

# Solitary neurofibroma of the floor of the mouth

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## ABSTRACT

We present a case of neurofibroma in the floor of the mouth. A 74-year-old female complained of painless swelling in the right side of the floor of the mouth for 3 months. Clinical examination and CT revealed a tumor involving the right side of the floor of the mouth. Histological features were characteristic of a neurofibroma. Such a tumor arising in the area of the mouth is very rare. We review the clinical and pathological features of a neurofibroma arising in the mouth.

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Oral peripheral nerve sheath tumors are rare, and include neurofibroma, schwannoma, palisaded encapsulated neuroma, nerve sheath myxoma, mucosal neuroma associated with multiple endocrine neoplasia type III, traumatic neuroma and granular cell tumor.<sup>1,2</sup> We rarely encounter solitary neurofibroma in the oral cavity. The sites in order of frequency are tongue (the most common), buccal mucosa, floor of the mouth, palate, lips and gingiva. Pathologic distinction between neurofibroma and schwannoma is essential, as the former shows a much greater propensity for malignant transformation, especially when associated with Von Recklinghausen's neurofibromatosis.<sup>3</sup> The objective of this paper is to discuss the clinical presentation, pathological characteristics, management and follow up of solitary neurofibroma in the floor of the mouth.

**Case Report.** A 74-year-old Bahraini female presented with a painless swelling in the right side of the floor of the mouth. It was noticed by the patient for the previous 3 months. It did not increase in size. Physical examination showed a firm non-tender submucosal, smooth surface mass, in the right side of the floor of the mouth. It was oval in shape and measuring 30 x 25 x 20 mm (**Figure 1**). The mass was bimanually palpated, and the right submandibular duct draining saliva appeared normal. There were no palpable lymph

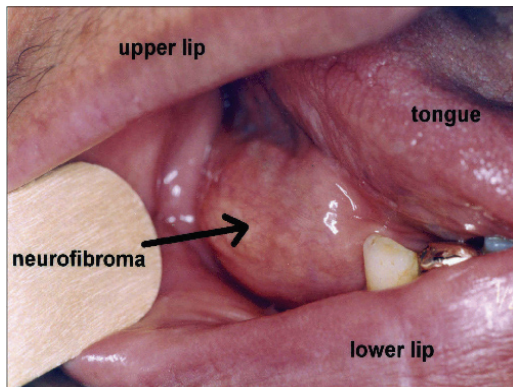
nodes in the neck. The rest of the physical examination and laboratory data were not contributory to the case under discussion. Computed tomography of the oral cavity and upper neck showed a soft tissue mass in the anterior part of the floor of the mouth on the right side with no evident adjacent bone changes. There were a few small lymph nodes in the neck, which were not significant (**Figure 2**). Excision-biopsy of the mass was performed under general anesthesia. At surgery, the lesion was firm, extending from the floor of the mouth to the submandibular space and not adherent to the mandible. The defect was closed primarily with interrupted rapid Vicryl sutures, taking care not to injure the submandibular duct. The post-operative period was uneventful, and she was discharged from the hospital and kept under regular follow-up in the outpatient clinic. The operation site healed very well by primary intention. Macroscopic examination of the specimen revealed a well defined yellowish firm mass, oval in shape and measuring 25 x 20 x 15 mm. Microscopic examination showed a capsulated lesion composed of spindle nucleated cells arranged in tight whorls focally and looser pattern in other areas (**Figures 3a & 3b**), a few of the nuclei were plump and enlarged. There was no mitosis, necrosis, or any foam cells seen within the lesion. Scattered mast cells were present as well. Skeletal muscle

