The Journal of Laryngology & Otology
January 2002, Vol. **116,** pp. 49–51

Plexiform neurofibroma of the larynx in a child

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Abstract

A case of a female child of six years of age with a plexiform neurofibroma of the larynx caused by von Recklinghausen's disease is presented. Laryngeal involvement in neurofibromatosis type 1 (NF1) is rare and only 19 paediatric cases have been reported. The tumour was biopsied and lateral pharyngotomy with supraglottic hemilaryngectomy was performed in order to relieve obstructive symptoms. Problems related to this unusual tumour localization are discussed and a review of the literature is presented.

Key words: Laryngeal Neoplasms; Neurofibromatosis 1; Neurofibroma, Plexiform; Child

Introduction

Neurofibromatosis Type 1 is characterized by multiple café-au-lait spots and neurofibromatosis as well as somesomatic abnormalities. Friedrich von Recklinghausen was the first who showed in 1882 that multiple and cutaneous fibromas existed simultaneously.[1] NF1 is one of the most common single gene disorders, occurring in approximately 1/3000 live births. It passes in an autosomal dominant fashion and affected parents have a 50 per cent chance of passing the gene to their own child. NF1 also has the highest gene mutation rate for an autosomal dominant disease, with half of reported cases being new mutations, the others being inherited.[1] Through recombinant DNA analysis, a gene deviation has been mapped on the proximal long arm of chromosome 17.[2]

Plexiform neurofibroma of the larynx in children is rare, 18 cases with laryngeal involvement were reported by different authors in the otolaryngologic literature. These cases were presented with details by Masip in 1996 with his 19th case and this is the 20th case in the literature.[3]

Case report

A six-year-old-girl was referred to our department with laryngeal obstructive symptoms. The duration of symptoms was five years and nine months and first appeared when she was three months old. The presenting symptoms were dyspnoea and coughing. These symptoms worsened when she was in a supine position especially in the evenings. Her voice was normal at rest, but a 'hot potato' voice could be heard when she strained. As can be expected, dyspnoea was increased during deep inspiration. On physical examination with a rigid telescope, a large mass was seen that originated from the left aryepiglottic fold, prolapsed medially over the glottic inlet and resulted in obstructive symptoms. The mass was pale with normal mucosa and had smooth contours. Café-au-lait spots were seen on her body but no other neurofibromas were observed. Magnetic resonance (MR) imaging demonstrated a supraglottic mass originating from the left aryepiglottic area. T2 relaxation time was prolonged and it enhanced brightly. (Figures 1 and 2).

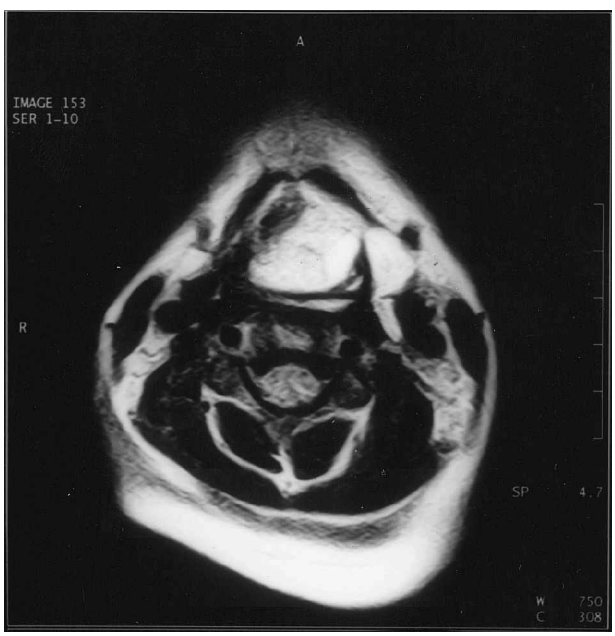

Fig. 1
Axial MR image showing the tumour originating from the left aryepiglottic fold.

Since the patient had obstructive symptoms and no histopathological diagnosis, a decision was made to debulk the lesion and take biopsy specimens. During the operation a solid mass was observed originating from left aryepiglottic fold and prolapsing into the glottic inlet. The lesion was removed endoscopically for histopathologic examination.

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Accepted for publication: 14 June 2001.

