

SOLITARY NEUROFIBROMA OF THE MAXILLARY SINUS: A RARE CASE REPORT

C. Adnane¹, T. Adouly², L. Elhani³, M. Mahtar⁴

1, 2,3,- Resident, 4- Prof & Chief surgeon, Dept. of ENT, 20 Aout Hospital, IBN Rochd
University hospital, Casablanca, Morocco

ABSTRACT:

Neurofibroma is a benign slow growing tumour of the peripheral nerve sheath; frequently associated with neurofibromatosis type I. Solitary neurofibroma of maxillary sinus is exceedingly rare tumour, with only seven cases described in the literature. We report a case of a 14-year-old male patient presented in our ENT department with a painless swelling of the left cheek for the last six months. The diagnosis was a solitary neurofibroma of the maxillary sinus and we try to describe the clinical, histopathological and radiological characteristics of this tumour. CT scan showed a heterogeneous expansive lesion in the left maxillary sinus destroying the lateral, medial, anterior and posterior walls with infiltration of the fosse infratemporalis. A maxillary resection was performed to excise the tumour. Histological examination of the specimen showed a neurofibroma. No sign of recurrence was noted after 6 months of follow-up. Despite the rareness and the difficulty diagnosing this disease, otolaryngologists should keep this diagnosis in mind within the range of tumours of the paranasal sinuses.

KEYWORDS: Neurofibroma; Maxillary sinus.

Cite this Article: C. Adnane, T. Adouly, L. Elhani, M. Mahtar. Solitary Neurofibroma of maxillary sinus- A rare case report: Journal of Head & Neck physicians and surgeons Vol 2, Issue 2, 2014 : Pg 136-141

INTRODUCTION:

Neurofibroma is a benign slow growing and encapsulated tumour of the peripheral nerve sheath. It arises from the connective tissue of their sheaths, especially the endoneurium¹. It may occur as isolated sporadic lesion, but is much more common in association with neurofibromatosis type 1, also known as von Recklinghausen's disease. Solitary neurofibroma of the maxillary sinus is exceedingly rare tumour^{2,3,4}. We report a case of solitary neurofibroma of the maxillary sinus and we try to describe the clinical, histopathological and radiological characteristics of this tumour.

CASE REPORT:

A 14-year-old male patient presented in our ENT department with a painless swelling of the left cheek for the last six months (figure 1). History revealed a postnasal discharge and left unilateral nasal obstruction for the last for years. There was no history of epistaxis or headache. The examination of the facial region showed a painless bulge of the left cheek with a healthy skin. It was firm in consistency. Endobuccal examination revealed a tumefaction of the left upper gum with palatal and vestibular extension. Ophthalmic examination found a diplopia and a left exophthalmia without decreased visual acuity. Endoscopic examination of the nose revealed mucopurulent discharge and tumefaction in the left nasal cavity. Anatomical landmarks of the left nasal cavity were changed and the tumefaction had a diffuse margin with normal nasal mucosa. The CT scan showed a heterogeneous expansive lesion in the left maxillary sinus destroying the lateral, medial, anterior and posterior walls with infiltration of the fosse infratemporalis (figure 2). Multiple biopsies with histopathological examination were no specific with no signs of malignancy. The tumour excision was the final decision. Exposure of the left maxillary sinus by Caldwell-Luc approach revealed a white colored thick and firm mass filling the maxillary sinus with marginal attachment especially in the inferior wall. The mass was removed in totality with a partial maxillectomy (figure 3). Reconstruction was made by buccal fat pad flap (figure 4). Final histopathological examination found a neurofibroma. There was no sign of recurrence after 8 months of follow-up.



Figure 1: Image of patient objective a swelling of the left cheek.

