SVOA Dentistry

ISSN: 2753-9172

ScienceVolks Open Access

Case Report

Schwannoma (Neurilemmoma). Ectopic Position in Maxillary Sinus

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DOI: https://doi.org/10.58624/SVOADE.2024.05.0179

***Received:** June 21, 2024* ***Published:** July 09, 2024*

Abstract

A Schwannoma (Neurilemmoma) is a benign nerve tumor arising from a myelinated nerve fibers sheath, that can appear in any part of the body. Forty-five percent of the cases occur in the head and neck region. Intraoral Schwannomas are rather uncommon, intranasal and intrasinus Schwannomas are even rarer (less than 1%). The clinical case reports a young, man 14-years-old male with a Neurilemmoma the left maxillary sinus, involving the left third molar. The reports include case information, radiological findings, the surgery treatment, the histological characteristics of this particular tumor, a differential diagnosis, the post-operation process, and a review of the available literature.

Keywords: *Neurilemmoma, Benign Neurogenic Tumor, Ectopic Position, Schwannoma of the Maxillary Sinus, Third Molar.*

Introduction

A Neurilemmoma or Schwannoma is a benign neoplasia derived from Schwann cells – a lesion first described by Verocay in 1910. Back then, he called it a "Neurinoma". In 1935, Stout coined the term "Neurilemmoma"[7].

A Schwannoma is a benign neoplasm derived from the neuroectoderm, produced by unknown causes or stimuli. It develops from the Schwann cells (originating in the neural crest) of the peripheral nerve sheath. As the Schwannoma grows, the nerve is displaced and is not infiltrated by the tumor [2,3,4,5]. The lesion is painless, slow-growing, encapsulated, can appear and grow at any age, and is most commonly located in the soft tissues of the head and neck.[2,4,5,6] It usually affects sensitive nerves.[15,16] An important characteristic of the Schwannomais that its malignancy rate is very low, whereas this rate is relatively high for Neurofibromas (Von Recklinghausen Skin Disease)

The second most common Schwannomas appear on the trigeminal nerve, accounting for 0.8 to 8% of these intracranial neoplasms[1,8,9,10,11].

Trigeminal Schwannomas tend to affect middle-aged patients, mainly those from 38 to 40 years in age, with cases ranging from ages 30 to 60 [16,17,18]. The incidence in women is twice that of men [1,12,13,14,15,16,17].

Trigeminal Schwannomas (TS) patients sometimes experience mild symptoms in the areas served by nerves from the affected trigeminal branch; however, they can also be asymptomatic.[1] The highest incidences are between the ages of 38 and 40 years, and are more common in women [1,12,13,14].

Case Presentation

A 14-years-old male patient residing in of Santiago, Chile, exhibitingDysphasic Syndrome - with mentaldeficiency and disability- caused by hypoxia during fetal development.The patient had not been treated forthese pathologies. Possible epilepsy, untreated. In addition, the patient had been defined as acarrier of Ligamentous Hyperlaxity.

During a routine dental check-up, the patient had apanoramic x-ray exam (Fig. 1). The exam showed a radio-opaque lesion in the left maxillary sinus cavity. As result,a High-Resolution Computed Tomography (HRCT) Scan was requested for a better view and understanding, to thereby determine a course of treatment. (Fig. 2) HRCT of Neurilemmoma associated with the upper- left third molar. In the image, a radio-opaque lesion can be observed in the left maxillary sinus that displaces the upper-left third molar relative to its crown.

Tooth in formation, classified as dental follicle located close to the nasal and posterior wall of the maxillary sinus. The lesion surrounding this molar reaches to the inside of the maxillary sinus walls. Apparently, it does not involve this sinus mucosa. (Fig. 3)

The diagnostic hypotheses proposed were: Maxillary Cyst, Dentigerous Cyst; and Keratocyst.

