DOI: 10.14260/jemds/2015/381

CASE REPORT

UNUSUAL NEUROFIBROMA OF LARYNX: A CASE REPORT

S. Surya Prakasa Rao[1], T. V. S. S. N. Leela Prasad[2], D. G. Harikrishna[3], N. Veeraswamy[4], H. Kishore Dora[5]

HOW TO CITE THIS ARTICLE:
S. Surya Prakasa Rao, T. V. S. S. N. Leela Prasad, D. G. Harikrishna, N. Veeraswamy, H. Kishore Dora. "Unusual Neurofibroma of Larynx: A Case Report". Journal of Evolution of Medical and Dental Sciences 2015; Vol. 4, Issue 15, February 19; Page: 2641-2644, DOI:10.14260/jemds/2015/381

ABSTRACT: Neurofibromatosis originates from the Schwann cells of the nerve sheath of the nerve which is benign in nature. In von Recklinghausen's disease multiple masses arises all over the body. It may occur from internal nerves sporadically. These are the white glistening round or lobulated masses arise in the course of nerves involved. Neurofibroma of larynx is seldom reported. Women are more affected than men in the ratio 3:2. Hoarseness is the earliest and common symptom, the patient present with. If the mass increase in the course of time, the dyspnoea, later dysphagia symptoms wills result. The arytenoid is the common site of origin, later aryepiglottic fold, false vocal cord, ventricle of the larynx. As the tumor is benign in nature, total excision of tumor is recommended. In the literature, very few cases of laryngeal involvement are reported. This article presents a case report of neurofibroma of larynx involving false vocal cord and ventricle presented submucosally with symptom of hoarseness, primarily with mild stridor on exertion. In this large endolaryngeal mass, laryngofissure surgical approach is selected as a choice.
KEYWORDS: Neurofibroma, endolaryngeal tumor, neurofibromatosis.

INTRODUCTION: Endolaryngeal neurofibromas are very rare tumors. The neurofibroma in the subcutaneous tissue are common which does not require any surgical intervention except when they give cosmetic defect and interfere with function as a result of its anatomical position. Neurofibroma may occur anywhere in the head and neck, including lips, palate, tonsil, tongue, posterior pharyngeal wall, parapharyngeal area, parotid and larynx. Neurofibroma is a neurogenic tumor originating from the peripheral nerve, which occurs sporadically or in part with von Recklinghausen's disease. It presents in two forms, plexiform and non plexiform. It occurs rarely in larynx reported first time in 1952 as an isolated endolaryngeal neurofibroma. Commonly it presents with symptoms like hoarseness, dyspnoea, dysphagia.(1)

It presents more commonly in women than men.(2,3) It affects larynx in descending order as arytenoids, aryepiglottic folds, rarely subglottic area. These are non-encapsulated, nerve fibers traverse, and usually present in the mass. Histologically, spindle cells with enlarged nuclei seen. Sometimes it may present with neurologic abnormalities like spin bifida or glioma.(4) According to Supance, Quenelle and Crissman, solitary neurofibroma of larynx are frequently present than combinedly occurring with von Recklinghausen's disease.(5) As the tumor is benign in nature, total excision conservatively is adequate. Most commonly affected cranial nerves are (CN) II and (CN) VIII. Neurofibroma associated with von Recklinghausen's disease will have 4.6 to 16% chance of sarcomatous changes.

CASE REPORT: CASE: In November 2014, a 15 year old male named L. Ganesh came to the Government ENT hospital with complaint of hoarseness of voice for 6 months. It is insidious in onset and gradually increasing in the course of one month. It is associated with difficulty in breathing on

DOI: 10.14260/jemds/2015/381

exertion and dysphagia for solids. On Indirect Laryngoscopic examination, a smooth lobulated mass of size 3x2x1cm seen on the left aryepiglottic fold extending to false cord was detected. The mass had completely obscured the view of endolarynx and pushed the epiglottis to opposite side. CT scan showed a well circumscribed soft tissue mass in the left aryepiglottic folds extending to false cord. On Direct Laryngoscopy, a sub mucosal mass on left aryepiglottic fold extending to false cord was revealed. The surface is smooth, glistening and reddish pink in colour. There is no ulceration and signs of inflammation. Preoperative tracheostomy was done.