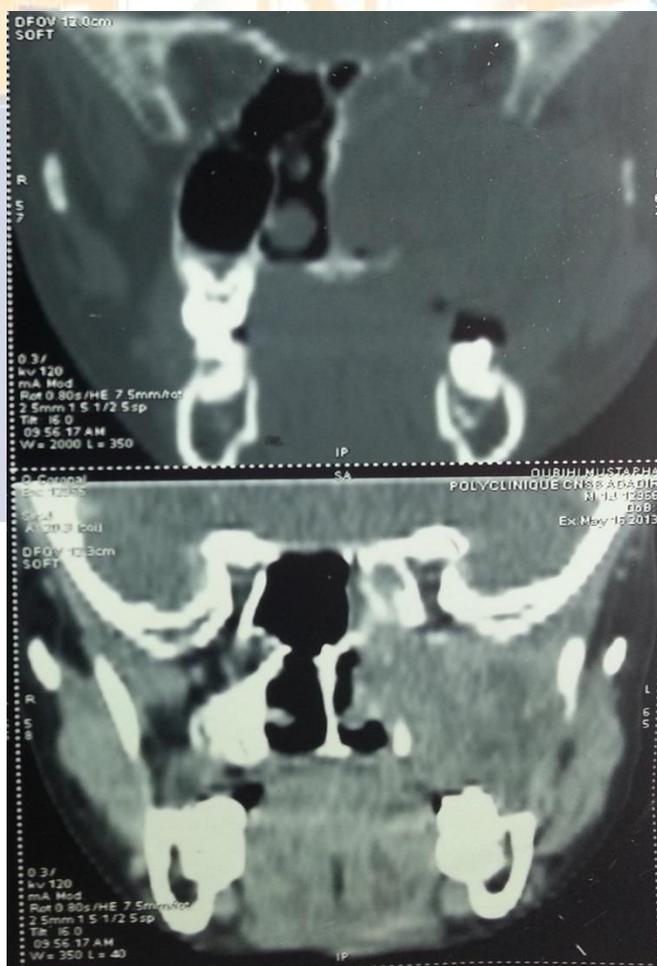


Figure 2: Coronal CT scan showed a heterogeneous expansive lesion in the left maxillary sinus.



Figure 3: Macroscopic view of the removed mass.

