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CASE REPORT

Neurofibroma of the Vagus Nerve in the Cervical Portion

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Abstract: *Objective:* Neurofibromas of the vagus nerve on the neck are very rare. They are asymptomatic, slowly growing. We have seen only one case of neurofibroma of the vagus nerve in the mentioned location during last 22 years.
Clinical presentation: 33-years old patient with negative family history. She has observed increasing swelling on the right side of the neck for about 2 years. She complained of the disorders of swallowing, expectoration, aspiration episodes, stridor, intermittent palpitations, breathlessness, frequent airway infections. Magnetic resonance (MR) and angiography (AG) showed expansion in the mentioned location.
Conclusion: We present our experiences with neurosurgical management of neurofibroma of the vagus nerve in the cervical portion using microscopic technique. We found only 9 described cases of surgical treatment of the neurofibroma of the vagus nerve in the neck location in available literature till 2007 *(Fig. 6, Ref. 9)*. Full Text (Free, PDF) *www.bmj.sk*.
Key words: neurofibroma, nervus vagus.

Diagnosis

A visible, well-bounded palpable expansion with a dimension of 11.5 x 5 cm was observed on the right side of the neck. We have not detected coffee spots on the skin. Electromyographic examination showed paresis of plicae vocalis L.dx. Carotid angiography excluded a tumour of carotid wall (Fig. 1). We observe low T1 signal (Fig. 2) and high T2 (Fig. 3) on MRI. MR showed a deviation of the common and internal carotic arteries and of internal jugular vein ventromedially. According to MR the tumour outgrew the vagus lumen. Oppression of the carotic artery was more evident on coronal scans while ventromedial compression of internal jugular vein on axial ones. Histological examination (Fig. 4) showed that the typical features of neurofibromatic tumour are spiral cells and highly collagenised stroma.

Resection

The patient has been informed before the surgery on the nature of surgical solution and on potential complications.

The neck was in horizontal position during the surgery. M. sternocleidomastoideus was transferred out of longitudinal section laterally and the tumour was uncovered consequently. Preparation and surgical removal of the tumour were done with caution due to intimate relation of the nerve to the adjacent structures. The tumour was whitish-brown, with smooth surface and rubber consistence and was ingrown in the cicatricial fibrotic tissue. The expansion has spread approximately 1 cm from cranial base below the carotid bifurcation and enlarged lymphatic nodes were present. We have been clarified n. hypoglossus and cut out m. digastricus. Following the vessel identification the tumour of the vagus nerve was gradually separated and identified under the microscope. Virtually the whole lumen was infiltrated. Removed neurofibroma (Fig. 5) had dimensions of 10 x 4 x 1.4 cm. Both stubs were treated by reconstructive surgery.

Removal of neurofibroma of the vagus nerve using internal decompression is considered by some authors (1, 2, 3) as more effective. Capsula preservation ensures prevention of the neck structure damage.

Although under the microscope, a blunt preparation is dangerous.

Temporary bradycardia and hypotension were observed during the preparation in the vicinity of the bifurcation. Right vocal cord paralysis persisted in the postsurgical period.

The patient has been discharged after 10 days; after 8 months she indicates regression stridor, dysphagia and breathlessness. She does not indicate weight loss; pulmonary complications; EMG showed persisting right vocal cord paralysis.

Discussion

Neurofibromas of the vagus nerve on the neck are extremely rare (2, 4, 5). They belong to benign tumours of nerve sheaths. Generally they are incorporated in the tumour and have fusiform