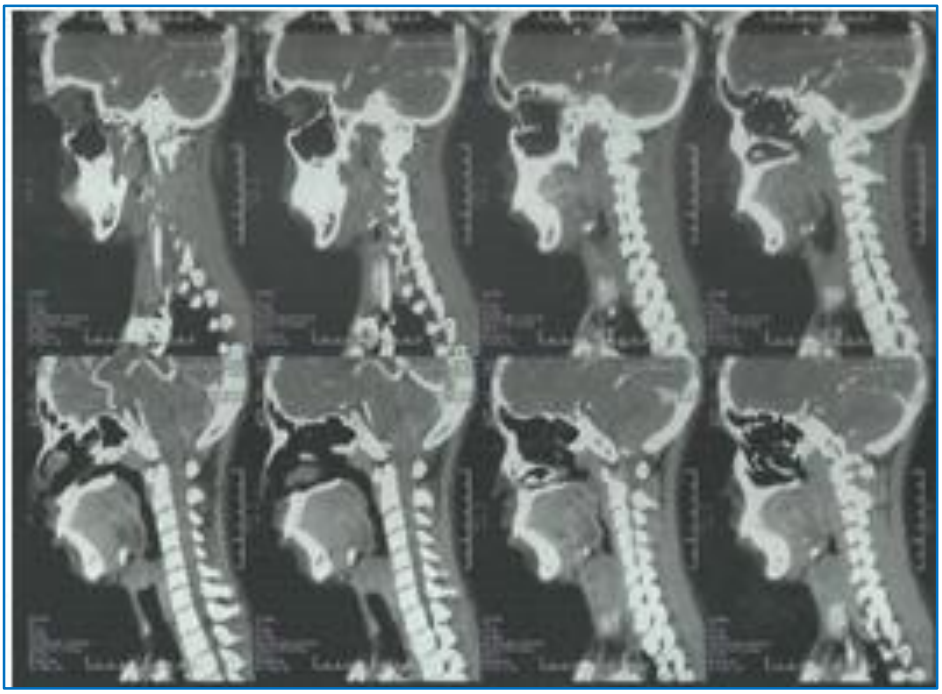


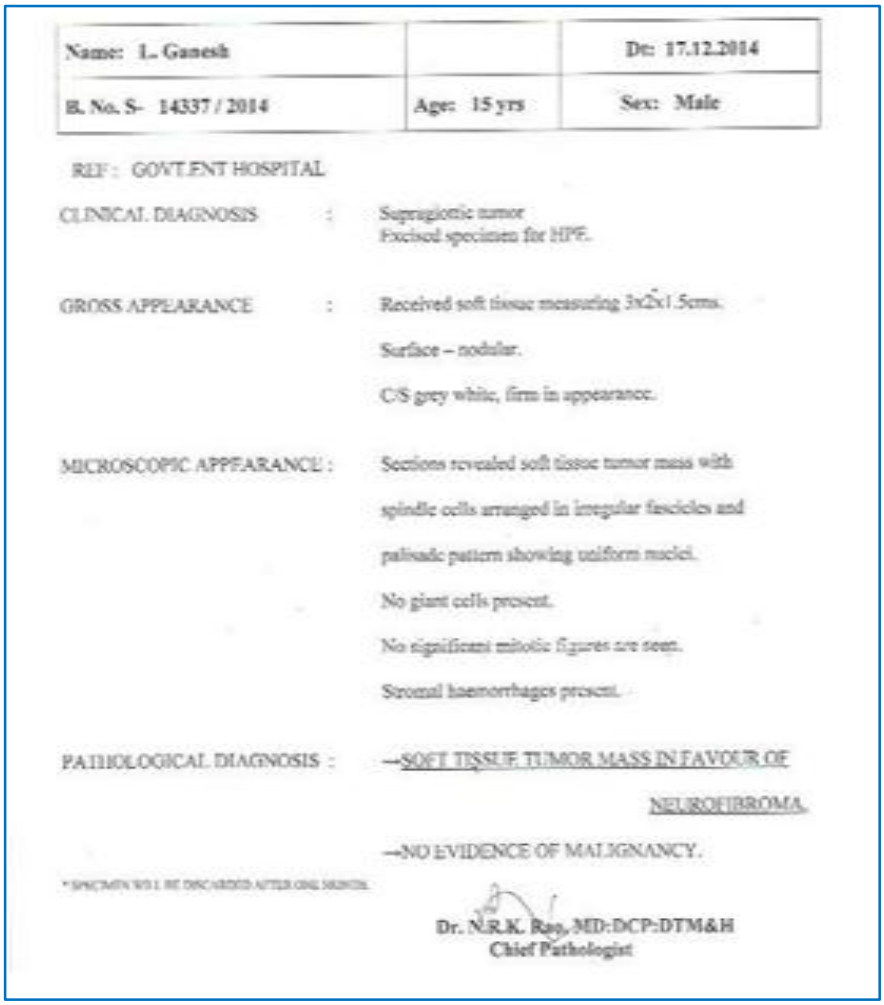
Fig. 1

MANAGEMENT: Surgery is the treatment of choice for laryngeal neurofibroma. Endoscopically small sized tumors can be excised, but for large tumors external approach is the preferred choice. Depending on site and size of tumor, three types of external approaches are used like Laryngofissure, lateral pharyngotomy and sub-hyoid pharyngotomy. In this case, Laryngofissure approach was selected. Under General Anesthesia, patient kept in supine position with neck extended, a horizontal skin crease incision given. Thyroid cartilage exposed and opened in the midline. A whitish glistening 3x2x1.5 cm mass seen on the left aryepiglottic folds extending on to the false cord. Mass was removed by sub mucosal dissection and wound closed in layers.

