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Solitary Neurofibroma of Maxillary Sinus: A Rare Clinical Entity

Dr. Lakshmi Krishnan[1], Dr. Anannya Tripathy[2], Dr. Manu S Babu[3], Dr. Akshita Goyal[4]

[1]Junior Resident, Department of ENT and Head and Neck Surgery,
Dr. D.Y. Patil Medical College, Hospital and Research Centre,
Dr.D.Y.Patil Vidyapeeth,Pimpri chinchwad ,Pune,Maharashtra-411018
[2]Junior Resident, Department of ENT and Head and Neck Surgery,
Dr. D.Y. Patil Medical College, Hospital and Research Centre,
Dr.D.Y.Patil Vidyapeeth,Pimpri chinchwad ,Pune,Maharashtra-411018
[3] Associate Professor, Department of ENT and Head and Neck Surgery,
Dr. D.Y. Patil Medical College, Hospital and Research Centre,
Dr.D.Y.Patil Vidyapeeth,Pimpri chinchwad ,Pune,Maharashtra-411018.
[4]Junior Resident, Department of ENT and Head and Neck Surgery,
Dr. D.Y. Patil Medical College, Hospital and Research Centre,
Dr.D.Y.Patil Vidyapeeth,Pimpri chinchwad ,Pune,Maharashtra-411018

CORRESPONDING AUTHOR:

Dr. Anannya Tripathy

Junior Resident, Department of ENT and Head and Neck Surgery,
Dr. D.Y. Patil Medical College, Hospital and Research Centre,
Dr.D.Y.Patil Vidyapeeth,Pimpri chinchwad ,Pune,Maharashtra-411018

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ABSTRACT:

Neurofibromas are slow growing, benign tumours of peripheral nerve tissue seen frequently in association with neurofibromatosis type 1. Although the third decade of life is when presentations are at their peak, cases have been documented in people as young as 10 months to as old as age of 70. The true nature of the disease is often revealed by histopathological evaluation. The isolated occurrence of neurofibroma in maxillary sinus is rare with less than 10 cases reported in literature. Here, we report a rare case of solitary neurofibroma of left maxillary sinus without generalised syndrome of neurofibromatosis in a 27-year-old female patient.

KEYWORDS: Neurofibroma, Maxillary sinus, Head and neck cancer

INTRODUCTION:

A benign, heterogeneous, slow growing tumour termed as neurofibroma most commonly originates in endoneurium of peripheral nerve sheath. (1) Although isolated cases may sometimes occur, they are mostly linked to neurofibromatosis type 1 also known as von Recklinghausen's disease. One of the most common neurogenic tumours is neurofibroma.

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Although the third decade of life is when presentations are at their peak, cases have been documented in people as young as 10 months to as old as age of 70. The true nature of the disease is often revealed by histopathological evaluation. They have been broadly classified histologically into solitary, diffuse and plexiform. The last four cranial nerves, the facial nerve and the cervical sympathetic chain are the most often affected nerves in neurofibroma of head and neck region. [(2)] Maxillary sinus solitary neurofibroma is an exceptionally rare condition. [(3)] They often present as painless progressive slow growing sessile or pedunculated lesions but may at times be painful due to pressure effects. [(2,3,4,5)] Here, we report a unique case of solitary neurofibroma of left maxillary sinus in a 27-year-old female patient and try to describe the clinical, histopathological and radiological characteristics of this tumour.

CASE REPORT:

A 27-year-old female patient presented to the ENT OPD with complaints of left sided facial pain and headache for a duration of 6 months relieved only temporarily with medical management to begin again. She had no complaints of trauma to nose, fever, recurrent URTI, allergic symptoms, facial swelling, epistaxis, nasal obstruction, reduced sensations over face, any complaints of loosening of teeth or any restriction of eye movements. She had no significant past or family history.

ON CLINICAL EXAMINATION: Patient was conscious and well oriented to time, place and person. Systemic examination was normal. Sensory examination was normal along all divisions of trigeminal nerve. No obvious facial swelling was seen and skin over the face was normal.

