Case Report

Schwannoma of head-and-neck region: A clinical chameleon – Report of two cases occurring at rare sites with unusual clinical manifestations

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Abstract

Schwannomas are rare benign encapsulated neoplasms that usually do not arise in the nasal and oral cavities. Only about 25% of the schwannomas are located in the head-and-neck region. The preoperative diagnosis of schwannomas in the head-and-neck region is difficult, as they present with varied clinical manifestations and have nonspecific radiological findings, which can lead to a diagnostic dilemma. We report two cases of schwannomas, one in the nasal cavity and the other in the tongue, that were considered to be an infective lesion and malignant lesion respectively on clinical evaluation. The biopsy and subsequent histopathological examination led to the diagnosis of schwannoma. It is important to be aware of the unusual clinical manifestations of schwannomas and keep it in the differential diagnoses, even at relatively uncommon sites such as the nasal and oral cavities. Lesions that are suspected to be infectious lesions or malignant lesions on clinical and/or radiological evaluation can also be schwannomas. Unnecessary diagnostic evaluations and radical therapeutic measures can be avoided with early diagnosis.

Keywords: Head, neck, nose, schwannoma, tongue

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INTRODUCTION

Schwannomas, also known as neurilemmomas, are rare benign encapsulated neoplasms that arise from schwann cells. They are even rarer in the nasal and oral cavities, with only about 25% being located in the head-and-neck region. These lesions can present with varied clinical manifestations, which can lead to a diagnostic dilemma. We report two cases of schwannomas, one in the nasal cavity and the other in the tongue, that were considered to be an infective lesion and malignant lesion, respectively, on clinical evaluation. Since schwannomas almost never undergo malignant transformation and usually do not recur after complete surgical excision, it is important to be aware of the unusual clinical manifestations of this lesion and keep it in the differential diagnoses.

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CASE REPORTS

Case 1

A 32-year-old male presented with a mass in the right nostril for 3 months. It was about 5 cm in maximum dimension and caused deviation of the nasal septum along

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with decrease in the lumen of the nasal cavities, leading to difficulty in breathing. The lesion was suspected to be an inflammatory polyp clinically.

Computed tomography (CT) scan showed the presence of an enhancing mass lesion in the right nasal cavity with few necrotic components, causing bone remodeling changes [Figure 1]. It appeared to arise from the nasal septum, however, no evidence of any bone destruction was present. Anteriorly, the mass lesion was extending into the vestibule; and posteriorly, it was confined within the nasal cavity. A radiological diagnosis of neoplastic lesion was offered, with papilloma (likely fungiform), malignant mass lesion, fungal infection and finally, schwannoma as differential diagnosis. On clinical re-evaluation, a differential diagnosis of rhinosporidiosis was kept.

A biopsy was sent for histopathological examination, which showed cellular spindle cell tumor comprising of hypocellular and hypercellular areas. The tumor cells were ovoid to elongate with fine chromatin, inconspicuous nucleoli and moderate amounts of ill-defined cytoplasm. Several Verocay bodies were noted. No atypical mitosis or necrosis was identified. On immunohistochemistry (IHC), the tumor cells were strongly for S-100 protein [Figure 2]. The lesion was diagnosed as schwannoma. The subsequent excised specimen also showed similar morphological features.

Case 2

A 28-year-old male presented with a nodular lesion on the tongue that was first noticed 1 year ago [Figure 3]. The patient complained of difficulty in chewing, swallowing and phonation. There was no history of weight loss or trauma.

On examination, an ulcerated, nontender, noncompressible, nonreducible swelling was present that involved predominantly the left half of the tongue. It was 3 cm × 2 cm in size and was covered with necrotic slough. The tongue mobility and protrusion were slightly reduced. The adjacent mucosa showed no abnormalities. Cervical lymphadenopathy was present, with palpable lymph nodes on both sides of the neck.

Based on the clinical findings, the possibility of a malignant lesion was considered as the first differential diagnosis.

CT scan revealed a heterogeneously enhancing irregular marginal lesion, involving the anterior tip and left side of the tongue, crossing midline [Figure 4]. No infiltration into the genioglossus-geniohyoid complex, hyoglossus and mylohyoid muscles was seen.

