Neurofibroma of the Recurrent Laryngeal Nerve*

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A case of von Recklinghausen's disease is described in which involvement of the recurrent laryngeal nerve by a neurofibroma at the level of the aortic arch caused dysarthria. The tumor was successfully removed at thoracotomy. The literature dealing with this extremely rare occurrence is reviewed.

Abnormalities of the brain, spinal cord, meninges and peripheral nerves commonly occur in von Recklinghausen's disease. These may take the form of benign or malignant tumors, although it is suggested that most of the benign lesions are in fact developmental abnormalities or hamartomas. Intrathoracic lesions of neurogenic origin, neurofibromas and neurolemmomas, occur almost exclusively in the posterior mediastinum and usually arise from intercostal nerves or their branches. Tumors of the intrathoracic vagus nerve are rare in contrast with the much more frequent involvement of cervical vagus. The first reported case of the former was by Blades and Dougan (1943) and has been followed by other reports. An extremely unusual occurrence is involvement of the recurrent laryngeal nerve causing hoarseness. A patient with von Recklinghausen's disease is described in whom a neurofibroma involving the left recurrent laryngeal nerve caused paralysis of the left vocal cord.

Case Report

In 1963 a 32-year-old man developed multiple raised cutaneous lesions, biopsy of which confirmed a diagnosis of neurofibromatosis. The family history was entirely negative and a chest radiograph taken at this time was normal. Four years later he was investigated for headaches, but no cause was found. In May, 1969, he developed hoarseness and an unproductive cough and he was again referred for investigation. Clinical examination revealed multiple skin nodules up to two centimeters in diameter with numerous café-au-lait patches. Apart from a hoarse voice there were no detectable neurologic abnormalities. His cardiovascular system and respiratory system showed no abnormal signs apart from a pectus malformation of the lower sternum.

Routine hematologic and biochemical analyses were normal. Chest radiographs showed a rounded projection on the left side of the superior mediastinum, adjacent to the aortic arch (Fig 1, 2). In view of the patients' clinical history an intrathoracic neurofibroma was diagnosed but an aortic aneurysm could not be excluded. Tomography supported this view and aortography showed a normal aortic arch. At thoracotomy, after confirmation of paralysis of the left vocal cord by laryngoscopy, an oval tumor was found lying on the...
aortic arch in continuity with the vagus nerve. The vagus nerve and its recurrent branch emerged from the lower end of the tumor. The tumor was removed by sacrificing the involved length of the vagus nerve.

Histologic examination confirmed an $8 \times 6 \times 6$ cm tumor which microscopically showed nerve fibers passing through interlacing strands of connective tissue. Connective tissue overgrowth was present at the upper and lower cut ends of the vagus nerve. No palisading, hyalinization or cuffing of vessels was present and there was no evidence of malignancy. Histologically the lesion was a neurofibroma and a skin nodule removed at the same time showed the same pathologic features.

The patient remains well and his voice is unchanged.

**Discussion**

Histologic differentiation between neurofibromas and neurilemmomas ("Schwannomas") is said to be easier in peripheral than in intrathoracic nerves. Neurofibromas are not encapsulated, with traversing nerve fibers and are expanded by endoneurial, perineurial or Schwann cell components.

Separation into benign and malignant forms may be extremely difficult and it has been suggested that previously described criteria of malignancy would not now be accepted. It is generally agreed, however, that sarcomatous changes may occur. Neurogenic tumors of the posterior mediastinum are more likely to be malignant if other features of von Recklinghausen’s disease are present. In a series of 48 patients with intrathoracic neurogenic tumors reported by Ackermann and Taylor in 1951, four had von Recklinghausen’s disease. Three of these had malignant Schwannomas and one a neurofibroma. Hoarseness in the presence of suspected intrathoracic neurogenic tumors does not of itself indicate malignancy. Vocal cord paresis from pressure on the recurrent laryngeal nerve caused by bleeding into a benign neurogenic tumor has been described. It would seem, however, that in view of the danger of malignant change in the presence of von Recklinghausen’s disease, surgical removal is indicated and provided it is unilateral, little functional impairment occurs, although bilateral removal may cause laryngeal, cardiovascular and gastrointestinal complications.

The presence of a pectus malformation of the chest which also occurred in a case reported by Gerbode and Margules (1953) caused no technical difficulties at operation and required no treatment.
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Perforation of the Aorta as a Complication of Mediastinal Sepsis*

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An unusual form of perforation of the aorta following mediastinal infection is recorded. A fishbone perforation of the esophagus produced a mediastinal abscess and the local sepsis led to weakening and necrosis of the aortic wall with fatal secondary hemorrhage. This report stresses the need for urgent prophylactic therapy in patients who have swallowed a foreign body and especially with the onset of mediastinitis.

CASE REPORT

A 42-year-old Indian man was admitted with fever and chest pain of four days’ duration. He had swallowed a fishbone eight days earlier. On examination, his temperature was 38.5°C. There was no surgical emphysema in the neck.

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