Nose: Bilateral nasal mucosa was normal. Deviation of nasal septum to right with cartilaginous spur on the right side and left frontal and maxillary sinus tenderness were observed. No mass or discharge was seen in bilateral nasal cavities. Anterior rhinoscopy findings were confirmed by doing a diagnostic nasal endoscopy.

Oral cavity: Examination was within normal limits except for caries in left upper 2^{nd} premolar and 1^{st} molar tooth.

Oropharyngeal examination was normal and no lymph nodes were palpable.

Ear: Bilateral tympanic membrane was intact and hearing was normal.

Ophthalmological examination was within normal limits in bilateral eyes.

X-RAY OF THE PARANASAL SINUSES (WATER'S VIEW) showed a radiopaque shadow (homogeneous opacity) in left maxillary sinus.

MRI BRAIN WITH MR VENOGRAPHY(PLAIN) showed expansion of left maxillary sinus with T1-hyperintense and T2-Hypointense content within the left maxillary sinus s/o? mucocele.

COMPUTERIZED TOMOGRAPHY OF THE PARANASAL SINUSES revealed a unilocular and expansile iso-dense lesion of 2.7*3.2*3.5cm in left maxillary sinus nearly occluding it and causing thinning of posterolateral wall of left maxillary sinus. Inferiorly, it

was seen to be related to roots of left upper 2nd premolar and 1st and 2nd molars- s/o? periapical cyst. Superiorly, it was seen to cause superior bowing of floor of left orbit. Rest of the paranasal sinuses were within normal limits.

Making the preoperative diagnosis of left maxillary cyst, surgical excision of the lesion was done by Caldwell Luc approach under GA. Caldwell-Luc exposure of the right maxillary sinus revealed a cystic mass with a thick wall in left maxillary sinus filled with pus. Pus aspirated from the cystic lesion was sent for culture and sensitivity. Cyst wall was removed in toto and sent for histopathological examination. Left maxillary antrum was inspected using 0^0 Hopkin's scope. Left upper 1st molar tooth was extracted and the surgical wound was closed using 3-0 vicryl.

HISTOPATHOLOGICAL EXAMINATION OF INTRA-OP SAMPLE showed pseudostratified ciliated columnar epithelium with sub-epithelium showing a tumour comprised of spindle cells arranged in vague fascicles, with elongated wavy nuclei and pointed ends. Diagnosis: Benign spindle cell neoplasm-neurofibroma.

Postoperatively, the patient had mild facial oedema on the left side of the face which subsided over a weeks' time. Patient is being followed up on OPD basis and follow up done at 2 months post-surgery was normal.

DISCUSSION:

Neurofibroma is a benign nerve sheath tumour arising from the peripheral nervous system. Solitary and multiple neurofibromas are seen to occur in patients with neurofibromatosis type 1, such as von Recklinghausen's disease. Because they lack a Schwann cell sheath, they can develop on any cranial nerve, with the exception of the optic nerve. Nasal and paranasal neurofibromas originate from the ophthalmic & maxillary divisions of trigeminal nerve and autonomic plexus. (6) Before they can be clinically diagnosed, maxillary sinus nerve sheath tumours reach a certain size. The exact location and extent of the lesion greatly determine the symptoms, which are frequently nonspecific. (7) Tumours of the trigeminal nerve frequently do not cause any neurological impairment. However, one common feature of these lesions is bone destruction. Pre-operative CT scan done shows heterogeneous soft tissue density and destruction of paranasal sinuses. Microscopically, they are composed of Schwann cells, perineural cells, mast cells, fibroblasts & axons embedded in an extended extracellular matrix. (1) Neurofibromas are characterised by immunoreactivity for S-100, vimentin and neuron specific enolase. (2) Malignant peripheral nerve sheath tumours and schwannomas must be distinguished from neurofibromas. In contrast to neurofibromas that display sparse tumour cells with a mucoid extracellular matrix, schwannomas exhibit unique areas that are classified as Antoni types A and B, a higher tumour density & typical palisade pattern of nuclei. (8) Pleomorphic tumour cells and nuclei, and hypercellularity are characteristics of malignant peripheral nerve sheath tumours; these features are not present in this tumour. Complete surgical excision remains the gold standard of treatment. (9) The infiltrative nature of the disease may make total surgical excision challenging for the surgeon. These procedures may occasionally necessitate the sacrifice of vital structures, which can result in a significant functional deficit or cosmetic defect that raises the disease's morbidity and mortality. (10)