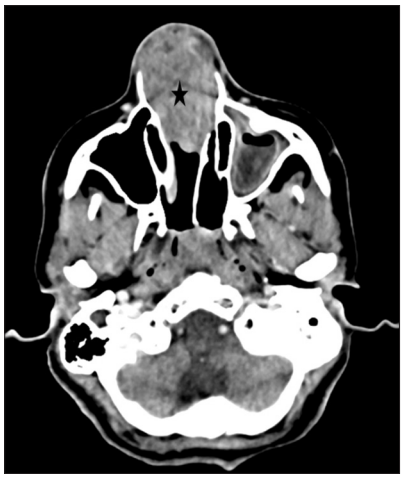

Figure 1: Contrast-enhanced computed tomography (axial section) at the level of nasal cavity showing a heterogeneously enhancing soft-tissue mass lesion (*) distending the right nasal cavity, displacing the nasal septum to the left side

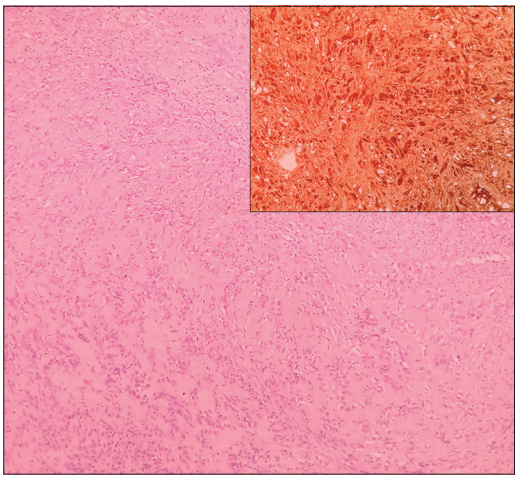

Figure 2: Photomicrograph showing cellular spindle cell tumor comprising of hypocellular and hypercellular areas, with the presence of several Verocay bodies (H and E, ×100). Inset– The tumor cells expressed strong immunopositivity for S100 protein

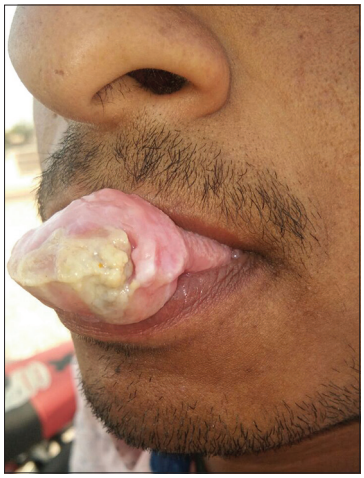

Figure 3: Clinical photograph showing nodular lesion on the tongue

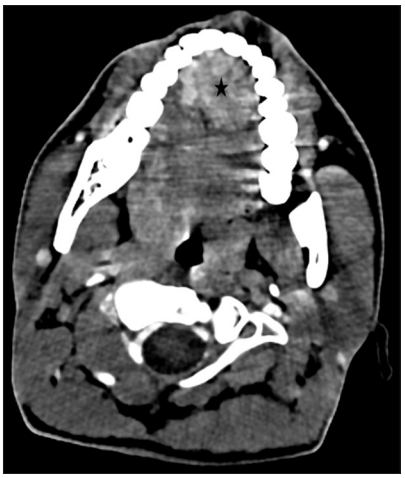

Figure 4: Contrast-enhanced computed tomography (axial section) of the neck showing a heterogeneously enhancing soft-tissue mass lesion (*) at the tip of the tongue. The enhancement of the lesion is more than the tongue muscles

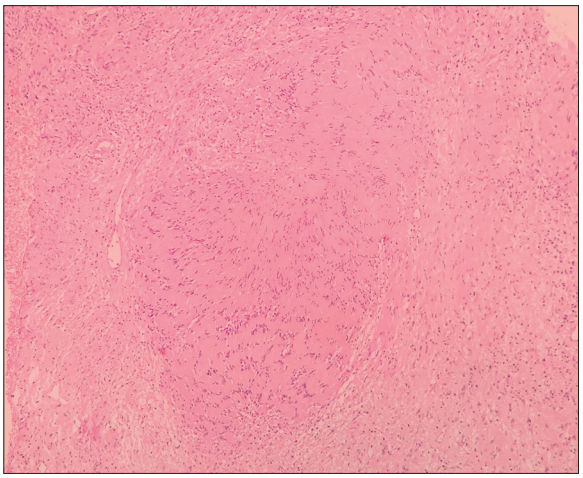

Figure 5: Photomicrograph showing cellular spindle cell tumor, with the presence of Verocay bodies (H and E, ×100)

