The Journal of Laryngology and Otology
November 1996, Vol. **110,** pp. 1065–1068

Airway obstruction in children due to plexiform neurofibroma of the larynx

HASSE EJNELL, M.D., PH.D., MIA JÄRUND, M.D.*, MARTIN BAILEY, B.SC., F.R.C.S.†,
PONTUS LINDEMAN, M.D., PH.D.

Abstract
Plexiform neurofibroma of the larynx is a rare disease. Three cases in children with airway obstruction are presented. After tumour reduction via a lateral pharyngotomy their tracheostomies could be closed. The surgical treatment of plexiform neurofibroma of the superior laryngeal nerve is discussed and conservative, subtotal resection is recommended.

Key words: Head and neck neoplasms; Neurofibroma; Larynx; Child

Introduction

In 1882 Friedrich von Recklinghausen presented a neurocutaneous syndrome which now bears his name as von Recklinghausen's disease (VRD). It is characterised by multiple café-au-lait spots and generalised neurofibromatosis as well as a variety of somatic abnormalities (Holt, 1978).

It is a predominantly hereditary, autosomal dominant disease but is also found as a spontaneous mutation (McKennan and Bard, 1991). It occurs in about 1:3000 births.

Three primarily benign tumours are associated with VRD: schwannoma, neurofibroma and plexiform neurofibroma. The plexiform neurofibroma is an unpleasant tumour due to its slow but relentless growth along nerves in a poorly circumscribed fashion, restricting the possibility of radical surgical resection without post-operative morbidity. Sarcomatous degeneration occurs in five per cent of neurofibromas (Holt, 1978).

The presence of a plexiform neurofibroma is pathognomonic of VRD. Large tumours in the neck are usually of the plexiform type and may involve cranial nerves VIII-XII as well as the brachial plexus (Lusk *et al.*, 1987) and sympathetic chain.

Plexiform neurofibroma of the larynx in children is rare with only a few cases described in the literature (O'Connor, 1980). We present three children with plexiform neurofibroma involving the superior laryngeal nerve.

Case report

Case 1

A girl born in 1986 with a history of snoring and episodes of apnoea at night underwent adenoidectomy at two years of age and tonsillectomy at four years of age. Despite these measures her symptoms persisted. At the age of seven her problems increased noticeably in severity with inspiratory stridor and globus sensation. A sleep study showed mild obstructive sleep apnoea syndrome (OSA) and spirometry indicated the presence of intermittent upper airway obstruction. A $3 \times 2 \times 1$ cm tumour was identified in the left supraglottic region involving the false vocal fold but not reaching the true vocal fold (Figure 1). The tumour involved the left posterior part of the cricoid cartilage leaving the upper oesophageal sphincter tumour free. Involvement of the lateral hypopharyngeal wall was restricted to the pharyngo-epiglottic folds. The tumour thus included the area supplied by the superior laryngeal nerve.

Histological examination showed soft tissue with multiple nerve fascicles, some of which were enlarged with oedematous endoneurium with intermixed collagen and bipolar cells consistent with the diagnosis of plexiform neurofibroma.

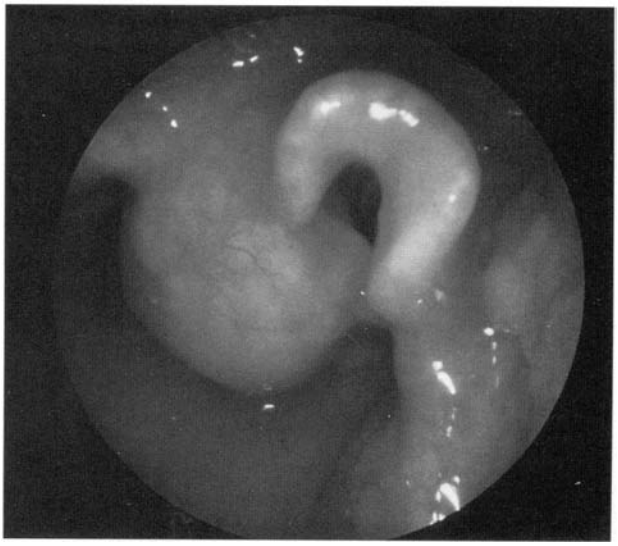

FIG. 1
Direct laryngoscopy showing the supraglottic tumour.

From the Departments of Otorhinolaryngology, Sahlgren's University Hospital*, Göteborg, Sweden, Great Ormond Street Hospital for Children†, London, UK and Mölndals Hospital, Mölndal, Sweden.
Accepted for publication: 22 July 1996.