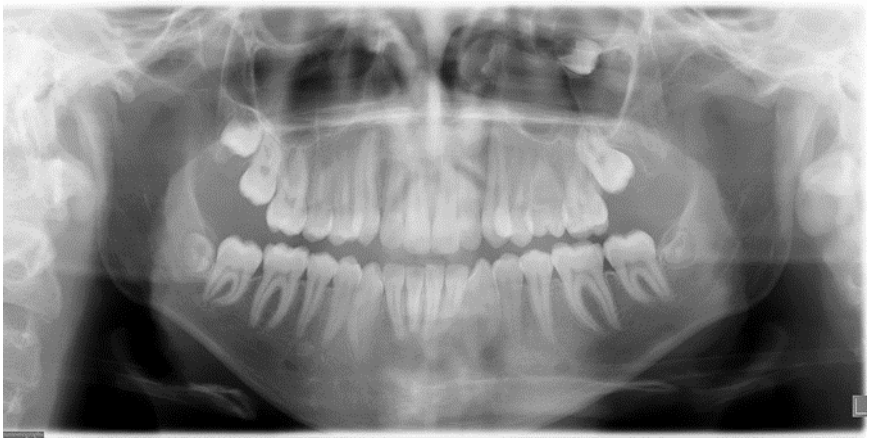

Figure. 1 *Patient initial Panoramic x-ray.*

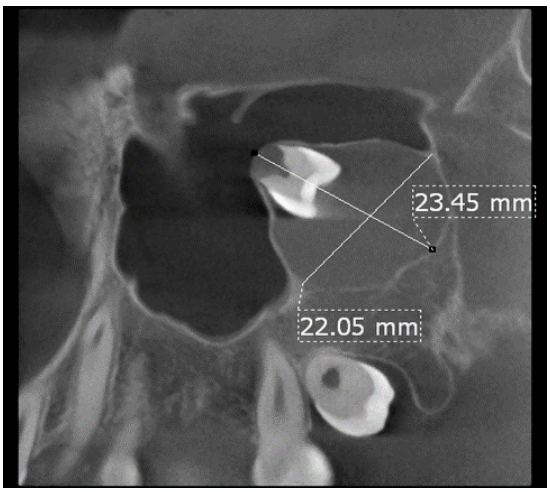

Figure. 2 *CT Cone-Beam Neurilemmoma associated with and displacing the upper left third molar.*

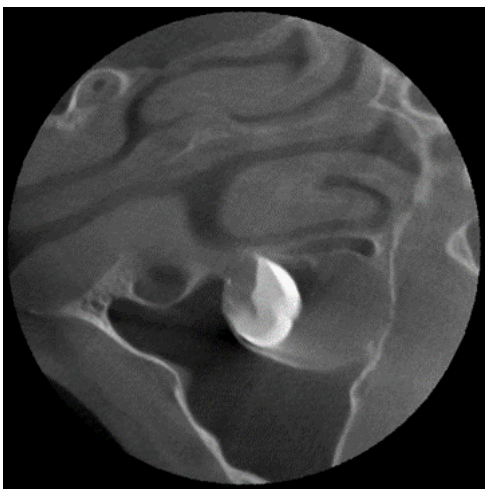

Figure. 3 *CT Cone-Beam axial view, Neurilemmoma surrounding molar and reaching maxillary sinus walls without involving sinus mucosa*.

Through complementary tests, blood exams and imaging scans, along with a clinical exam and the patient's asymptomatic characteristics, a clinical imaging diagnosis was made of Intrasinus Cyst Lesion, to be treated bysurgery.

