

Delayed Diagnosis

Glossopharyngeal schwannoma presenting as gagging dysphagia

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Summary: A case of cervical glossopharyngeal schwannoma presenting with gagging dysphagia is presented. Treatment was by total excision of the schwannoma following which the patient made a good recovery. To our knowledge, this is the first report of gagging dysphagia associated with a glossopharyngeal schwannoma.

Introduction

Schwannomas are benign tumours arising from the Schwann cells of nerve sheath. Glossopharyngeal schwannomas are rare entities and can arise from either the intracranial or extracranial portion of the cranial nerve. Extracranial schwannomas usually present as a painless parapharyngeal space mass. We describe a case of glossopharyngeal schwannoma in a man with von Recklinghausen's disease who presented with an exaggerated gag reflex.

Case report

A 25 year old man presented in 1984 with a history of gagging dysphagia. He complained of pain over the right side of the neck associated with swallowing. Palpation over the right anterior triangle of the neck caused him to gag.

In childhood and early adulthood, he had operations beginning with thoracic spinal fusion for scoliosis at age 11 years followed by operations in the neck to remove neurofibromas, leading to the diagnosis of von Recklinghausen's disease. On examination, he had café-au-lait spots and subcutaneous neurofibromata, but no neurological deficit. The patient had a computed tomographic (CT) scan of the neck and a barium swallow. Neither investigation showed any abnormality. Various procedures were tried unsuccessfully over the next few years to control his symptoms, including local anaesthetic injection to the right cervical plexus.

A repeat CT scan of the neck in 1988 showed a suspicious low-density lesion within the right carotid sheath, at the level of the carotid bifurcation. The lesion did not show evidence of vascularity on carotid arteriogram. Exploration of the right side of the neck was undertaken in 1988. The carotid sinus, internal and external carotid arteries and internal jugular vein were explored, but the only abnormality noted was thickening of the carotid sheath. No specific mass was seen. Post-operatively he developed a right hypoglossal nerve palsy and continued to gag when pressure was applied to his neck.

In early 1992, his symptoms became worse and he developed difficulty swallowing. Once again pressure on the right anterior triangle of the neck caused extreme sensitivity and gagging response. The CT scan continued to show an abnormal area at the right carotid bifurcation which did not enhance following intravenous contrast (Figure 1).

A repeat exploration of the neck was carried out in September 1992. A well-encapsulated irregular rubbery mass measuring 4 × 3 × 2 cm at the right common carotid artery bifurcation was found. It was lying between the internal and external carotid arteries and produced marked displacement of the ascending pharyngeal artery. The glossopharyngeal nerve was firmly attached to tumour but the vagus nerve trunk which was displaced medially was intact. The mass was totally excised sparing the vagus nerve.

Postoperatively, the patient had total relief of the gagging dysphagia and the pain over the right side of the neck, but he developed dysphonia due to a right vocal cord palsy. There was also decreased sensation over the right side of his oropharynx. Histological analysis of the mass revealed a well-encapsulated ancient schwannoma. In addition

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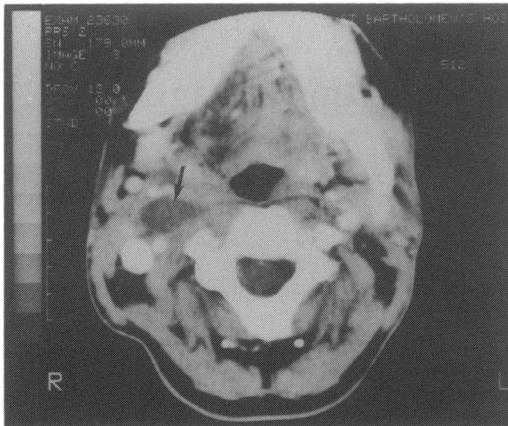


Figure 1 An axial contrast enhanced CT scan of the neck showing a low density mass separating the right internal and external carotid arteries. Arrow indicates the lesion.

there were two smaller schwannomas present within the capsule.

Discussion

Nerve sheath tumours are the second commonest tumour of the parapharyngeal space after salivary gland tumours. They may arise from the lower cranial nerves, cervical plexus, sympathetic chain and brachial plexus. The vagus is affected more commonly than any other nerve. They mainly occur in middle-aged adults and are more common in women than men. Nerve sheath tumours in the head and neck are a common manifestation of neurofibromatosis. Neurofibromas of the plexiform or non-plexiform type are common.¹ Although neurofibromas are often found in association with von Recklinghausen's disease, schwannomas can also occur in these patients, albeit less frequently, and they tend to be multiple. Malignant transformation of these tumours is rare.

A schwannoma is distinguished histologically from a neurofibroma by the characteristic Antoni A and Antoni B cellular pattern compared to the loosely arranged stroma of a neurofibroma. Antoni A areas consist of compact tissue with a high cellularity whereas Antoni B areas are composed of less cellular, loose reticular tissue. An ancient schwannoma shows atypical hyperchromatic nuclei. Immunohistochemical stains for neural-crest marker antigen S-100 protein has helped to

identify nerve sheath tumours from other tumours.

Among the intracranial schwannomas, the acoustic neuroma is by far the most common, although involvement of the fifth, seventh, 10th, 11th and 12th cranial nerves have been described, together with the jugular foramen neurinomas.^{2,3} Schwannomas of the glossopharyngeal nerve, both the intracranial and cervical portions, are rare tumours.

The clinical picture of intracranial glossopharyngeal schwannomas invariably shows a progressive neurological deficit with hearing loss, tinnitus, vertigo, balance disturbances, visual disturbances, pharyngeal and facial hypoaesthesia.⁴ In contrast, the cervical tumours tend to present as a persistent, non-tender, parapharyngeal mass pushing the tonsil and pharynx medially. They are less likely to cause nerve deficit, although they can cause dysphagia, hoarseness, shoulder drop and tongue atrophy. In this report, the main presenting symptom was gagging dysphagia which seemed to be triggered by pressure on the schwannoma. Cough reflex on performing fine needle aspiration in vagal nerve schwannoma⁵ has been described and glossopharyngeal neuralgia with syncope, hypotension, bradycardia has been associated with neck masses⁶ but there has been no previous report of gagging reflex associated with glossopharyngeal schwannoma.

The differential diagnosis of a parapharyngeal space schwannoma are deep-lobe parotid tumours and other salivary gland tumours, lymphoma, chemodectoma and paragangliomas. In most cases, a thorough radiological investigation enables a more precise diagnosis. In the case of a schwannoma, the CT scan will disclose a hypodense tumour mass⁷ with little or no enhancement after contrast administration. Angiographic examination demonstrates an avascular tumour with characteristic displacement of neighbouring vessels. Magnetic resonance imaging has been reported to be more specific than CT scan in the diagnosis of intracranial schwannoma⁸ and when used with GdDPTA can help to differentiate the various type of tumours in the parapharyngeal space.⁹

The treatment of choice of nerve sheath tumours is complete surgical excision. In some cases it is impossible to carry out total excision because of the proximity of vital structures or the residual deformity caused by the procedure. Repeated debulking of these cases has been described in neurofibromas without significant risk of malignant transformation.¹⁰

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