SECTION SHOWED: Soft tissue tumor mass with spindle cells arranged in irregular fascicles and palisade pattern showing uniform nuclei. No giant cells present. No significant mitotic figures seen. Stromal haemorrhages present.

PATHOLOGICAL DIAGNOSIS: A soft tissue mass in favour of neurofibroma. No evidence of malignancy.

DOI: 10.14260/jemds/2015/381



Name: L. Ganesh		Dt: 17.12.2014
B. No. S- 14337 / 2014	Age: 15 yrs	Sex: Male

REF : GOVT.ENT HOSPITAL

CLINICAL DIAGNOSIS : Supraglottic tumor
Excised specimen for HPE.

GROSS APPEARANCE : Received soft tissue measuring 3x2x1.5cms.
Surface – nodular.
C/S grey white, firm in appearance.

MICROSCOPIC APPEARANCE : Sections revealed soft tissue tumor mass with spindle cells arranged in irregular fascicles and palisadic pattern showing uniform nuclei.
No giant cells present.
No significant mitotic figures are seen.
Stromal haemorrhages present.

PATHOLOGICAL DIAGNOSIS : –SOFT TISSUE TUMOR MASS IN FAVOUR OF NEUROFIBROMA.
–NO EVIDENCE OF MALIGNANCY.

* SPECIMEN WILL BE DISCARDED AFTER ONE MONTH.

Dr. N.R.K. Raju, MD:DCP:DTM&H
Chief Pathologist

After 2 weeks tracheostomy tube was weaned successfully. Patient adviced for 1 month regular visit for a period of 6 months for any recurrence.

DISCUSSION: Endolaryngeal neurofibromas are seen occasionally, of which solitary neurogenic tumors are common than multiple neurofibromas. If a neurofibroma is seen, other manifestations like axillary freckling, café-au-lait spots and family history should be enquired carefully. On histopathological examination, neurofibroma arises from the Schwann cells of the nerve sheath. It is non-encapsulated and forms round white mass along the course of the nerve. Prominent collagen production, diffusely distributed mast cells and entrapped nerve fibres are seen in neurofibroma. Men are less commonly affected than women.[6]

The commonest symptoms are hoarseness of voice, dyspnea and dysphagia and commonest sites are aryepiglottic folds, arytenoids as seen in our case. Imaging prior to surgical excision is important. CT scan shows, mass in the supraglottic region. As the tumor is benign in nature, tumor is excised conservatively. The main aim of surgery is total excision with preservation of normal function. Large tumors can be excised through external approaches, like Laryngofissure, Lateral pharyngotomy and sub-hyoidpharyngology whereas small tumors are excised endoscopically. As there is no definite capsule for tumor, recurrence is common.

DOI: 10.14260/jemds/2015/381

REFERENCES:

1. El – Serafy S. Rare benign tumors of the larynx. J Laryngol Otol. 1971 Aug; 85 (8): 837-851.
2. Chen YW, Fang TJ, Li HY. A solitary laryngeal meurofibroma in a pediatric patient. Chang Gung Med J. 2004 Dec; 27 (12): 930-933.
3. Gstottner M, Galvan O, Gschwendtner A, Neher A. Solitary subglottic neurofibroma: a report of an unusual manifestation. Ear Arch Otorhinolaryngol.2005 Sep; 262 (9): 705-707.
4. Zobell DH. Massive neurofibroma of the larynx. Case report. Laryngoscope. 1964 Feb; 74:233-240.
5. Cummings CW, Montgomery WW, Balogh K Jr. Neurogenic tumors of the larynx. Ann Otol Rhinol Laryngol. 1969 Feb; 78 (1): 76-95.
6. Pearlmen SJ, Friedman EA, Appel M. Neurofibroma of the larynx. Arch Otolaryngol. 1950 Jul; 52 (1): 8-14.

AUTHORS:

1. S. Surya Prakasa Rao
2. T. V. S. S. N. Leela Prasad
3. D. G. Harikrishna
4. N. Veeraswamy
5. H. Kishore Dora

PARTICULARS OF CONTRIBUTORS:

1. Associate Professor, Department of ENT, Andhra Medical College, Government ENT Hospital.
2. Assistant Professor, Department of ENT, Andhra Medical College, Government ENT Hospital.
3. Assistant Professor, Department of ENT, Andhra Medical College, Government ENT Hospital.
4. Assistant Professor, Department of ENT, Andhra Medical College, Government ENT Hospital.
5. Post Graduate, Department of ENT, Andhra Medical College, Government ENT Hospital.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:

Dr. T. V. S. S. N. Leela Prasad,
D. No. 43-21-41,
Venkataraju Nagar,
TSN Colony,
Visakhapatnam-16.
E-mail: leelaprasaddoctor@gmail.com

FINANCIAL OR OTHER COMPETING INTERESTS: None

Date of Submission: 27/01/2015.
Date of Peer Review: 28/01/2015.
Date of Acceptance: 10/02/2015.
Date of Publishing: 19/02/2015.