University Journal of Surgery and Surgical Specialities

ISSN 2455-2860 2020, Vol. 6(3)

EUROFIBROMA OF THE SUPRAGLOTTIS - A CASE REPORT

VINOD P
Department of ENT,CHRISTIAN MEDICAL COLLEGE

Abstract : Neurofibromatosis is a benign neurogenic tumor, originating from schwann cells of the nerve sheath. It occurs sporadically or in association with Von Recklinghausens disease. Women are more predisposed to this condition (FM 32). Laryngeal involvement is rare and the symptoms depend upon the size and location of this tumor varying from irritation, hoarseness, dysphagia, a feeling of fullness in the throat to dyspnoea, or combinations of these. Owing to the slow growth of these tumors, the symptoms may take a few months to several years to manifest , hence eluding early diagnosis.The literature to date has reported only a few cases of laryngeal involvement, with the aryepiglottic fold being the most frequent site of occurrence. The other sites involved are the ventricular folds and the free edges of the true vocal cord. The site and the size of the tumor may obscure the endolaryngeal inlet, making it impossible to visualize the vocal cords. However being a benign tumor, conservative surgery is the preferred management .We present a patient with of supraglottic neurofibroma and a brief review of literature.

Keyword : Laryngeal neurofibroma, laryngoscope, laser, pharyngotomy.

INTRODUCTION

Laryngeal neurofibromas are extremely rare, accounting for 0.03-0.1% of benign tumours of the larynx (1). Less than 30 cases of neurofibromas of the larynx have been reported in the literature (2). The confirmation of laryngeal neurofibroma is based on the histopathologic demonstration of the characteristic spindle cell and immunohistochemical staining for S-100 protein. We present a case of laryngeal neurofibroma in a 19 year old man excised initially by endoscopic transoral laser excision followed by lateral pharyngotomy to facilitaite complete tumor removal.

Case report

A 19 year old gentleman presented with change in voice since childhood. He had laboured breathing which became worse at night for the past 1 year. On physical examination he was comfortably seated and had multiple subcutaneous nodules and *cafe-au-lait* spots on the entire body. A history of familial neurofibromatosis was asked for, which was negative. Flexible nasopharyngolaryngoscopy revealed a smooth mucosa covered mass on the right aryepiglottic fold obscuring the right vocal cord and narrowing the supraglottic airway (Fig 1)

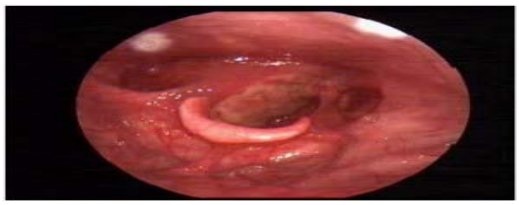

Fig 1: Mass in the right aryepiglottic fold

A contrast enhanced Computed tomography of the head and neck revealed a well-defined 4x3.2x4cm (TRxAPxCC) poorly enhancing lesion in the right side of the supraglottis with craniocaudal extent from upper border of C4 to lower border of C6 superiorly, causing complete obliteration of the right pyriform sinus with asymmetric thickening of the right pharyngoepiglottic fold. The lesion obliterated the right parapharyngeal space . The right vocal cord was medially displaced and the subglottis was free .(Fig 2)

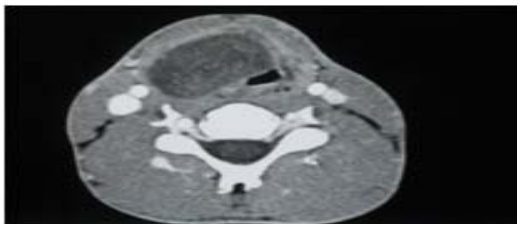

Fig 2: CT neck with contrast, axial cuts, shows enhancing mass arising from supraglottis.

Given the concern for airway obstruction and based on the size and location of the mass, the patient was planned for direct laryngoscopy and transoral CO2 laser resection under general anaesthesia. Intraoperatively a mucosa-covered 7.5 x 5 cm mass was seen involving the medial surface of the right aryepiglottic

fold, false cord, right arytenoid, paraglottic space, and extending submucosally inferiorly up to the cricoid cartilage. An endoscopic laser excision was planned. However peroperatively, the posterior paraglottic component could not be completely excised, hence a lateral pharyngotomy was done. The intraoperative and postoperative period was uneventful and he had no post-operative complications. Histopathological examination revealed a nodular lesion composed of spindle cells in a myxoid stroma of collagen fibres and myelin producing Schwann cells (Fig 3). Spindle cells were positive for S-100 indicating neurogenic origin (Fig 4). There was no dysplasia or evidence of malignancy.

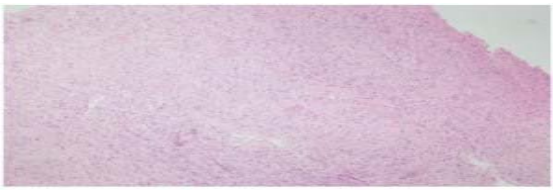

Fig 3: Schwann cells intertwined with fibroblasts

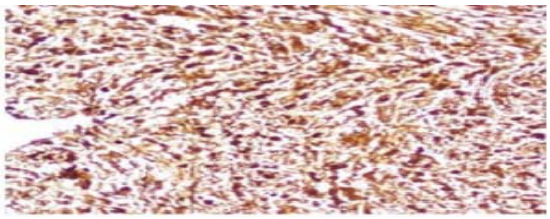

Fig 4: S-100 IHE

He followed up four months later in our OPD and was asymptomatic. A flexible endoscopy done on follow up showed a right vocal cord palsy, with no recurrence.