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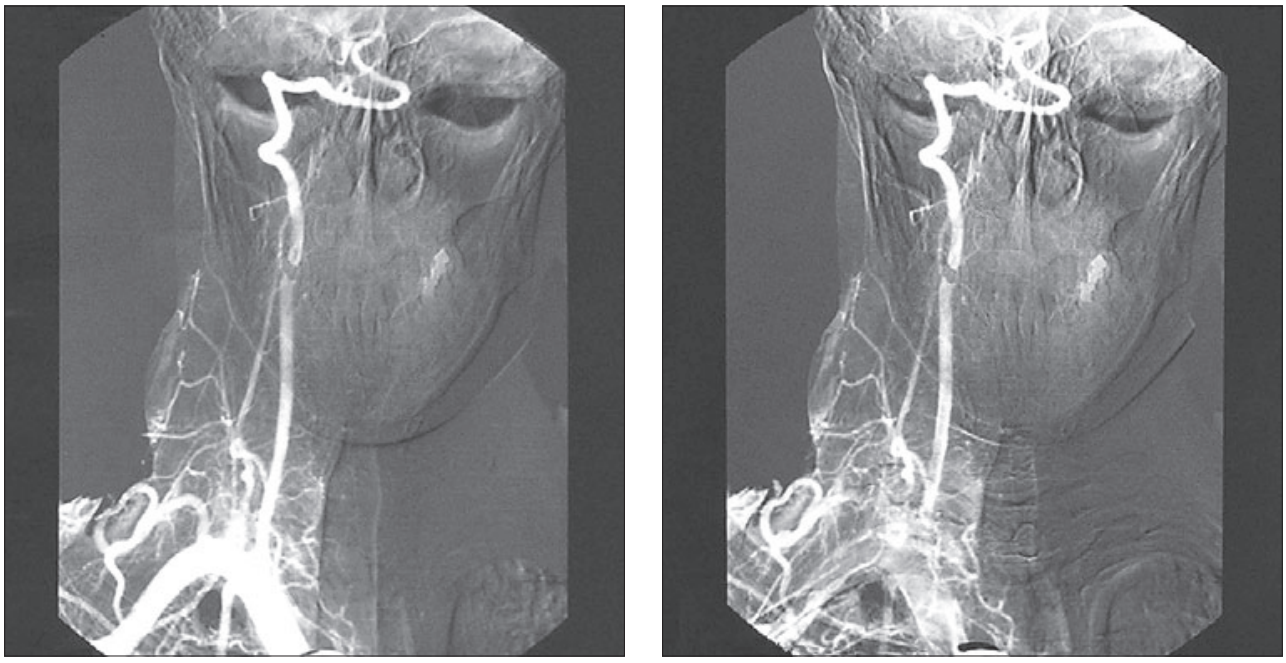

Fig. 1. Comon carotid angiogram, location of the tumor in relation to the carotid vessels.