A biopsy was performed, which revealed a tumor composed of oval to spindle-shaped cells with hypocellular and hypercellular areas. The tumor cells were ovoid to elongate with moderate eosinophilic cytoplasm, fine chromatin and 0–1 nucleoli. Focal sporadic mild atypia was present. Many Verocay bodies were seen [Figure 5]. Separate small necrotic bits with exudate were also noted. A diagnosis of schwannoma was made on histopathological examination.

Direct fine-needle aspiration was done from the right cervical lymph node and showed features of reactive lymphoid hyperplasia.

DISCUSSION

Schwannomas are slow-growing, benign neoplasms that can arise from any nerve that is covered by a sheath of schwann cells. These include cranial nerves (except optic and olfactory nerves, as they are extensions of the white matter of the brain), spinal nerves and nerves of the autonomic nervous system.[1,2]

They were first described by Verocay and the schwannian derivation of these tumors was recognized by Stout.[3]

Schwannomas usually present as single, firm, circumscribed lesions of variable size, that are not associated with pain or ulceration.[1] The average size ranges between 2 and 3 cm.[4] About 25%–40% of all schwannomas occur in the head and neck, with 90% or more arising from the vestibulocochlear nerve.[5] Only 1%–12% of these affect the intraoral area, for which tongue is the most common site. Other intraoral sites include the palate, the floor of the mouth, gingiva, buccal mucosa, lips and vestibular mucosa.[4] Within the tongue, these lesions may arise from the hypoglossal nerve, the glossopharyngeal nerve or the lingual nerve. However, only 50% of the tumors have a direct relation with a nerve.[6]

The preoperative diagnosis of schwannomas in the head-and-neck region is difficult.[7] The clinical presentations of lingual schwannomas vary, depending on the size and location of the lesion. More than 50% of the patients present with an asymptomatic mass. The common complaints in symptomatic cases include throat pain or discomfort, dysphagia, change in voice, snoring or difficulty in breathing. Ulceration and pain are present in <2% of the patients. The symptoms are present more commonly in patients who had lesion in the posterior one-third of the tongue.[1]

The differential diagnosis of these tumors usually includes soft-tissue neoplasms such as fibroma, lipoma and leiomyoma, paraganglioma, carotid artery aneurysm, branchial cleft cyst, angioma and other neurogenic tumors.[7]

However, sometimes the clinical manifestations can be deviant, as was seen in the current cases, which can lead to diagnostic dilemmas.

Though radiology is helpful in reaching the most probable diagnosis, as seen in the current cases, radiology might not always provide with a definite diagnosis, and therefore, biopsy becomes mandatory in such cases.

The histopathological appearance of the lesion is characteristic, comprising of two patterns of tissue growth – Antoni type A and B areas. Type A areas are densely packed, composed of elongated schwann cells

whereas type B areas have less number of cells that are dispersed in a loose, myxoid and disorganized matrix. Variations in the distribution of type A and B areas have no clinical significance.[2,8] S-100 protein is the most sensitive IHC marker for these lesions.[9]

It should be emphasized that differential diagnosis of schwannoma should be kept in mind for mass lesions, even at relatively uncommon sites such as the nasal and oral cavities. Lesions that are suspected to be infectious lesions or malignant lesions on clinical and/or radiological evaluation can also be schwannomas, which should, therefore, be considered in the differential diagnosis. This is necessary to avoid unnecessary diagnostic evaluations and radical therapeutic measures and would lead to better patient care.

CONCLUSION

Schwannomas are rare tumors of the nasal and oral cavities, which can present with unusual clinical manifestations, ranging from infectious lesions to malignancies. Therefore, they should be considered in the differential diagnosis while evaluation of any case of nasal or oral cavity mass, since the prognosis is good and favorable outcome likely, and unnecessary diagnostic evaluations and radical therapeutic measures are not required, leading to better patient care. Complete resection is sufficient, following which recurrence is rare.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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