The size of the tumour demanded excision via a lateral pharyngotomy and a peroperative tracheotomy was performed because of anticipated post-operative swelling. There was no tumour outside the entry of the internal branch of the superior laryngeal nerve into the thyro-hyoid membrane. The left half of the hyoid bone was excised and the hypopharynx opened. The tumour was dissected from the mucous membrane down to the left false fold. Similarly it was possible to remove the extralaryngeal part of the tumour without mutilation of the larynx. The mucous membrane was sutured. A week post-operatively laryngoscopy showed almost total regression of the post-operative oedema. The tracheostomy was closed the next day. Two months later direct laryngoscopy showed no tumour growth.

To this date two years post-operatively the patient has had neither breathing nor feeding difficulties. Her voice is normal.

Case 2

A girl born in 1990 arrived at the age of nine months at the casualty department with acute respiratory distress in connection with a cold. She had fever, inspiratory and expiratory stridor. Steroids and antibiotic treatment had no effect. At laryngoscopy a 1.5 × 1 cm tumour was identified in the area of the right aryepiglottic fold, arytenoid cartilage and pyriform fossa. The laryngeal inlet was hidden beneath the protruding tumour. A biopsy of the tumour, a tracheostomy to secure the airway and a computed tomography (CT) scan were done.

A thorough history revealed that the girl had had noisy breathing since birth, thought to be due to laryngomalacia. Physical examination showed multiple café-au-lait spots as well as Lisch noduli and hamartomas of the iris, consistent with VRD. Histological examination of the tumour showed a plexiform neurofibroma. During the next four years the tumour was debulked by frequent treatments with the CO_2 laser but decannulation was not possible. Repeated laryngoscopies as well as magnetic resonance imaging (MRI) showed an increase in tumour size paralleling the growth of the child (Figures 2 and 3).

At the age of five she still depended on the tracheostomy for breathing, and had difficulty swallowing solid foods. The decision to excise the tumour via a lateral pharyngotomy was taken. This child had tumour enveloping the vagus nerve throughout the right side of the neck. However, there were no symptoms due to this involvement and the vagus nerve was left intact peroperatively. A large tumour was located and excised anterior to the thyro-hyoid membrane in the area of the superior laryngeal nerve along with the hypopharyngeal and laryngeal tumour. The latter part of the tumour was excised in the same fashion as in *Case 1* and measured 5 × 3 × 2 cm. About 90 per cent of the tumour could be excised. This girl was successfully decannulated four weeks post-operatively. She has, a year later, no breathing nor swallowing difficulties and her voice is normal.

Case 3

A girl born in 1980 was transferred to our hospital at the age of five months with a history of increasing stridor and feeding difficulties from three weeks of age, requiring

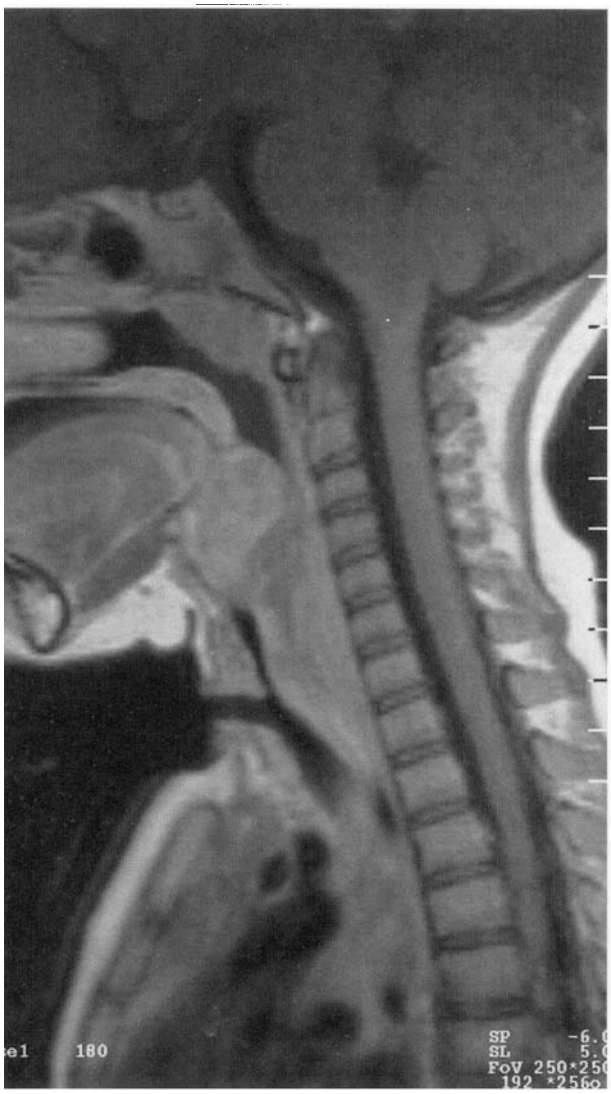

FIG. 2
MRI, a sagittal view of the supraglottic tumour.