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Despite the fact that intralesional excision would decrease morbidity, it typically results in recurrence. Because lesions may recur, follow-up is required. Rarely can solitary neurofibromas undergo malignant change. In 2-5% of cases of neurofibromatosis type-1, neurofibroma can develop into a malignant peripheral nerve sheath tumour. [(1)]

CONCLUSION:

Otorhinolaryngologists should keep this diagnosis in mind within the range of paranasal sinus tumours despite the disease's rarity and difficulty in diagnosis. Because the single tumour may be the beginning of neurofibromatosis type 1, it is critical to pay attention to it.

REFERENCES:

1. Ferner RE, O' Doherty MJ. Neurofibroma and schwanomma.Curr Opin Neurol 2002;15:679-84.
2. LAE. Pereira, YB. Choo, “Schwanomma of the infraorbital nerve,” Ear, Nose and Throat Journal, Vol. 58, 1979, pp. 236-239
3. M. Hirao, T. Gushiken, H. Imokawa, S. Kawai, H. Inaba, M. Tsukuda, “Solitary neurofibroma of the nasal cavity: resection with endoscopic surgery,” J Laryngol Otol, Vol. 115, No. 12, 2002, pp. 1012-1014.
4. RJ. Poupard, S. Mintz, “Solitary intrabony neurofibroma of the maxilla,” J Oral Maxillofac Surg, Vol. 55, No. 7, 1997, pp. 768-772.
5. CC. Boedeker, GJ. Ridder, G. Kayser, J. Schipper, W. Maier, “Solitary Neurofibroma of the Maxillary Sinus and Pterygopalatine Fossa,” Otolaryngol Head Neck Surg, Vol. 133, No. 3, 2005, pp. 458-459.
6. CB. Majoie, FJ. Hulsmans, JA. Castelijns, LH. Sie, A. Walter, J. Valk, KW. Albrecht, “Primary nerve-sheath tumours of the trigeminal nerve: clinical and MRI findings,” Neuroradiology, Vol. 41, No. 2, 1999, pp. 100-108.
7. Gómez-Oliveira G, Fernández-Alba Luengo J, Martín-Sastre R, Patiño-Seijas B, López-Cedrún-Cembranos JL. Plexiform neurofibroma of the cheek mucosa. A case report. Med Oral. 2004;9:263.
8. Hillstrom RP, Zarbo RJ, Jacobs JR. Nerve sheath tumors of the paranasal sinuses: Electron microscopy and histopathologic diagnosis. Otolaryngol Head Neck Surg 1990;102:257-63.
9. Deka, et al. Neurofibroma of the maxillary sinus. Indian journal of Otolaryngology 1988;40:28-29.
10. Meyer U, Kleinheinz J, Handschel J, Kruse-Lösler B, Weingart D, Joos U. Oral findings in three different groups of immunocompromised patients. J Oral Pathol Med. 2000;29:153–8.

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Figure 1: Preoperative picture showing no obvious facial swelling

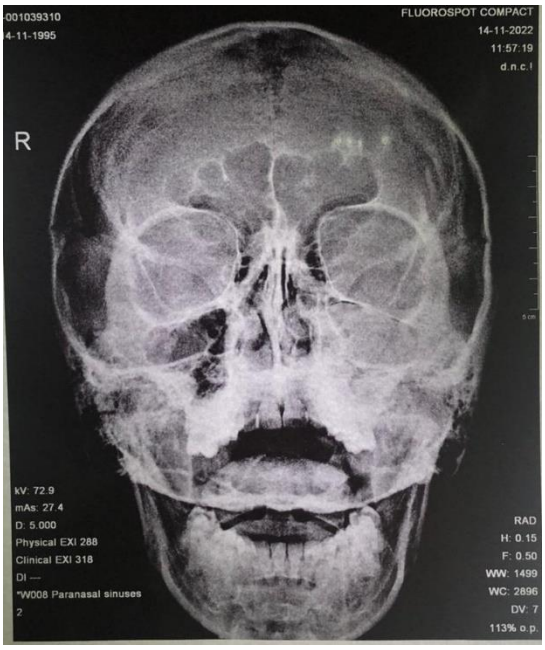

Figure 2: X-Ray Water's view showing homogenous opacity of left maxillary sinus.

