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LARYNGEAL NEUROFIBROMA - A RARE MANIFESTATION OF NEUROFIBROMATOSIS 1

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ABSTRACT

Neurofibromatosis type 1(NF-1) is an autosomal dominant neurocutaneous disorder characterised by tumors in skin, bone and nervous system[1]. Laryngeal neurofibroma is a rare manifestation of NF-1 and one of the rarest benign tumours of larynx. The management of laryngeal neurofibroma varies according to the site of involvement in larynx and extent of the lesion. The most common site of involvement is arytenoid or aryepiglottic fold which was similar in our case. Complete removal of larger tumours often require external approach.

KEYWORDS

Neurofibromatosis 1, Laryngeal Neurofibroma, Benign Tumour, Lateral Pharyngotomy

1. INTRODUCTION

First introduced by von Recklinghausen in 1882, neurofibromatosis type I is an inherited neurocutaneous disorder characterised by multiple café-au-lait spots and neurofibromas. Neurofibromas are benign peripheral nerve sheath tumours arising from fibrous covering of peripheral nerve which are rare in larynx[2]. The most common site of neurofibroma in larynx is supraglottis.

2. Case Report

18 year old male, known case of neurofibromatosis type I and occult spina bifida presented with complaints of change in voice of 3 years duration and snoring of 2 years duration. Patient had multiple neurofibromas in the upper limb and café-au-lait macules in trunk. There was no difficulty in breathing or swallowing and noisy breathing when awake. Videolaryngoscopic examination showed smooth surfaced globular mass involving right aryepiglottic fold and arytenoid, obscuring view of glottis **(Fig 1).**

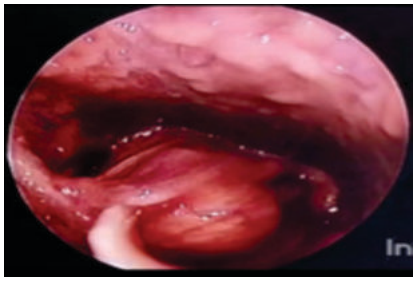

Figure 1: Videolaryngoscopic image

MRI Neck showed a well defined encapsulated T2 hyperintense lesion in the right paraglottic space predominantly in supraglottic and glottic region of size 3.5 x 3 x 4.9 cm displacing epiglottis, right aryepiglottic fold and vocal cord medially **(Fig 2).**

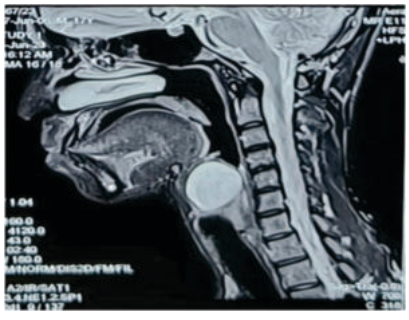

Figure 2: MRI Neck

Patient underwent elective tracheostomy followed by lateral pharyngotomy of right side. A horizontal incision was put at the level of lower border of thyroid cartilage on right side. Subplatysmal flap elevated and strap muscles divided. Superior part of thyropharyngeus separated from insertion. Mucosa of pyriform fossa exposed, lateral pharyngotomy done, tumour identified which was attached to right aryepiglottic fold and epiglottis, same removed and sent for histopathological examination **(Fig 3)**.

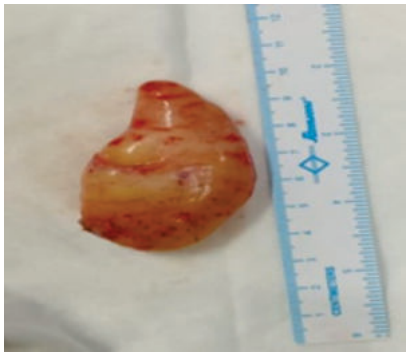

Figure 3: Excised specimen

Pharyngeal mucosa repaired by intermittent absorbable sutures, strap muscles sutured and wound closed with suction drain. Post operative period was uneventful. Post operative videolaryngoscopy showed right vocal cord palsy with adequate glottic space **(Fig 4).**

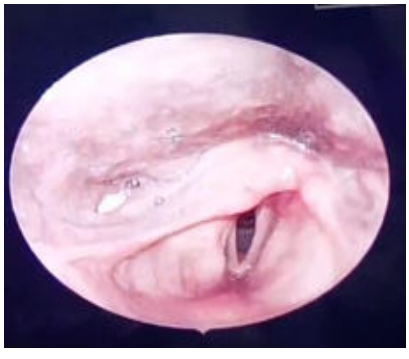

Figure 4: Post op videolaryngoscopy image

Histopathological examination showed loosely arranged spindle cells with wavy serpentine nuclei and pointed ends in the background of fibrocollagenous stroma **(Fig 5)**.

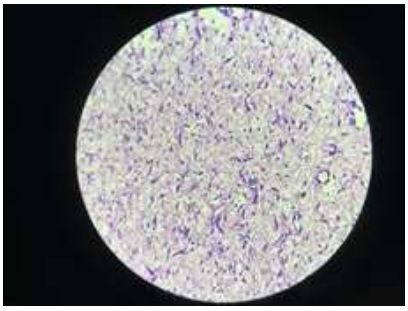

Figure 5: Histopathology examination showing spindle cells in fibrocollagenous stroma

3. DISCUSSION

Neurofibromas are extremely rare in larynx. They may occur in isolation, but more commonly associated with NF1 or NF 2[3]. The first case of laryngeal neurofibroma was reported by Jackson and Coates in 1929. The most common sites of laryngeal involvement include arytenoids and aryepiglottic fold which are rich in terminal nerve plexuses. The origin of tumour in supraglottis is commonly from superior laryngeal nerve or from anastomoses between the superior laryngeal nerve and recurrent laryngeal nerve[4].

Laryngeal neurofibromas are usually associated with NF 1 and not with NF 2. The presentations of NF 1 are café-au-lait macules, neurofibromas, axillary or inguinal freckling, Lisch nodules, skeletal and neurological abnormalities.

Due to its slow growing nature, laryngeal neurofibroma can remain asymptomatic for years. Various presenting features include hoarseness, dysphagia, foreign body sensation and stridor.On laryngoscopy, neurofibromas present as sessile or pedunculated bulges impinging on the airway[5].

Magnetic resonance imaging provides superior soft tissue characterisation. The appearance of solitary neurofibromas varies with high or low signal on T2-weighted images, depending on the admixture of fibrous tissue, and enhancement can be moderate and heterogenous or homogenous and intense[3]. Histologically, neurofibroma is composed of spindle cells embedded in a fibro-myxoid stroma. Immunohistochemically, neurofibroma is S100 and SOX10-positive tumour.

Surgical excision, endoscopically or via external approach is advocated based on the location, extension of tumour and the severity of symptoms. Endoscopic laser excision is recommended when the tumour is small and localised. An open approach (lateral thyrotomy, lateral pharyngotomy or laryngofissure) may be required for extensive lesions[3] and often requires tracheostomy for establishing adequte airway.

4. CONCLUSION

Neurofibroma, though rare, should be kept as a differential diagnosis in patients with a submucosal laryngeal mass. Based on its location and severity of symptoms, surgical excision via endoscopic or open approaches may be advocated. Long term follow up is necessary due to high chances of recurrence.

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