PLEXIFORM SCHWANNOMA OF THE LOWER LIP, TREATMENT AND MANAGEMENT AND A REVIEW OF LITERATURE
Subha Dhua¹, D. R. Sekhar²

HOW TO CITE THIS ARTICLE:
Subha Dhua, D. R. Sekhar. “Plexiform Schwannoma of the Lower Lip, Treatment and Management and a Review of Literature”. Journal of Evidence based Medicine and Healthcare; Volume 2, Issue 33, August 17, 2015; Page: 4968-4974, DOI: 10.18410/jebmh/2015/693

ABSTRACT: AIMS: To treat and manage a swelling on the lower lip which was insidious in onset and as gradually progressive on a 18 year old female. This has been a rare case and could be the first one in India as it was located on the lower lip with very few cases reported in the literature.

MATERIALS AND METHODS: An 18 year old female was referred to our Out Patients Department (OPD). The patient was apparently normal 10 years back when she developed a swelling of the lower lip which was insidious in on-set gradually progressive. Initially, it was of the size of 1x1 cm and gradually increased to 3x3cms. OBSERVATION: It was noted that in the Antoni A area typical Verocay bodies composed of palisading nuclei and surrounding spaces filled with eosinophilic filaments. No necrosis was noted and there were no atypical mitotic figures. In the Antoni B region a closely textured matrix with areas of edema, myxomatous changes cystic degeneration and dilated vessels were noted, Fig. 4a, b & c. On the basis of these histopathologic findings a diagnosis of Plexiform Schwannoma was inferred. On the basis of histopathologic findings and immunohistochemical profiles, a diagnosis of Plexiform Schwannoma was confirmed.

CONCLUSION: The excised mass revealed benign microscopic features and confirmed complete removal. Malignant transformation of a benign Plexiform Schwannoma which has been controversial by some authors was not considered. Surgical removal of the mass is considered to be the best course of treatment.

KEYWORDS: Antoni A, Verocay Bodies, S-100 Protein, T1 Weighted Images.

INTRODUCTION: Plexiform Schwannoma of the lower lip are rare neurogenic tumours arising from Plexiform Schwannoma neural sheath and are also termed neurilemmoma or neurinoma, is an uncommon benign true nerve-sheath neoplasm formed entirely of Schwann cell proliferation that occurs in the central and peripheral nerves. These are solitary benign, slow growing and smooth surfaced and are known to predominantly affect young adults and there is no sex predilection and commonly manifests as asymptomatic, slow growing solitary nodule. (Kun et al 1993, Michida et al. 1995, Fitzpatrik 1996).¹ Plexiform Schwannoma in the lower lip is a rare location with very few cases reported in the literature²,³,⁴,⁵,⁶ as shown in the table I below:
MATERIALS AND METHODS: An 18 year old female was referred to our Out Patients Department (OPD). The patient was apparently normal 10 years back when she developed a swelling of the lower lip which was insidious in on-set gradually progressive. Initially, it was of the size of 1x1cm and gradually increased to 3x3cms.

A careful physical examination revealed a flow, smooth surface, non-tender marks measuring 3x3cms located in the mucous of the lower lip, Fig. 1 below.

There was no history of discharge/pain. The medical history of the patient was unremarkable.

Abbreviation: NA – data not available.
The results of all laboratory tests were indicative of peripheral hypercellular (Antoni A) and central hypocellular (Antoni B) regions.

The differential diagnosis of this mass of the lower lip was fibroma, neurofibroma, Plexiform Schwannoma, leiomyoma, minor salivary gland tumour and other benign mesenchymal tumours. Physical examination revealed a firm non tender mass with a fairly smooth surface located in the vermilion area of the lower lip.

PROCEDURE: The mass was completely excised under general anesthesia. It was an encapsulated tumor mass measuring 3x3cms with a fairly firm and smooth surface, see Fig 2 below.

RESULT: It was found that in the Antoni A area typical Verocay bodies composed of palisading nuclei and surrounding spaces filled with eosinophilic filaments. No necrosis was noted and there were no atypical mitotic figures. In the Antoni B region a closely textured matrix with areas of
edema, myxomatous changes cystic degeneration and dilated vessels were noted, Fig. 4a, b & c. On the basis of these histopathologic findings a diagnosis of Plexiform Schwannoma was inferred. On the basis of histopathologic findings and immunohistochemical profiles, a diagnosis of Plexiform Schwannoma was confirmed.

