malignancy. Other more probable explanation should be considered and examined, so that the patient will not be treated improperly.