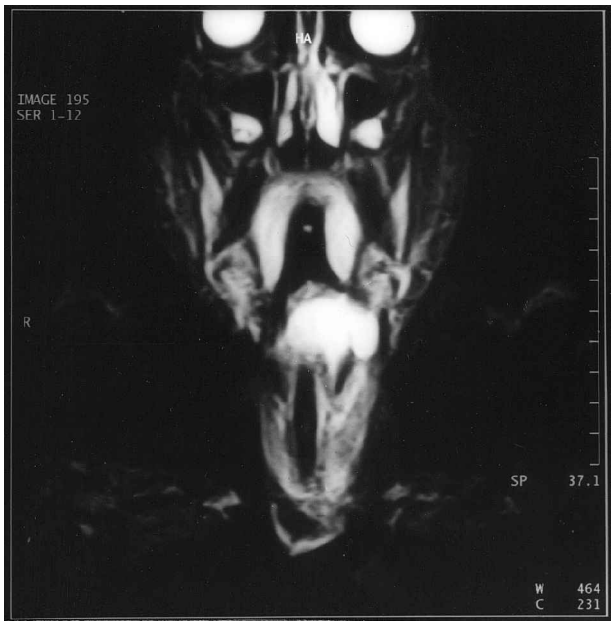

FIG. 2

A supraglottic solid mass is seen on coronal MR section.

Histopathological examination

Macroscopically the specimen was pinkish-grey covered with mucosa on one side and measuring 1.5 × 1.5 × 0.8 cm. Microscopically the surface was covered with an intact mucosa. The subepithelial tissue contained a tumour that was divided by fibrous elements to form nodules. In the nodules, uniform, spindle-shaped Schwann cells formed bundles with a loose stroma. Immunohistochemical reactions for S100 protein and vimentin proved to be positive for tumour cells (Figures 3 and 4).

After the histopathological diagnosis of plexiform neurofibroma was made a cranial MR scan was obtained and a right optic glioma and several hamartomas were seen (Figure 5). The patient underwent a lateral pharyngotomy and left supraglottic hemilaryngectomy without tracheotomy. The post-operative course was uneventful and the patient was discharged on the fifth post-operative day. She has been followed-up for two months with complete cure.

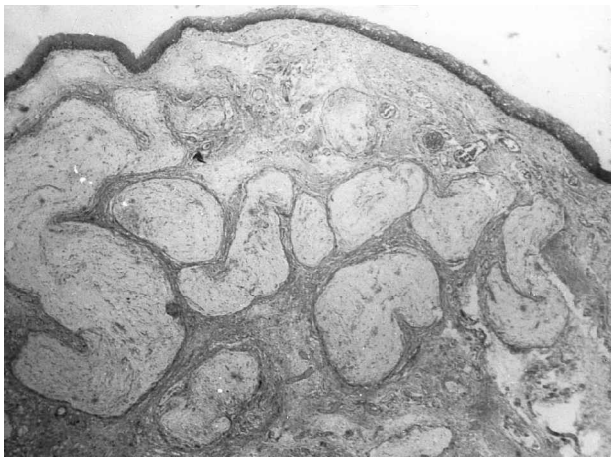

FIG. 3

An intact multilayered squamous epithelium is observed on the surface. The subepithelial zone contains a tumour separated by fibrous septa. (H & E; ×32)

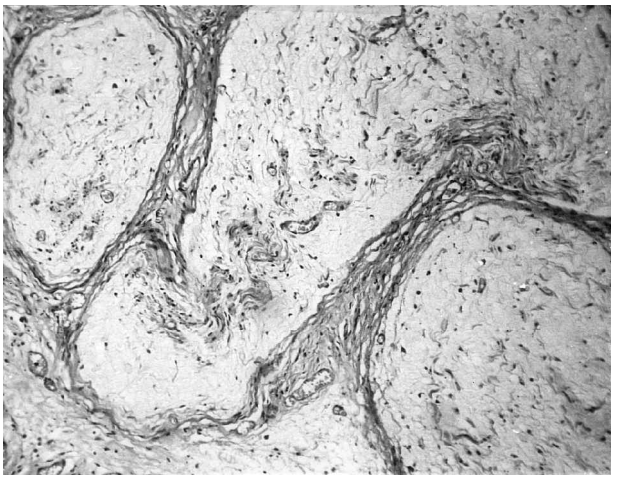

FIG. 4

With higher power magnification uniform spindle-shaped tumour cells forming bundles can be seen. (H & E; ×125)

