Schwannoma of Left Infra Orbital Region – A Case Report With Emphasis On Advanced Imaging In Diagnosis

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ABSTRACT: Schwannomas are uncommon neurogenic tumors that are typically benign, slow growing, and asymptomatic. The preoperative imaging provides us valuable information on location and origin of the tumor aiding in near accurate preoperative diagnosis. In rare cases such as schwannoma, it is critical to determine the origin of the tumor to preserve nerve function which is done with the help of the Magnetic resonance imaging modality.

Keywords: schwannoma, peripheral nerve sheath tumor, Imaging of schwannoma

Introduction:
The head and neck schwannomas are fairly common benign tumor of the Schwann cells. Around 3-4% of these cases are found during autopsy. The Central Brain Tumor Registry of the United States (CBTRUS) USA has reported an incidence rate for all primary nerve sheath tumors of the brain/CNS of 0.8 per 100,000 person/year for the years 1990–1994 and 1.1 per 100,000 person/year for the years 1995–1999. Among Indian patients with vestibular schwannoma, 56 per cent had giant tumors and another 41 per cent had tumors measuring 25 to 40 mm in size. Once termed “neurilemoma” (Stout, 1935), this most common variant of schwannoma, presents as a globoid, encapsulated tumor associated with a spinal or cranial sensory nerves. This occurs most commonly in sixth decade of life. Schwannoma involves the nasal cavity and paranasal sinuses and represents less than 4% of soft tissue tumors of head and neck region occurring commonly in middle aged adults with equal gender distribution. The cases are of interest due to the relative rarity of the pathology and presence of non-significant symptoms for a presumed initial clinical diagnosis.

CASE REPORT:
A 46 year old female patient (fig.1) reported with a chief complaint of pain in left upper back tooth region since 15 days. Patient was known hypertensive and diabetic since 5 years and under medication. On general examination patient was normal. Extra oral examination revealed a small oval shaped swelling of size 2*2 cm on the left infra orbital region which was of same size since the inception to the present day, skin over the swelling appeared normal. Surrounding structures appeared normal with no secondary changes. On palpation all inspectory findings with respect to size, shape and extent were confirmed the swelling was soft to firm in consistency, non-fluctuant and non-tender. Intra oral examination revealed no significant findings. With the above clinical findings a provisional diagnosis of Lipoma on left infra orbital region was made and since it was a soft tissue mass the patient was subjected to MRI. MRI revealed a sharply defined ovoid mass, homogeneously isointense to the muscle on both T1&T2w sequences located deep to temporalis muscle, superiorly up to the supra orbital margin inferiorly up to hard palate. The sharply demarcated homogenous soft tissue mass arising from the left pterygopalatine fossa and deep to temporalis muscle was suggestive of Peripheral Nerve sheath tumor most likely schwannoma. On correlating the clinical and radiologic findings a final diagnosis of schwannoma was given. (fig2 & 3)

DISCUSSION:
Oral schwannomas are rare and present most commonly in patients with 30-60 years old with a slight female predisposition in few studies, no sex predilection in few studies. Intra orally tongue is the most commonly affected site, base of the tongue.
Clinically pain, tenderness, and paresthesia may be expected if the tumor is large or, by virtue of a deep-seated location, is impinging on neighboring structures. Symptoms have been reported in up to one third of patients. Neurolemmomatosis or schwannomatosis, a variant of NF2, is an autosomal dominant disorder with full penetrance. The spectrum of schwannomas which include conventional schwannoma, a histologically benign tumor which destroys the surrounding osseous structures, cellular schwannoma, a tumor that histologically simulates malignant peripheral nerve sheath tumor (MPNST) plexiform schwannoma which, particularly in cellular form and when occurring in childhood, simulates MPNST and lastly Melanotic schwannoma a variety which is often mistaken for melanoma. The psammomatous form of the latter is often associated with Carney complex, a rare heritable disorder that includes cutaneous lentiginous, myxomas of skin, subcutaneous tissue, heart and endocrine neoplasms.(3)

MACROSCOPIC FEATURES OF SCHWANNOMA

Schwannoma (conventional schwannoma) in its baseline form include globoid shape, presence of a capsule, tan color and homogeneously firm texture.

RADIOGRAPHIC FEATURES

CONVENTIONAL IMAGING

Conventional imaging modalities does not give a significant description of the soft tissue masses. These soft tissue masses are better seen with the high resolution magnetic resonance imaging on comparison to a conventional imaging.

ADVANCED IMAGING

Schwannomas appear as enhancing homogenous or heterogeneous well circumscribed soft tissue density mass on a computed tomography. The lesions margin may not be seen on computed tomography. Schwannomas are well-circumscribed lesions. On magnetic resonance imaging appear isointense or hyper intense to muscle on T1-weighted imaging and hyper intense on T2-weighted imaging. This gives the appearance of a target lesion. A Cystic schwannoma on magnetic resonance imaging will appear isointense on T1-weighted imaging, hyper intense on T2-weighted imaging, and will often feature an area of peripheral enhancement after fat suppression.(5). MRI orbital schwannomas appear a sharply marginated, oval or fusiform intracanal or extracanal masses that demonstrate moderate to marked enhancement. The optic nerve is always displaced and often engulfed by the tumor. The lesions in jaw may appear as solitary radiolucency associated with the inferior alveolar nerve canal or as multilocular radiolucency’s that produce extensive bone damage with cortical remodeling. A schwannoma with in the nerve (inferior alveolar canal) may cause bulbous enlargement of the canal.(6). Overall MR imaging enables better delineation of schwannomas because of superior soft tissue contrast.

CONCLUSION

It is evident that dental diagnosis does not confine itself to the oral cavity but also the head and neck region as a whole. This case implies the significance of ideal investigations necessary for the condition there by helping in appropriate diagnosis and management

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