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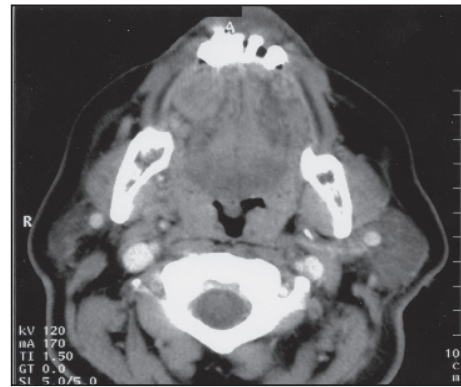
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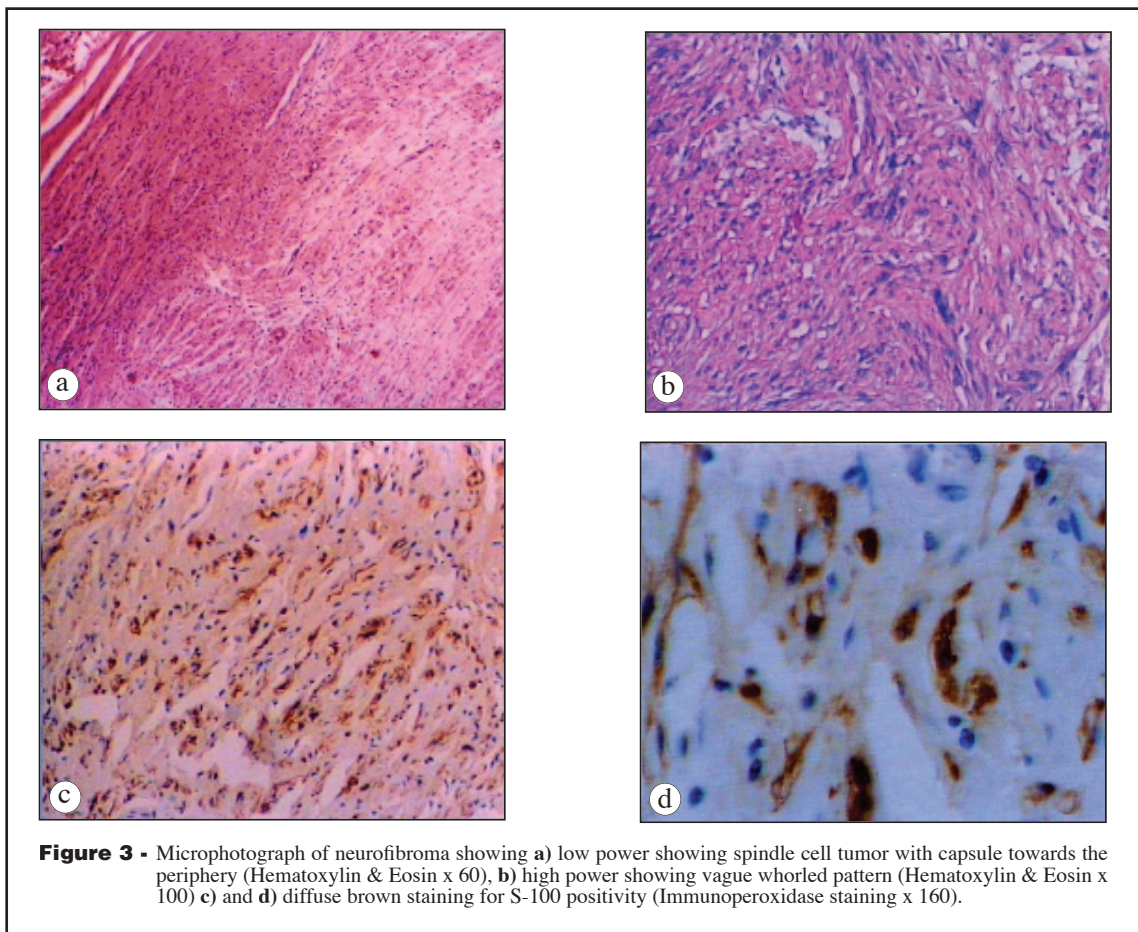
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**Figure 1** - Pre-operative photograph of the tumor arising from the right side of the floor of the mouth.



**Figure 2** - A CT scan of the oral cavity and the neck at the level of the floor of the mouth, showing the tumor localized in the floor of the mouth.