DISCUSSION

Neurogenic tumours of the larynx are extremely rare. They account for 0.03-0.1 % of all benign laryngeal tumours, neurofibromas being less common than schwannomas (1). Less than 30 cases of endolaryngeal neurofibromas have been reported in literature since its first description by Hollinger in 1950 (3). The ratio of females to males affected by laryngeal neurofibromas is 3:2 (4). Its presenting symptoms are variable depending on the patient's age, size and location of the lesion. Children often present with stridor, whereas in adults a variety of symptoms ranging from progressive change in voice to dyspnoea and even throat discomfort on swallowing are seen. The most frequently affected area is the supraglottic region, with the majority involving the arytenoids and the aryepiglottic fold, followed by the false vocal cords (4, 5) owing to a rich supply in terminal nerve plexuses (6). Supraglottic involvement is the rule, although subglottic tumours involving the trachea have also been reported (4,5) . The tumours arise from terminal nerve plexuses in the submucosal space of the supraglottic larynx. Because of the predominant supraglottic location, it is believed that the neurofibromas arise from the superior laryngeal nerve. It appears as a smooth round mass and is firm or elastic during a laryngeal examination under anaesthesia (7). Pathologically, most lesions of the larynx in patients with von Recklinghausen disease are neurofibromas. Schwannomas may also occur in patients with neurofibromatosis. Neurofibromas possess collagen-producing fibroblasts as well as myelin-producing schwann cells and are not as well encapsulated as schwannomas (4, 7). Malignant transformation into neurofibrosarcomas and malignant schwannomas has been reported in the larynx, usually in patients with von Recklinghausen disease (8). Our experience with this patient has taught us that the presence of a well-defined unilateral submucosal mass should raise the suspicion of a neurogenic tumor. The treatment of choice for laryngeal neurofibromas is surgical removal. Excision of the tumor using a laryngoscope or through an external approach depends on the size and location of the lesion. Smaller lesions can be approached endolaryngeally with or without laser .Larger tumours may require external approaches like lateral pharyngotomy, lateral thyrotomy or laryngofissure technique (9, 11). Non-plexiform neurofibromas have a good prognosis after excision, especially if complete removal is achieved during primary surgery (10). However plexiform neurofibromas which are pathognomonic of neurofibromatosis carry malignant potential and require extensive surgical exposure to get wider margins and complete extirpation can be achieved only by external approach (12).In the case reported here, we initially excised the tumour by endoscopic transoral laser excision followed by lateral pharyngotomy for complete tumor removal.

CONCLUSION

Neurofibromas should be considered as one of the differential diagnosis, not only in children but also in adults presenting with a submucosal laryngeal mass, even if other clinical signs of NF1 or NF2 are absent. Diagnosis can be only confirmed by direct laryngoscopy & biopsy. Endoscopic approach for resection is ideal for the preservation of laryngeal function. Long term follow-up is essential due to the high rate of recurrence and residual disease as well as the possibility of malignant change.

REFERENCES

1) Jones SR, Myers EN, Barnes L, Benign neoplasm of the larynx. Otolaryngol Clin North Am 1984: 17:151-178.

2) David M.Yousem, Carl Oberholtzer, Neurofibroma of the aryepiglottic fold. AJNR Am J Neuroradiol 1991 Nov –Dec:12 (6):1176-8.

3) Jiaying Liu, Chin Fong Wong, Francoise Lim, Jeev Kanagalingam. Glottic neurofibroma in an elderly patient : A case report. J Voice 2013 Sep:27(5):644-6.

4) Supance JS, Queneue DJ, CJ, Endolaryngeal neyrofibroma. Otolaryngol Head Neck Surg 1980, 88:74-78.

5) Chang-Lo M. Laryngeal involvement in Von Recklinghausen's disease: a case report and review of the literature, Laryngoscope. 1977: 87:435–442.

6) Helg JR. Neurinomas of the larynx and neurofibromatosis of von Recklinghausen's. Pract Otorhinolaryngol (Basel) 1950: 12:165-84.

7) Maisel RH, Ogura JH. Neurofibromatosis with laryngeal involvement Laryngoscope 1974; 84:132- 40.

8) Stines J, Rodde A, Carolus JM, Perrin C, Becker S. CT findings of laryngeal involvement in von Recklinghausen disease. J Comput Assist Tomogr 1987;11:141-3.

9) Ramakrishnan, Issing Laryngeal Schwannoma: Case Report and Literature Review. ISRN Otolaryngology Volume 2011, Article ID 540643

10) Maship MJ,Esteban E,Alberto C,Menor E,Cortina H. Laryngeal involvement in pediatric neurofibromatosis: a case report and review of the literature. Pediatr Radiol.1996:26:488-492.

11) Nakahira M, Nakatani H, Sawada S, Mtsumoto S, Neurofibroma of the larynx in neurofibromatosis. Arch Otolaryngol Head Neck Surg 2001:127:325-328.

12) Mitsuhiko Nakahira, MD; Hiroaki Nakatani, MD; Shoichi Sawada, Neurofibroma of the Larynx in Neurofibromatosis Preoperative Computed Tomography and Magnetic Resonance Imaging *Arch Otolaryngol Head Neck Surg.* 2001;127(3):325-328.

An Initiative of The Tamil Nadu Dr. M.G.R. Medical University
University Journal of Surgery and Surgical Specialities

An Initiative of The Tamil Nadu Dr. M.G.R. Medical University
University Journal of Surgery and Surgical Specialities