Treatment

A biopsy was conducted with the surgical removal ofthe lesion, performed under general anesthesia andlocal infiltration using Mepivacaine 2% with 1:200,00adrenaline. A mucoperiosteal vestibular flap was raised, followed by an osteotomy with a saw to opena rectangular bone window in the outside wall of themaxillary sinus cavity that compromised the ascending apophysis of the same side (Fig 4). This allowed suitable access to the maxillary antra, as well as to the tumor lesion to be able to remove it together with the affected third molar. A goodcleavage plane was obtained, enabling very little sinus mucosa to dry out, along with the tumor tissue,which appeared encapsulated. (Fig. 5)

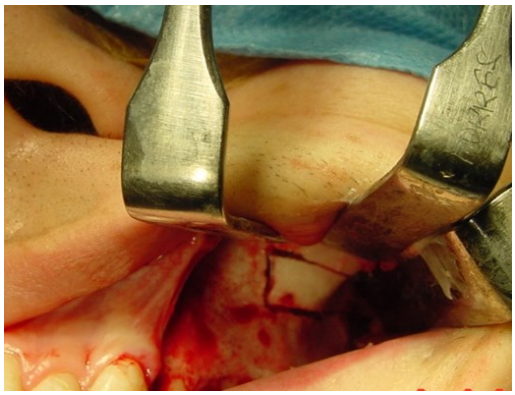

Figure. 4 *Mucoperiosteal flap and Bone Window to theMaxillary Sinus.*

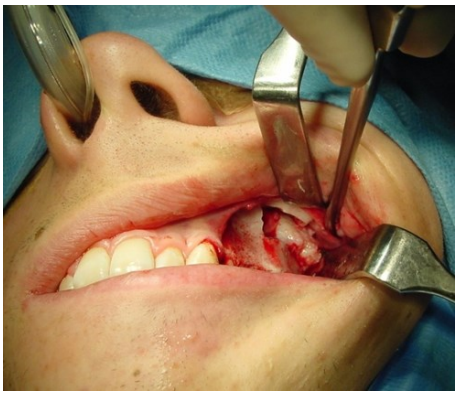

Figure. 5 *Vestibular Bone Window to the Maxillary Sinus, withthe tumor* removal.

The surgical bed was examined, then the entire area was cleaned. The piece of bone extracted from the bonewindow during initial osteotomy was returned and sutured in place. Afterward, the mucoperiosteal flap was closed. (Fig. 6)

The tissue samples, or specimens, were sent in threeseparate vials to Pathological Anatomy.

In their macroscopic analysis, they are referred to as Vial #1, Vial #2 and Vial #3. Vial 1 Specimen. Contains several pieces of soft, whitish-brown, irregular tissue; together toy measureapproximately 30 x 15 x 10 mm. In addition, various hard pieces of papyraceous tissue and a dental follicle (not processed) were found. (Fig. 7)

Vial #2 Specimen. Contains a piece of soft whitetissue measuring approximately 10 x 5 x 5 mm.

Vial #3 Specimen. Consists of a piece of soft tissuemeasuring approximately 7 mm.

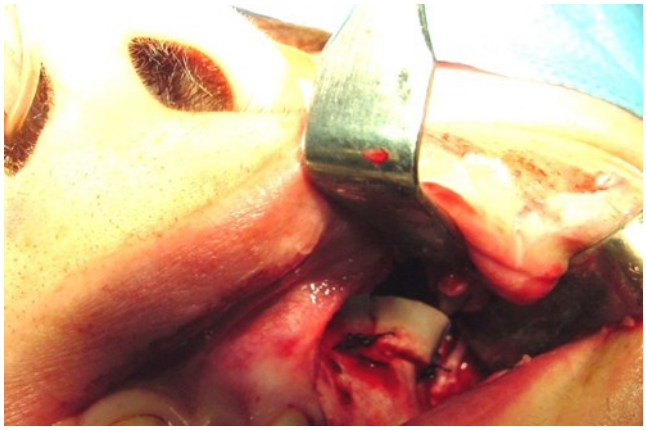

Figure. 6 *Bone window being sutured closed.*

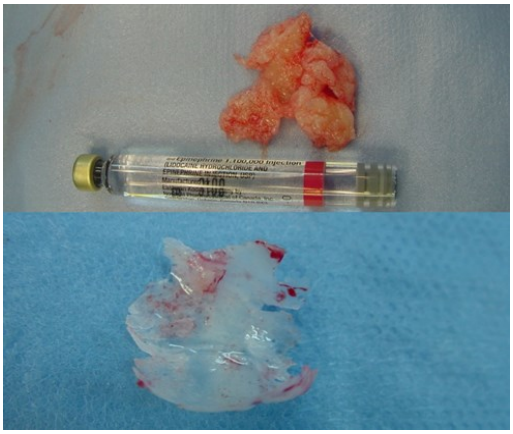

Figure. 7 *Tumor: "Surgical Piece of Neurilemmoma," Histologic Report prepared for the three samples sent for analysis.*