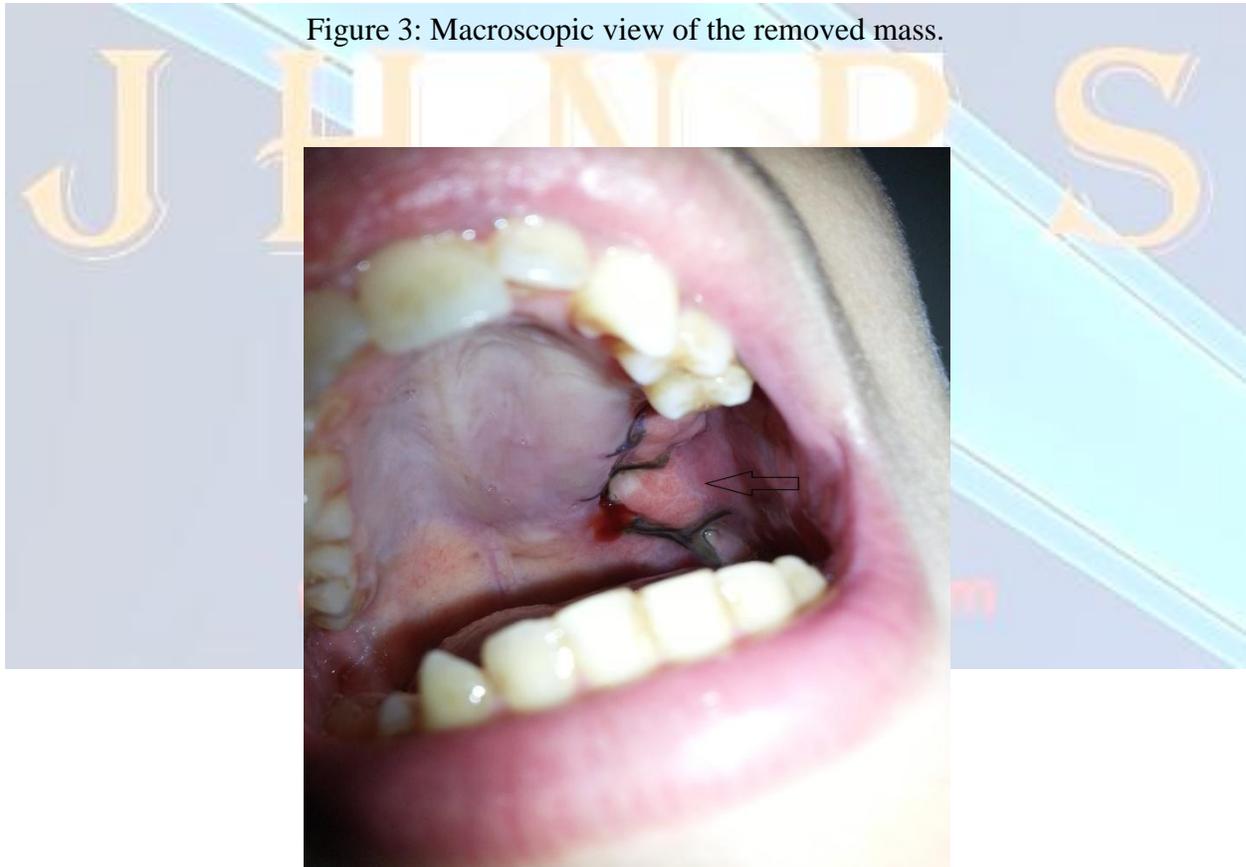


Figure 4: Post-operative view after buccal fat pad flap reconstruction.

DISCUSSION:

Neurofibroma is a benign nerve sheath tumour arises from the peripheral nervous system. It is divided into solitary and multiple. The last one occurs more frequently especially in association with neurofibromatosis type 1, such as Von Recklinghausen's disease. Neurofibromas may arise on any cranial nerve except the optic nerve as they do not possess Schwann cell sheath. Nasal and paranasal neurofibromas arise from the ophthalmic and maxillary division of the trigeminal nerve⁵. The symptoms are non-specific and depend greatly on the exact location and extent of the lesion⁶. In this location, it is often clinically silent until reaching considerable size before diagnosis⁴. Trigeminal nerve tumours often produce no neurological deficit and bone destruction is a common feature of these lesions. CT scan show heterogeneous soft tissue density and destruction of paranasal sinuses. Microscopically, they are composed of Schwann cells, fibroblasts, perineural cells, axons and mast cells¹. Immunoreactivity for S-100 protein and vimentin is characteristic for neurofibromas⁵. Complete surgical resection is the treatment of choice^{7,8}. The transformation of neurofibroma into malignant tumour has been observed in 2 to 5% of cases of neurofibromatosis type 1¹. Follow-up is essential because the lesion may recur and very rarely undergo malignant change. Isolated occurrence in a maxillary sinus is very exceptional, with only seven cases described in the literature⁹.

CONCLUSION :

Despite the rareness and the difficulty diagnosing this disease, otolaryngologists should keep this diagnosis in mind within the range of tumours of the paranasal sinuses. A prolonged monitoring is mandatory to avoid recurrence and malignant transformation.

REFERENCES:

- [1] RE. Ferner, MJ. O'Doherty, "Neurofibroma and schwannoma," *Curr Opin Neurol*, Vol. 15, No. 6, 2002, pp. 679-684.
- [2] RJ. Poupard, S. Mintz, "Solitary intrabony neurofibroma of the maxilla," *J Oral Maxillofac Surg*, Vol. 55, No. 7, 1997, pp. 768-772.

[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] 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.

[5] LAE. Pereira, YB. Choo, "Schwanomma of the infraorbital nerve," Ear, Nose and Throat Journal, Vol. 58, 1979, pp. 236-239.

[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] DA. Tandon, RC. Deka, "Neurofibroma of the maxillary sinus," Indian journal of Otolaryngology, Vol. 40, No. 1, pp. 28-29.

[8] RN. Hachem, A. Bared, J. Zeitouni, RT. Younis, "Single-stage total endoscopic resection of a plexiform neurofibroma of the maxillary sinus in a child with type 1 neurofibromatosis," Int J Pediatr Otorhinolaryngol, Vol. 74, No. 4, 2010, pp. 426-429.

[9] FCN. Maria, ADM. Jose, VR. Enrique, IA. Jesus, "Solitary Neurofibroma of the Maxillary Sinus," Acta Otorrinolaringol Esp, Vol. 62, No. 5, 2011, pp. 395-397.

Acknowledgement- None

Conflict of Interest- None Declared

Ethical Approval- Not Required

Correspondence Addresses :

Choaib ADNANE:
Resident, department of ENT, 20 août hospital,
Ibn Rochd university hospital,
Casablanca, Morocco.
Email: adnanechoaib@gmail.com