Perineural Invasion through the Maxillary Division of the Right Trigeminal Nerve in a Rare Case of Nasolabial Malignant Peripheral Nerve Sheath Tumor

C. Sempere-Ortega
J. Martinez-San-Millan

SUMMARY: We report the CT and MR imaging findings in a patient with malignant peripheral nerve sheath tumor in the right nasolabial area, which exhibited typical imaging features of cystic adenoid carcinoma.

We present a rare case of a patient without neurofibromatosis type 1 (NF-1) and with a malignant peripheral nerve sheath tumor (MPNST) arising in the terminal branches of the maxillary nerve in the right nasolabial area with retrograde perineural invasion of the entire nerve all the way to the pterygopalatine fossa. The location, behavior, and imaging features of our patient prompted the radiologic diagnosis of cystic adenoid carcinoma; however, pathologic analysis of the lesion confirmed the diagnosis of MPNST. To our knowledge, there is no previous imaging report of a lesion of this nature with this location and behavior.

Case Report

We present the case of a 62-year-old man with a previous history of facial trauma who complained of nose pain and nocturnal breathing problems. Findings of the clinical examination were completely normal. Several months later, the patient came back to our center because pain and breathing problems had increased and he could not continue his normal activities. In the new clinical examination, a soft-tissue mass was observed swelling on the right nasolabial fold and on the vestibule of the mouth, with no associated mucosal lesion; therefore, a submucosal lesion was suggested. CT and MR imaging revealed the presence of a homogeneous enhancing solid mass in the right nasolabial area (Fig 1A, -B), apparently located in the labial submucosa (Fig 1A), which invaded the right maxillary nerve all the way to the pterygopalatine fossa (Fig 1A, -B). The maxillary nerve was enlarged (Fig 2), with a homogeneous tubular-shaped increase of its diameter (Figs 3A and 1A, -B), resulting in a remodelling of the infraorbital canal (Fig 3B). A cystic adenoid carcinoma was suggested on the basis of the imaging and clinical findings. The patient underwent surgery, and the pathologic examination, surprisingly, was consistent with MPNST. Neither family history nor stigmata of NF-1 was found in the patient.

Discussion

MPNSTs are rare lesions derived from the mesenchymal cells of the neural crest. The term “MPNST” is now preferred instead of terms like “neurofibrosarcoma,” “neurogenic sarcoma,” or “malignant schwannoma” because the originating cell is not clearly understood.1 MPNSTs represent approximately 6% of all malignant soft-tissue tumors.2,3 These lesions have a tendency to affect the trunk and extremities, and only 10% of all MPNSTs are located in the head and neck region.7 The most common nerve affected is the sciatic nerve, followed by the brachial plexus.2 Between 38%–50% of these malignant tumors are associated with NF-1, and this entity is also related to multiplicity.2,5 The risk for developing an MPNST is 4600 times greater in patients with NF-1 than in the general population.4
The optimal treatment is combined surgical excision with radiation therapy. The nerve infiltration resulted in a nerve enlargement and a homogeneous tubular-shaped increase of the nerve diameter, but not in the typical spindle shape of MPNST. Multidetector CT allowed curved reconstructions that were very illustrative of the particularly perineural invasion exhibited by the tumor. However, MR imaging is necessary in this type of lesion because CT is a limited tool for demonstrating intracranial extension to the cavernous sinus, Meckel cave, and the cisternal and pontine portions of the cranial nerve V.

The differential diagnoses in the presence of a swelling in the nasolabial fold involve nasolabial cyst, odontogenic inflammatory process, odontogenic tumor, squamous cell carcinoma, soft-tissue sarcoma, bone lesion, lymphoproliferative disorder, and salivary gland tumor. On the basis of the location, solid nature, and enhancing pattern of the lesion after intravenous contrast administration and the perineural invasion exhibited, the differential diagnosis was narrowed to solid malignant tumors. Given the absence of an associated mucosal lesion, cystic adenoid carcinoma was suggested as the first diagnostic possibility.

**References**


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**Fig 3.** A. Curved axial reconstruction of a contrast-enhanced CT scan shows the original mass in the right nasolabial (arrow) area followed by a tubular-shaped enlargement of the maxillary nerve (curved arrow). Same image as in A with bone windowing. B. Note the infraorbital canal enlargement (arrow).