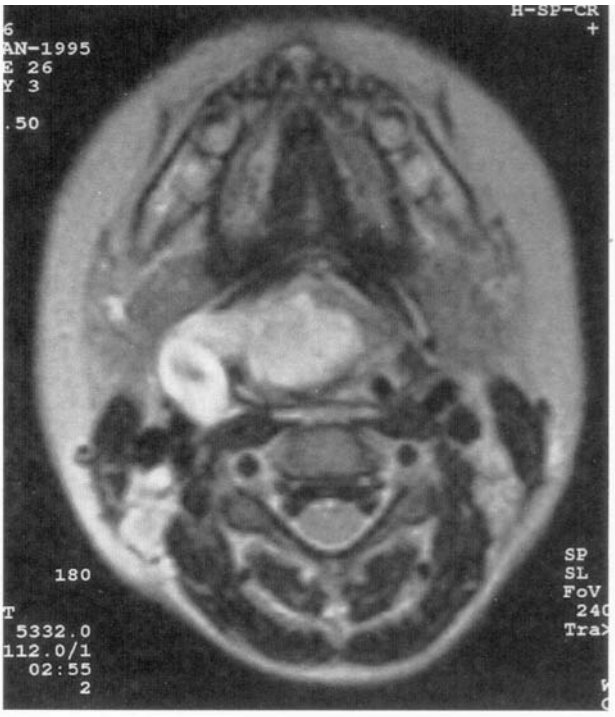

FIG. 3
MRI, a transverse section showing the supraglottic tumour.

https://doi.org/10.1017/S0022215100135765 Published online by Cambridge University Press

several admissions to the local hospital. Intubation shortly after arrival proved difficult due to a large supraglottic swelling, and a tracheostomy was performed later the same day. Subsequent microlaryngoscopy revealed a large, soft, apparently cystic right-sided supraglottic mass distorting the larynx and largely occluding the airway: this was thought at the time to be a lymphangioma (no biopsy was taken) and it was planned to defer excision via a lateral pharyngotomy until two years of age.

The child subsequently developed a localised swelling in the floor of the mouth anteriorly on the right side, displacing the tongue, and at the age of two years and five months this was excised.

The specimen proved to be a plexiform neurofibroma, and subsequent endoscopic biopsy of the laryngeal mass showed the same histology. Cutaneous café-au-lait patches were noted and a diagnosis of neurofibromatosis was confirmed.

At the age of two years and nine months a right lateral pharyngotomy was undertaken, and a large neurofibroma mass was sub-totally excised from the pre-epiglottic space on the right side and from within the right aryepiglottic fold. Following this procedure her swallowing dramatically improved, but residual swelling of the aryepiglottic folds and interarytenoid region persisted and the airway remained obstructed.

Four years later she developed some discomfort in the throat with a sensation of food 'sticking', and endoscopy revealed resolution of the aryepiglottic fold swelling but enlargement of the mass in the posterior wall of the larynx. This was then endoscopically excised down to just above the cricopharyngeus using the CO_2 laser.

Following this procedure the larynx appeared virtually normal, with just some bulging of the right false fold remaining, and decannulation was finally achieved at the age of nine years. Two years later an asymptomatic right parapharyngeal swelling was noticed: this enlarged a little over the ensuing three years with the development of an uncomfortable neck lump deep to the pharyngotomy scar. MRI scanning showed a 4×3 cm parapharyngeal mass extending up to the skull base, surrounding the internal carotid artery and internal jugular vein. The superficial part of this mass was excised and confirmed histologically to be a neurofibroma, but no attempt was made to remove the deep, parapharyngeal component in view of the inevitable morbidity involved.The patient remains well at the age of 15 years.

Discussion

The children in our report all presented with breathing difficulties in the early years of their lives as the first symptom and dysphagia at a later stage. *Case 1* lacks family history of VRD and may therefore be due to spontaneous mutation.