**Figure 3** - Microphotograph of neurofibroma showing a) low power showing spindle cell tumor with capsule towards the periphery (Hematoxylin & Eosin x 60), b) high power showing vague whorled pattern (Hematoxylin & Eosin x 100) c) and d) diffuse brown staining for S-100 positivity (Immunoperoxidase staining x 160).

fibers appear to be compressed toward the periphery. Immunohistochemistry for S100 was positive (**Figure 3c & 3d**) while for smooth muscle actin was negative. Thus, a diagnosis of a neurofibroma was further supported by immunohistochemistry profile.

**Discussion.** We can classify peripheral nerve sheath tumors into benign and malignant. The benign tumors include schwannoma and neurofibroma, and the malignant tumor comprises neurogenic sarcoma. Furthermore, we can divide neurofibroma into solitary tumors and multiple tumors, which include neurofibromatosis type I and type II. We generally think that Schwann cells and perineural cells derived from neuroectoderm are the origin of all these neoplasms.<sup>4,5</sup> Neurofibromas are usually solitary tumors, however, up to a tenth of patients have multiple lesions. They may undergo malignant degeneration, especially when associated with von Recklinghausen's disease, whilst Schwannomas very rarely do so. In the head and neck, malignant transformation ranges between 5-12%, which therefore requires thorough attention and knowledge of the manifestation of neurogenic tumors.<sup>6</sup> Macroscopically, neurofibromas appear as firm, circumscribed, mucosally covered red or grey masses. In the head and neck they are usually painless but may compress surrounding vital structures, and so interferes with phonation, deglutition or respiration; therefore, necessitating early surgical intervention. Although history and examination are helpful, biopsy establishes a definitive diagnosis.<sup>7</sup> Histopathologic features of solitary and multiple neurofibromas are essentially identical. Neurofibromas contain spindle-shaped cells, with fusiform or wavy comma-shaped nuclei distributed on a background of delicate connective tissue matrix. This matrix is rich in mucopolysaccharides and is usually myxomatous. We usually find mast cells scattered within the specimen,<sup>8</sup> as noted in our case. In comparison with a schwannoma, which is a close differential diagnosis, absent Verocay's bodies (Antoni type A areas) and hyaline thickening of the blood vessels in our case, further supports the histologic features of a neurofibroma on light microscopy. We often use immunohistochemistry to aid in confirming the diagnosis made on histology. The tumor's cells are uniformly positive for S-100 protein, signifying that they originate from neural crest-derived tissue. Antibodies to epithelial membrane antigen, CD57, and collagen IV are of secondary value and used only when histologic differentiation with other neural tumors is difficult.<sup>2</sup> Oral neurofibromas usually present as submucosal, nontender, discrete masses that range in size from a few millimeters to several centimeters. The tumors tend to grow slowly, and patients are

usually asymptomatic. The lesion is rarely painful; however, patients may experience pain if there is secondary trauma of lesion due to its location in the oral cavity. Neurofibroma of the floor of the mouth is an extremely rare tumor. We carried out a Medline search for reports of benign tumors, nerve sheath tumors, neurofibromas, or head and neck neoplasms of the floor of the mouth, and in the papers retrieved and on analysis of their references cited, did not find an isolated neurofibroma arising from the floor of the mouth without any other manifestations of Von Recklinghausen's Disease.

The treatment of solitary neurofibroma is complete resection of the tumor, because neurofibromas may infiltrate extensively. Recurrence is rare, although neurofibroma recurs more often than schwannoma. Reports state malignant transformation of neurofibroma to be at the rate of 10%,<sup>9</sup> therefore, long term follow up is essential with careful clinical examination and CT scanning and histological evaluation if suspicion arises. We completely excised this unusual tumor located in the floor of the mouth surgically through an oral incision. Follow up regularly for 3 years failed to demonstrate any recurrence.

In conclusion, we present a rare case of solitary neurofibroma arising in the floor of the mouth of an elderly lady. Immunohistochemistry supports the pathological diagnosis of this case.

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