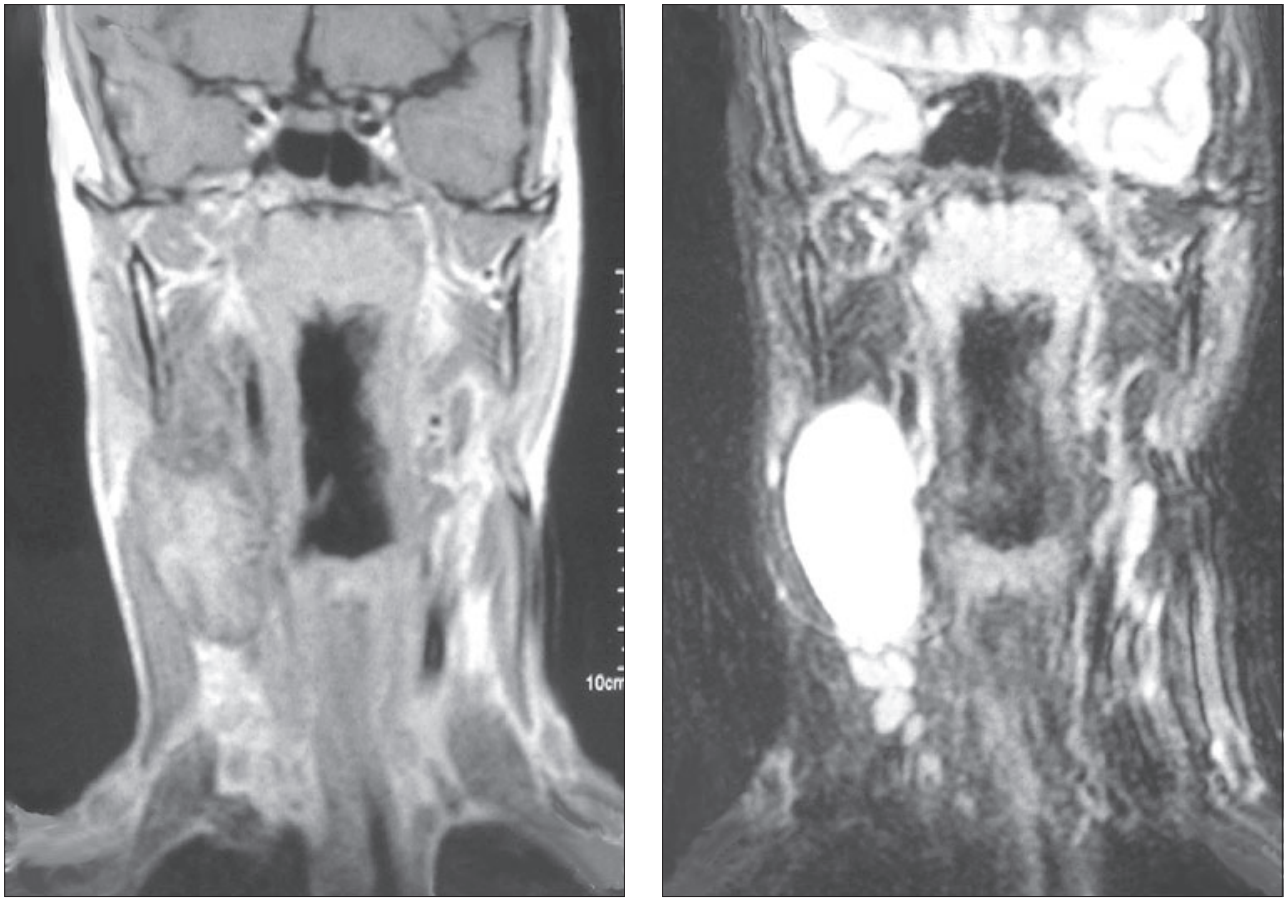

Fig. 2. Sagittal view of the patients MRI scan, showing location of the tumor.