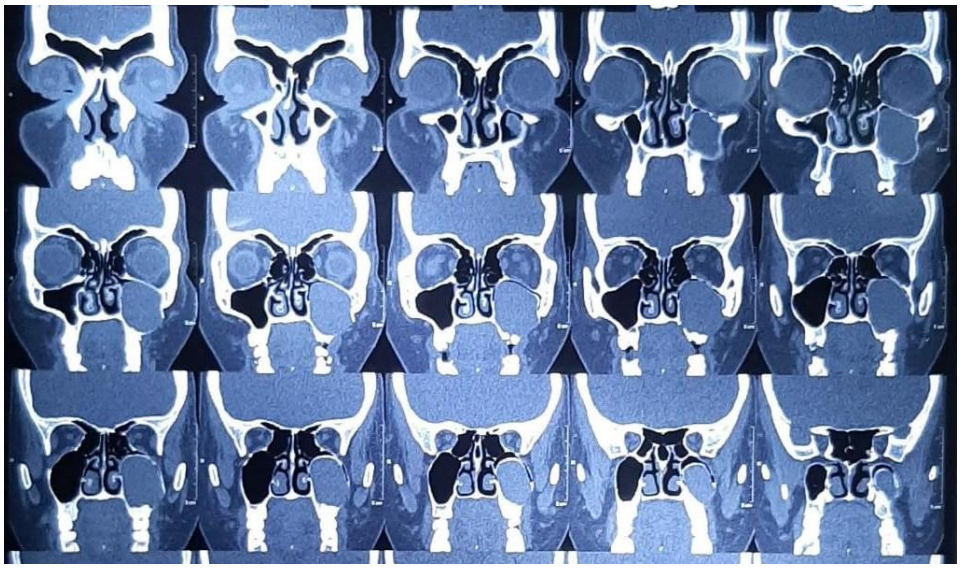

Figure 3: NCCT-PNS showing expansile lesion involving left maxillary sinus.

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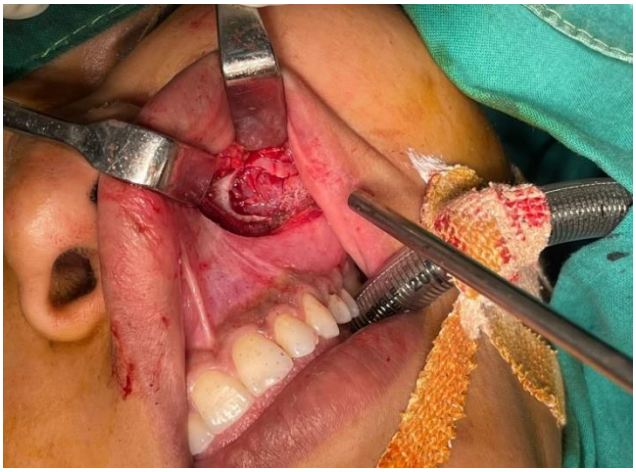

Figure 4: Intraoperative picture showing cystic lesion in left maxillary sinus.

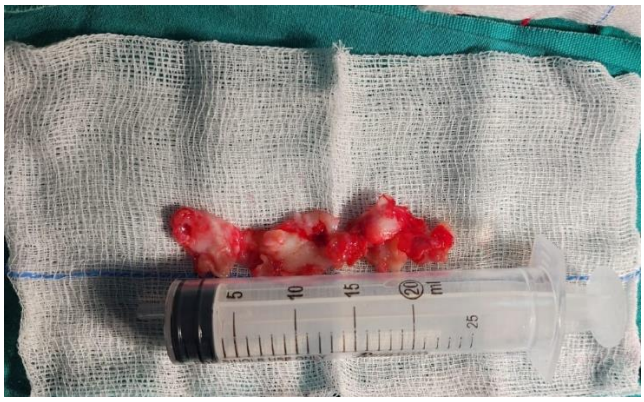

Figure 5: Specimen excised and sent for histopathological examination.