Discussion

Benign nerve sheath tumours may be divided into neurofibromas and schwannomas. Schwannomas, previously known as neurilemmomas, arise from Schwann cells. They are usually solitary and encapsulated. On the other hand, neurofibromas are non-encapsulated and can be solitary or multiple.[4,5] Neurofibromas originate from several cells, including Schwann cells and fibroblasts. Neurogenic tumours of the larynx are rare. Laryngeal neurofibroma was first reported by Colledge in 1930.[6] Since that report only 35 cases have been reported, 19 in paediatric patients.[2]

Head and neck manifestations of neurofibromatosis have been reported by White *et al.* The most common findings in 257 NF patients were as follows: cafe-au-lait spots (95 per cent); head and neck lesions (87 per cent); positive family history (42 per cent); Lisch nodules (35 per cent); scoliosis (30 per cent). He found only one laryngeal neurofibroma in that patient group.[7]

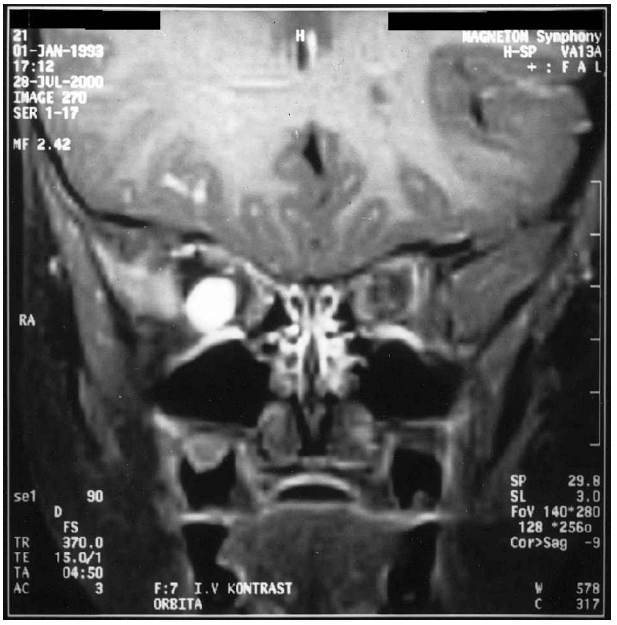

FIG. 5

Coronal MR section showing a right optic glioma.

https://doi.org/10.1258/0022215021910122 Published online by Cambridge University Press

The most common signs and symptoms in the 19 reported paediatric laryngeal cases were dyspnoea and stridor (55 per cent), dysphagia and change of voice quality (26 per cent).[2] The most common sites of involvement were the left aryepiglottic fold (seven cases), the right aryepiglottic fold (six cases) and the pyriform fossa (five cases). The tumour was located on the left aryepiglottic fold in our case as well. This location implies that the tumour originates from the superior laryngeal nerve and/or from anastomoses between the superior laryngeal nerve and the recurrent laryngeal nerve (Galen anastomoses).[8]

Because conservative treatment such as endoscopic partial resection allows further tumour growth, surgery was recommended.[9] Lateral pharyngotomy and excision or lateral pharyngotomy and supraglottic hemipharyngolaryngectomy have been recommended by several authors. Garabedian *et al*. reported successful results of seven cases that all underwent partial laryngectomy without tracheotomy.[8] Excision of laryngeal neurofibroma using the CO_2 laser has been described by Willcox but it does not seem to be an effective treatment modality because of the growth pattern of plexiform neurofibroma.[9] It may be used, however, to relieve laryngeal obstruction as a palliative measure in order to avoid total laryngectomy in large tumours.

In conclusion, if a patient with a positive family history presents with dyspnoea and change in voice quality the possibility of laryngeal neurofibroma should be considered. An MR scan should be obtained with axial and coronal sections to see the extensions of the tumour in the larynx.[10] Diagnosis is confirmed by direct laryngoscopy and biopsy. Finally, surgical resection by lateral pharyngotomy is the treatment of choice.

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Dr E. Yucel takes responsibility for the integrity of the content of the paper.
Competing interests: None declared