Microscopically, the characteristic histological features for a schwannoma of the lip are similar to those analogs found at other sites. These include complete tumor encapsulation and composition consisting of alternating regions of hypercellularity and hypocellularity known as Antoni A and Antoni B areas, respectively. In general, the relative proportions of the 2 regions vary to the extent that either may occupy nearly the entire tumor. Usually, the
hypercellular Antoni A region consists of monomorphic spindle shaped Schwann cells with pointed basophilic nuclei and poorly defined eosinophilic cytoplasm. Occasionally, Verocay bodies, made up of palisading spindle-shaped cells around eosinophilic fibrils, are present in the Antoni A area, as noted in our case. (Fig. 4a, 4b, 4c).\textsuperscript{[5,13]} Further, secondary changes, such as cystic degeneration, vessels with thick hyaline walls and hemorrhage, Fig. 4a, b, c) may occur in the Antoni B tissue.\textsuperscript{[2,6,13,14]} These secondary changes were also observed in the present case.

Mitosis and necrosis were not detected. Alternatively, a loosely textured matrix was determined for the Antoni B tissue, with areas of edema, myxomatous changes, cystic degeneration, and the presence of dilated hyaline vessels. (Fig. 4) Immunohistochemical staining was strongly positive for S-100 protein.

Intense S-100 protein immune reactivity for cytoplasmic and nuclear patterns indicates neural origin.\textsuperscript{[1,6]} These staining results and the associated H&E microscopic findings confirmed the diagnosis of Plexiform Schwannoma in the reported case.

The overall prognosis of Plexiform Schwannoma is quite favourable. Conservative surgical removal is the treatment of choice, with wide excision has achieved normally and no recurrence.\textsuperscript{[2,5,7,12]} These staining results and the associated H&E microscopic findings confirmed the diagnosis of Plexiform Schwannoma in the reported case.

These rare Plexiform Schwannoma tumours being non-specific in presentation, clinical diagnosis becomes difficult. Therefore, differential diagnosis may include fibroma, pleomorphic adenoma and other salivary gland tumours.\textsuperscript{[5]}

**DISCUSSION:** Plexiform Schwannoma are uncommon benign solitary encapsulated neoplasms arising predominantly from both peripheral and intracranial points of cranial nerve site whereas peripheral cranial nerve Plexiform Schwannomas are usually located in parapharyngeal space of the neck and in soft tissues such as tongue, buccal mucosa, palate and gingiva.\textsuperscript{[3]} In parapharyngeal space, the most commonly involved nerves are the vagus and the cervical sympathetic chain.\textsuperscript{[15]} Dass Gupta et al\textsuperscript{[16]} in 1969 described Plexiform Schwannoma with the lip being a rare entity area of occurrence followed by the palate and buccal mucosa. (Barbosa J. Hnsen LS.

Most commonly Plexiform Schwannoma are characterized by their solitary occurrence relatively smooth surface and slow growth.

Most Plexiform Schwannoma are asymtomatic and if the regional tumour is small, most likely it is not identified with ease. However, in the event of larger tumour it does occur inside the epineural sheath and undergoes progressive enlargement with the nerve fascicles spreading out of the surface of the tumour.\textsuperscript{[7,11]}

The characteristic features of the Plexiform Schwannoma of the lip historically are fairly similar to those of analogues found at other sites. The tumour has two distinct patterns and is described as Antoni A and Antoni B areas and the relative portions of the two areas vary considerably. The Antoni A hyper cellular areas primarily consist of monomorphic spindle shaped Schwann cells with pointed basophilic nuclei and poorly defined eosinophilic cytoplasm.\textsuperscript{[2,6,12]} Antoni B areas consist of loosely arranged cells and small cystic spaces. In Antoni B area, the cystic degeneration, vessels with thick hyaline walls and hemorrhage were observed.\textsuperscript{[11,12]}
Immuno histochemically, positive S-100 protein and lev-7 antigen reactivity warrants Schwann cell nature of these tumors\(^1,4\)

For Plexiform Schwannomas of the lip, ultrasound, computed tomography and magnetic resonance imaging are also used for preoperative diagnosis. Homogeneous, hypo echoic features and posterior acoustic enhancement are seen on ultrasounds.\(^5\) Computerized tomography shows homogeneous soft\(^7\) tissues density mass with clear margins\(^4\) and magnetic resonance imagining shows a homogenous lesion with low intermediate signal intensity on T1 weighted images and high signal intensity on T2 weighted images.\(^6\)

The confirmatory tests lie with histopathological examination and in the case presented it was confirmed only with the histo-pathological examination.

