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Neurofibroma of the supraglottic larynx in childhood

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Abstract

The case of a male child with a benign neurogenic laryngeal tumour caused by von Recklinghausen's disease is presented. At the age of eight years, tracheostomy was necessary. The tumour, deforming the right half of the larynx and extending into the right half of the neck, was removed three years later. Following this procedure, decannulation was possible, and the patient has been free of laryngological complaints for six years.

Attention is drawn to the fact that autosomal dominantly inherited neurofibromatosis is a generalized benign tumour with a special form. If it involves any vital structure or forms a functional obstacle, meticulous surgical removal is necessary, and this provides good prospects over a long period.

Key words: Neurofibromatosis; Laryngeal neoplasms; Surgery

Introduction

Neurofibromatosis is an inherited autosomal dominant condition. Developments in diagnostic and molecular genetic research have led to the solution of problems in terminology (Barker, 1987; Seizinger and Roullau, 1987).

In 1987, two types of neurofibromatosis were differentiated (McKennan and Bard, 1991). Neurofibromatosis type 1 has an incidence of 1:4000 live births. It was previously known as von Recklinghausen's neurofibromatosis or peripheral neurofibromatosis. According to the brief historical review by Kimmelman (1979), the disease was first described by Tilesius in 1793. Its hereditary character was indicated by Virchow in 1847 and it was first called neurofibromatosis by von Recklinghausen in 1882. Multiple cutaneous and subcutaneous nodules that increase in number with age are situated along dermal nerves and neural plexus branches (Chang-Lo, 1977). Through recombinant DNA studies, a gene deviation has been mapped on chromosome 17 (McKennan and Bard, 1991).

Neurofibromatosis type 2, with an incidence of 1:50 000, is called central neurofibromatosis. Diagnostic criteria include acoustic neurofibromatosis, or two of the following: neurofibroma, meningioma, glioma, schwannoma, and juvenile superior subscapular lenticular opacity. Molecular genetic studies revealed a gene deviation near the centre of the long arm of chromosome 22.

The two types of neurofibromatosis may occur together, with a negative family history, and the condition can arise after a spontaneous mutation (McKennan and Bard, 1991).

Case report

Earlier reports on this male child (born in 1975) revealed that he had suffered from von Recklinghausen's disease for several years: hoarseness started at the age of six years. He was examined in three different hospitals, but no operation was performed.

He was admitted to our clinic in 1983 with increasing inspiratory dyspnoea, and emergency tracheotomy was performed. A longish, plum-like, firm lump could be felt on the right side of the neck, under the sternocleidomastoid muscle. In the right piriform recess and in the arytenoid region, a smooth-surfaced, 2 cm tumour was seen, obstructing the laryngeal inlet. In May 1983, medial pharyngotomy and partial extirpation of the right aryepiglottic fold were performed under general anaesthesia. Nevertheless, this did not permit decannulation. The boy was discharged with an indwelling, size 4, tracheal cannula.

When he was readmitted, in June 1986, he had been wearing his cannula for three years. He had the associated developmental abnormalities of pectus excavatum and scoliosis convex to the right. The typical 'cafe-au-lait' spots of von Recklinghausen's disease were widespread and most marked on the back (Figure 1). The right arytenoid and aryepiglottic regions were expanded with a smooth tumour narrowing the laryngeal airway. Movement of the right half of the larynx was limited. There was a 5 cm swelling deep into the right sternocleidomastoid muscle, which was not attached to the skin.

Surgery was indicated: 0.5–1 cm compact, white nodules which were lighter in colour than the fatty tissue, and which were indicative of von Recklinghausen's disease, were removed together with the involved sternocleidomastoid muscle by a functional right neck dissection-like approach (Figure 2). Almost all nerve fibres in the region were involved. The laryngeal space-occupying tumorous lesion compressing the piriform fossa and the aryepiglottic fold from the right side was removed separately, prepared along the thyroid cartilage and the hyothyroid membrane. The histological finding was the same on every occasion i.e. neurofibromatosis. Five weeks after the operation, decannulation and closure of the stoma were possible. Since then the patient has been reviewed at regular intervals. He has been tumour-free for six years. His breathing is free.