After a lateral pharyngotomy with excision of most of the tumour the children could return to a normal life without feeding difficulties and without a tracheostomy. None had post-operative neurological deficits. Conservative treatment only permits further tumour growth and surgery must be recommended although the degree of tumour resection can be debated. Successful excision of neurofibroma of the larynx with the CO_2 laser has been described (Willcox *et al.*, 1993), but this does not seem to be an effective treatment when it comes to plexiform neurofibroma due to its growth pattern. Is it crucial to remove all of the tumour at all costs (leaving the patient with large neurological deficits) or is it sufficient to surgically remove the symptomatic part of the tumour, with repeated surgery if symptoms recur? We feel that the latter is the method of choice in view of the tendency of the tumour to recur due to microinvasion of involved nerves and with regard to the quality of life for the patient. To minimise the negative effect on growth, speech and psychological functions as well as the post-operative morbidity, it is important that the tumour is treated early.

Head and neck neurofibromatosis may involve the petrous temporal bone, scalp, oral cavity, orbit and more rarely the larynx. Nerves affected may be the facial, auditory, glossopharyngeal, hypoglossal or vagus (Galli *et al.*, 1992) as well as the sympathetic chain, cervical plexus or brachial plexus (Decurtin *et al.*, 1988). In our cases the superior laryngeal nerve as well as the vagus nerve was involved and only a few similar cases have previously been reported (Chang-Lo, 1977; Czigner and Fekete-Szabó, 1994).

The superior laryngeal nerve, a branch of the vagus nerve passes from the inferior ganglion and divides into a motor (external) and a sensory (internal) branch. The external laryngeal nerve supplies the cricothyroid and cricopharyngeus muscles. The internal laryngeal nerve passes through the thyrohyoid membrane and innervates the mucous membrane of the pyriform fossa and upper part of the larynx.

All three cases had tumour growth corresponding primarily to the sensory branch of the superior laryngeal nerve. *Cases 2* and *3* also had vagus involvement. There is a marked difference between symptoms when only the vagus nerve is affected and when its branch the superior laryngeal nerve is involved. The former usually lacks symptoms except for a lateral cervical mass (Bocca, 1984), whereas the latter has a tendency to cause breathing problems, dysphagia and globus sensation.

Due to the slow growth rate of the tumour the diagnosis is often delayed and the tumour often reaches an astonishing size leading to difficulties at the time of surgical intervention. Often the children need a covering tracheostomy for the procedure. In our cases the children have done well without the tracheostomy after tumour excision via a lateral pharyngotomy.

Our first case shows the importance of considering benign tumours of the head and neck region in the differential diagnosis when confronted with a child with abnormal breathing patterns such as OSA despite having undergone adenoidectomy and tonsillectomy. The second case with breathing problems since birth stresses the importance of actively excluding other causes before accepting a diagnosis of laryngomalacia. Admittedly, however, plexiform neurofibroma of the larynx is extremely rare.

References

Bocca, E. (1984) Tumori parafaringei. *Tumefazioni cervicali:* 299–318.

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

Czigner, J., Fekete-Szabó, G. (1994) Neurofibroma of the supraglottic larynx in childhood. *Journal of Laryngology and Otology* **108:** 156–158.

Decurtin, C., Wey, W., Moll, C. (1988) Schwannoma and neurofibroma of the neck. *HNO* **36:** 437–444.

Galli, J., Almadori, G., Paludetti, G., Rosignoli, M., Corina, L., Ieraci, A. (1992) Plexiform neurofibroma of the cervical portion of the vagus nerve. *Journal of Laryngology and Otology* **106:** 643–648.

Holt, G. R. (1978) ENT manifestations of von Recklinghausen's disease. *Laryngoscope* **88:** 1627–1632.

Lusk, M. D., Kline, D. G., Garcia, C. A. (1987) Tumours of the brachial plexus. *Neurosurgery* **21:** 439–453.

McKennan, M. D., Bard, M. D. (1991) Neurofibromatosis type 2. *Laryngoscope* **101:** 109–113.
O'Connor, A. F. (1980) Neonatal laryngeal neurofibromatosis. *Ear, Nose and Throat Journal* **59:** 174–177.
Willcox, T. O., Rosenberg, S. I., Handler, S. D. (1993) Laryngeal involvement in neurofibromatous. *Ear, Nose and Throat Journal* **72:** 811–815.

Address for correspondence:
Hasse Ejnell, M.D.,
Department of Otorhinolaryngology,
Sahlgren's University Hospital,
S-413 45 Göteborg,
Sweden.