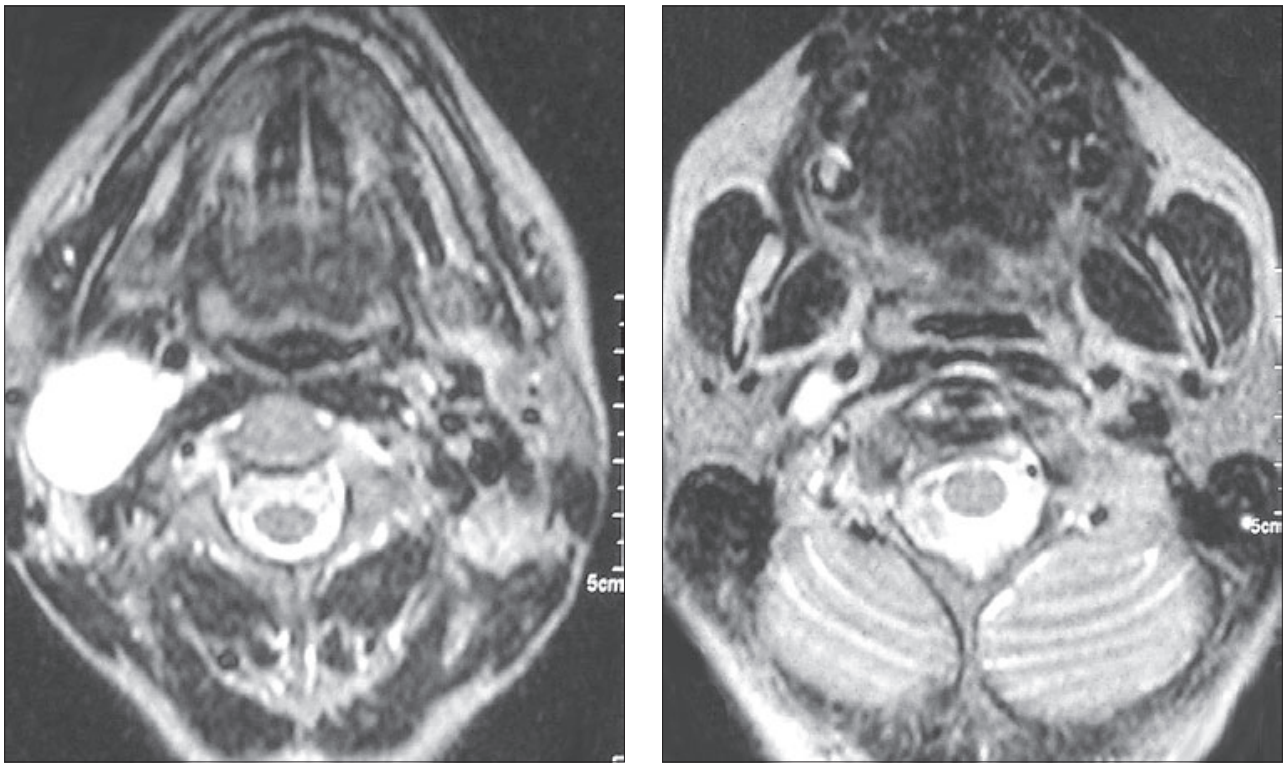

Fig. 3. MR axial T1 and T2 weighted MRI scan.

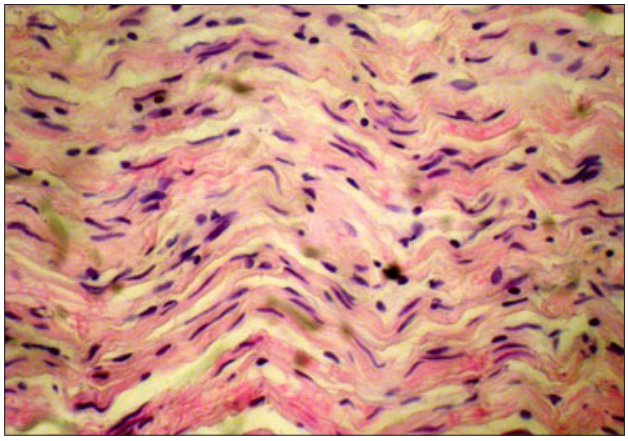

Fig. 4. Typical features (characteristics) of a neurofibroma.

configuration as if the nerve was coming out of the nerve trunk (3, 6, 7). Neurofibromas of the vagus nerve are usually avascular, without an own capsule. They are surrounded by a capsule which is an elongation of the neurolemma. They grow slowly, are painless and cause no clinical symptoms for a long period and therefore they are observed at first by ENT doctors (otorhinolaryngologists) and general surgeons, however neurosurgeons must recognize and treat them (3). Neurofibromas of the vagus nerve occur most frequently between 20–40 years of age in equal proportion between men and women.

Probable neurofibroma precursor – perineural fibroblast – comes from embryonic more primitive neuroectodermal cells than Schwann,s cells and their precursors. Their origin upgrows much more frequently from motoric fascicle or fascicles (3). Hence the neurofibroma removal is much more difficult than that of schwannomas (5).

In the case of small neurofibromas operated early in their development there are fewer fascicles outgrown and dissection is easier in comparison with large and old lesions where the whole nerve lumen may be outgrown in certain range.

Although they are generally benign, there were observed also malignant cases (5, 8, 9).

They often cause vocal cord plegia. Diagnostic gold standard is MR examination (1). Biopsy is contraindicated because of bleeding, adhesions, enclosed tissue layers and consequently difficult preparation of the surgery. The patient was after the surgery regularly followed by a neurosurgeon, ENT and neurologist. Currently, 12 months after the surgery, she indicates regression of stridor (despite EMG results), swallowing and breathlessness disorders. Stridor is persistent. Control MRI (Fig. 6) did not show any residual tissue, or tumour relapse. Electromyography examinations show persistence of paresis of plicae vocalis L.dx.