Starting from the child, a family tree showing the heredity of the illness was prepared (Figure 3). His sibling, mother, grandfather and great-grandmother (on the maternal side) suffer from von Recklinghausen's disease.

Discussion

Von Recklinghausen's disease is cited most often as the cause of bilateral acoustic neurinoma. Its familial incidence has been

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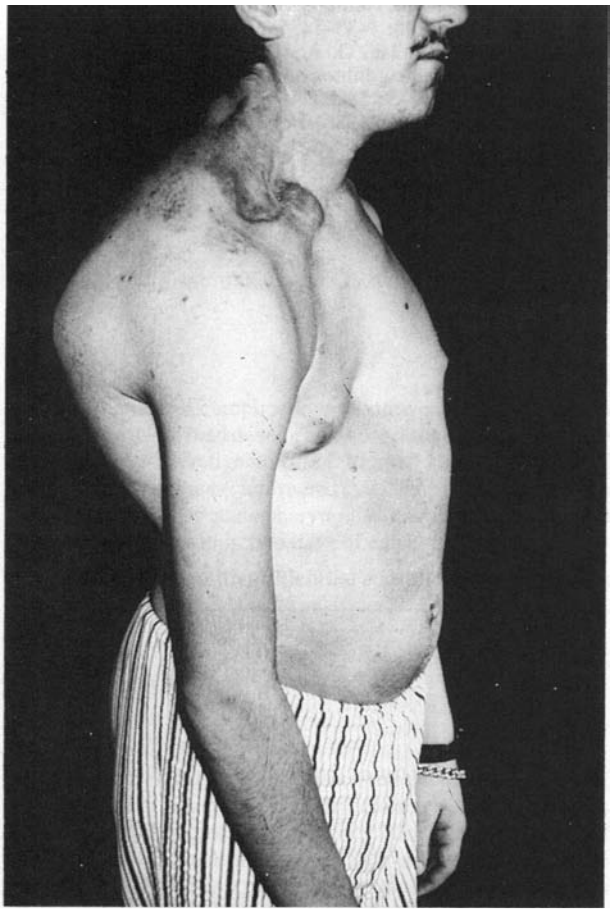

FIG. 1

Photograph showing the kyphoscoliosis and pectus and excavatum of the patient.

described (McKennan and Bard, 1991). Characteristics include large (more than six) 'café-au-lait' maculae (in 95 per cent of the cases) or a plexiform neurofibroma, Lisch nodules (iris hamartomas), and optic glioma. It is often accompanied by kyphoscoliosis, macrocephaly, broadening of the sella turcica, dysarthrosis, hyperostosis, hypertrichosis and vessel naevi (Holt, 1978; Kimmelman, 1979; White and Bigler, 1986).

Numerous otorhinolaryngological manifestations have been reported (Holt, 1978; White and Bigler, 1986). Two cases have been described with Horner's syndrome derived from neurofibromatosis starting from the cervical sympathetic plexus (Daly

FIG. 2

Operative removal of the tumour: (a) isolated tumour; (b) right internal jugular vein; (c) the extremely enlarged right vagal nerve; (d) right common carotid artery.

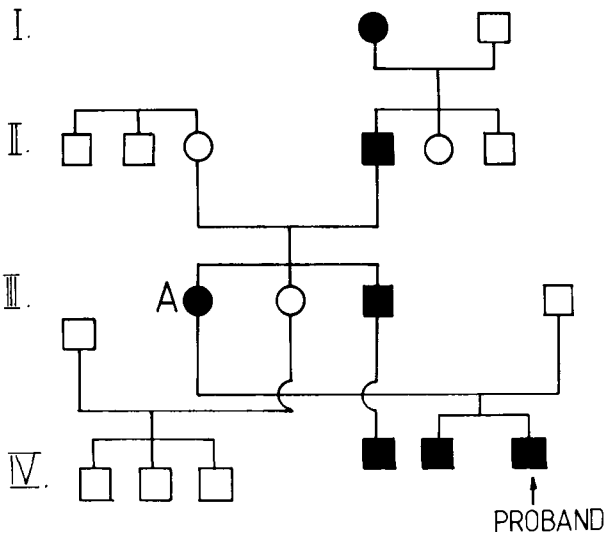

FIG. 3

Pedigree: the dominant heredity of the disease can readily be followed through the generations. □, males; ○, females. ■, males with disease; ●, females with disease.