In their microscopic analysis, the lesion wasdescribed in three parts as well:

Vial #1 Specimen. We observed a proliferation of fused cells (fusiform) generally organized in parallelor palisade arrangements, and sometimes contouredaround acellular eosinophil areas called "Verocay Bodies."

These structures are associated with two types of tissue patterns or areas called Antoni A and Antoni B.The Antoni A-type pattern was described in theprevious paragraph. The other pattern observed is the Antoni B tissue area, comprised of randomly distributed (not in a parallel pattern) fusiform cells in a light-colored fibrillar matrix.

Visible are nerve fibers cut transversally in different zones. Also, observable is a more myxomatous arrangement with irregular focal areas. In addition, sinus mucosa can be found covered by respiratory epithelium with ample hemorrhaging in nearbyconjunctive tissue.

Vial #2 Specimen. Presents are some trabeculae of lamellar bone tissue within normal limits.

Vial #3 Specimen. A highly collagenized piece of tissue, with characteristics similar to those of Specimen #1.

Certainty of Diagnosis

The histologic diagnosis provided by the Pathologist was Neurilemmoma (Schwannoma) of the MaxillarySinus.

This was confirmed by the presence of Verocay Bodies and of the Antoni A and Antoni B Areas in thespecimens labeled Vial #1 and Vial #3.

The patient was monitored clinically and radiographically panoramic view only- for more than 9 years without any recurrences. At no point during follow-up was it necessary to request a CT scan. After that, the patient did not return for further follow-ups.

Discussion and Conclusion

The solitary Neurilemmoma (Schwannoma) is a benign neural neoplasm of Schwann cell origin and are slow-growing tumors. It is relatively uncommon, although 25% to 48 % of all cases occur in the head and neck region. Sixty percent originate in the neurilemma of cranial nerves, Schwannomas or Neurilemmomasassociated with the trigeminal system range from 0.8% to 8% in frequency, with various reports indicatingthat approximately 10% are asymptomatic. Must be treated by Surgical Excision. The lesion should not recur.

Sensory nerves are particularly affected by Neurilemmomas, while motor involvement is usually associated with Neurofibromatosis.

Neurogenic tumors can occur in any part of the neck, but are most often located in the pharynx space, which, in a physical examination, can exhibit the displacement of the amygdala to the median line. Theydo not have special diagnostic characteristics[20].

Neurogenic tumors comprise a very low percentage of neoplastic head and neck lesions[20].

This heterogenous group is composed of benign tumors (neurofibromas, neurilemmomas, neuromas, neurogenic nevus and granular cells myoblastomas) and by malignant tumors (neurogenic sarcomas, malignant schwannomas, neuroepitheliomas and melanomas).

Neurilemmomas do not have distinctive characteristics that allow a more certain clinical diagnosis. In themouth, a differential diagnosis must include other benign mesenchymal neoplasms or salivary gland tumors.

For neurilemmomas located in the maxillary sinus, such as in the case of this article, the hypothesis diagnostic and differential diagnostic possibilities were aimed at the cystic aspects, to define the treatment to follow.

Acknowledgments

We would like to express our special thanks to Dr. Manfred Seidemann (OMS) for his generous help inreviewing the case for publication.

Conflict of Interest

The authors declare no conflict of interest.

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Citation: Montes H. JL, Gonzalez M. I. Schwannoma (Neurilemmoma). Ectopic Position in Maxillary Sinus. *SVOA Dentistry* 2024, 5:4, 109-114. doi:10.58624/SVOADE.2024.05.0179