Conclusion

Neurofibromas are relatively radiosensitive. Their removal is indicated mainly in early stages. Neurofibromas of the vagus nerve

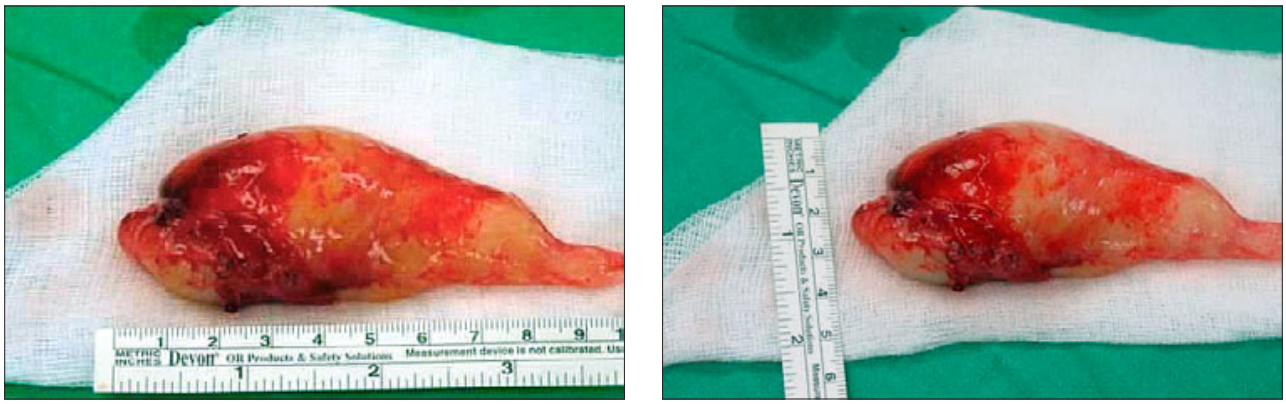

Fig. 5. Macroscopic view of the tumor, maximal length 10 cm, width 4 cm.

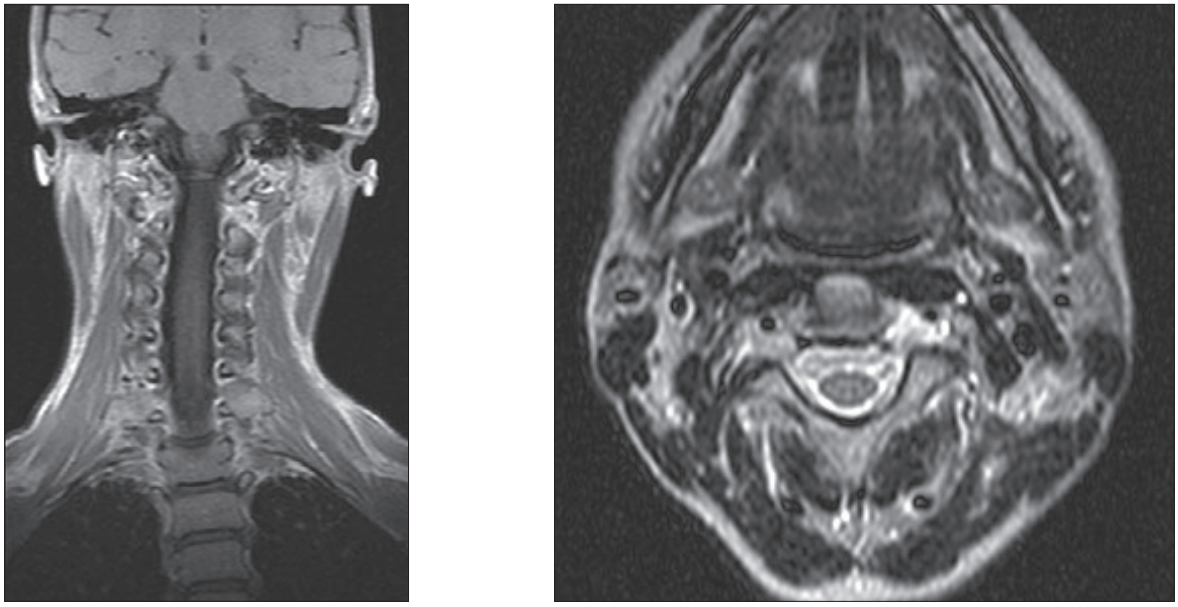

Fig. 6. MRI of the neck after operation. Condition after the vagus nerve neurofibroma extirpation reg colli 1.dx.

on the neck are rare. Epidemiologic characteristics, presentation, differential diagnostics and treatment are discussed. For postsurgical period, neurosurgical, neurologic and ENT monitoring is necessary.

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