and Roesler, 1963; Ruckley, 1986), and one case with Horner's syndrome caused by a solitary pharyngeal tumour. The tumour is derived from the connective tissue cells of the endo- and perineurium (Daly and Roesler, 1963). Fibroblast-like cells and nerve fibres can also be found in it, as residues of the original tissue. Malignant transformation has been reported in 30 per cent of all cases. Histologically, these malignancies prove to be schwannomas and fibrosarcomas. Neurofibroma is very rare in the larynx. Chang-Lo (1977) surveyed the 19 laryngeal neurofibromas reported up to that time. Kimmelman (1979) emphasized that neurofibromatosis occurs in the larynx on the recurrent laryngeal nerve and is generally initiated submucosally on the aryepiglottic plica (Kimmelman, 1979). Although, a solitary recurrent nerve neurofibroma has been reported earlier (Rees, 1971) the supraglottic neoplasm is the most frequent in childhood (Karja, 1982; White and Bigler, 1986) manifesting as a space-occupying process causing suffocation as in our case.

In accordance with the examination by White and Bigler (1986) our family tree follow-up revealed autosomal dominant familial inheritance. It was especially interesting that locally multiple cervical neurofibromatosis predominated in this child with generalized von Recklinghausen's disease. Involvement of the vagal nerve, which was extremely thickened was evident.

Although the recurrent laryngeal nerve was not explored, the space-occupying laryngeal tumour corresponds in localization to the level of the anastomosis between the recurrent and the superior laryngeal nerves (Galen Ansa). It must therefore have originated from these nerves.

There is no chance for either conservative or surgical causal treatment. Debate continues as to the role and place of the two methods in the management of the disease. This case of generalized von Recklinghausen's disease manifested multiple cervical lesions. As a space-occupying laryngeal tumour threatening asphyxia, it became an urgent surgical problem. It is our view that the tumour must be removed if it involves vital structures, forms a functional obstacle, commences rapid growth or causes unbearable pain. Surgical removal of the neurofibroma, which may give good results over a long time, is the only possibility for symptomatic treatment of the illness. In a lucky case, the tumour will not recur.

References

Barker, V. (1987) Gene for von Recklinghausen's neurofibromatosis is in the pericentromeric region on chromosome 17. *Science* **236:** 1100–1102.

Chang-Lo M. (1977) Laryngeal involvement in von Recklinghausen's disease: a case report and review of the literature. *Laryngoscope* **87:** 435–442.
Daly, J. F., Roesler, H. G. (1963) Neurilemmoma of the cervical sympathetic chain. *Archives of Otolaryngology* **77:** 262–267.
Holt, R. G. (1978) ENT manifestations of von Recklinghausen's disease. *Laryngoscope* **88:** 1627–1632.
Karja, J. (1982) Benign neoplastic tumours and space-occupying tumorous lesions of the larynx in children. *Acta Otolaryngology* **386 (Suppl.):** 181–183.
Kimmelman, Ch. P. (1979) Otolaryngologic aspects of neurofibromatosis. *Archives of Otolaryngology* **105:** 732–736.
McKennan, M. D., Bard, M. D. (1991) Neurofibromatosis type 2. *Laryngoscope* **101:** 109–113.
Rees, G. (1971) Neurofibroma of the recurrent laryngeal nerve. *Chest* **60:** 414–418.
Ruckley, R. W. (1986) Neurofibroma of the cervical sympathetic chain presenting with Horner's syndrome. *Journal of Laryngology and Otology* **100:** 1433–1436.
Seizinger, B. R., Roullau, G. A. (1987) Genetic linkage of von Rechlinghausen's neurofibromatosis to the nerve growth factor receptor gene. *Cell* **49:** 589–594.
White, K., Bigler, C. R. (1986) Head and neck manifestations of neurofibromatosis. *Laryngoscope* **96:** 732–737.

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