CONCLUSION: The excised mass revealed benign microscopic features and confirmed complete removal. Malignant transformation of a benign Plexiform Schwannoma which has been controversial by some authors was not considered. Surgical removal of the mass is considered to be the best course of treatment.

ACKNOWLEDGEMENT: The authors would like to thank Dr. Prasit Kumar Ghosh, PG Trainee and Dr. Rekha Sody, Associate Professor, Pathology Department, Vydehi Institute of Medical Sciences, Bangalore, India for their assistance in carrying out the histopathological investigations and Dr. Sanket Shetty, M.Ch Trainee, Plastic & Reconstructive Surgery Department

REFERENCES:
1. Kun Z, Qi DY. Zhang KH: A comparison between the clinical behavior of neurilemmomas in the neck and oral and maxillofacial region. J. Oral Maxillofacial Surg 51: 769-771, 1993.
2. Baderca F, Cojocaru S, Lazar E, Lazureanu C, Faur A, Lighezan R, Alexa A., Raica M, Valean M, Balica N: Schwannoma of the lip: case report and review of the literature. Rom J Morphol Embryol 49: 391-398, 2008.
3. Cardoso CL, Tolentino Ede S, Capeloza AL, Consolaro A: Schwannoma in the lower lip mucosa: unexpected diagnosis, Quintessence Int. 41:769-771, 2010.
4. Thurnher D, Quint C, Pammer J, et.al. Dysphagia due to a large schwannoma of the oropharynx: case report and review of the literature. Arch Otolaryngol Head Neck Surg 2002; 128: 850-852.
5. Cherrick HM, Eversole LR. Benign neural sheath neoplasm of the oral cavity. Report of thirty-seven cases. Oral Surg Med Oral Pathol 1971; 32: 900-909.
6. Asaumi J, Konouchi H, Kishi K. Schwannoma of the upper lip: ultrasound, CT, and MRI findings. J. Oral Maxillofac Surg 2000; 58: 1173-1175.
7. Yang, Shih-Wei, Lin, Chin-Yew, et al., Schwannoma of the Upper Lip: Case Report and Literature Review, Am J, Otolaryngol, 2003: 24, 351-354.
8. Yilmaz ND, Tokyal C. Derekozy FS Altuntas A: Schwannoma of the upper lip: a case report. Kulak Burun Bogaz Ihtis Derg 12-42-44, 2004.
9. Lobo I. Tomes T, Pina F, Dominguez M, Alves R. Barbos do Amara J, Selores M: Plexiform schwannoma of the lip mucosa J Eur Acad Dematol Venereol 23: 61-618, 2009.
10. Jukovic R, Stanko P, Galbavy S, Sieberova G, Babal P: Recurrent malignant epitheloid schwannoma of the lower lip. Bratisl Lek Listy 10: 116-119, 2009.
11. Pahwa R, Khurana N, Uma CK, et al. Neurilemmoma of tongue. Ind J Oral Surg 1968; 26: 651-658.
12. Michida A, Ryoke K, Ishkura S, et al. multiple schwannomas of the neck, mediastinum, and parapharyngeal space: Report of case. J Oral Maxillo Fal Surg 1995; 53: 617-620.
13. De Bree R, Westerveld GJ, Smeele LE. Submandibular approach for excision of a large schwannoma in the base of the tongue. Eur Arch Otorhinolaryngol 2000; 257: 283-286.
14. Triaridis C, Tsilighopoululos MG, Kououlas A, et al. Posterior Pharyngeal Wall schwannoma. J Laryngo Otol 1987; 101: 749-752.
15. Yang SW, Lin CY. Schwannoma of the upper lip: case report and literature review. American J Otolaryngology 2003; 24: 351-354.
16. Das Gupta TK, Brasfield RD, Strong EW, et al. Benign solitary schwannomas (neurilelemomas). Cancer 1969; 24: 355-366.

AUTHORS:
1. Subha Dhua
2. D. R. Sekhar

PARTICULARS OF CONTRIBUTORS:
1. Assistant Professor, Department of Plastic Surgery, Vydehi Institute of Medical Sciences & Research Centre, Bengaluru.
2. Professor & HOD, Department of Plastic Surgery, Vydehi Institute of Medical Sciences & Research Centre, Bengaluru.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Subha Dhua,
Assistant Professor,
Department of Plastic Surgery,
Vydehi Institute of Medical Sciences & Research Centre, Bengaluru.
E-mail: subhadhua@yahoo.com

Date of Submission: 26/07/2015.
Date of Peer Review: 27/07/2015.
Date of Acceptance: 01/08/2015.
Date of Publishing: 13/08/2015.