Incidental recurrent laryngeal nerve schwannoma: Treatment considerations for this asymptomatic finding

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ABSTRACT

Objective: To review the presentation, natural history, and diagnosis of recurrent laryngeal nerve (RLN) schwannomas, and to consider the implications of various treatment options.

Study design: Case report and literature review.

Case Report: A 56-year-old woman presented with elevated serum calcium and parathyroid hormone level consistent with primary hyperparathyroidism. A sestamibi scan was equivocal. Ultrasound revealed a left inferior parathyroid adenoma, as well as a second left neck mass. Magnetic resonance imaging demonstrated a 7.5x3.5x3.5 cm mass in the lower neck, extending into the mediastinum. Fiberoptic nasolaryngoscopy showed normal vocal cord function. She underwent a successful excision of a left inferior parathyroid adenoma. Further exploration revealed a mass arising from the left RLN. Careful dissection with RLN monitoring allowed for delivery of the mass from the mediastinum and complete resection with nerve preservation. However, at the termination of dissection, only distal stimulation of the RLN resulted in vocal cord movement. Histopathology was consistent with a schwannoma. The patient has persistent vocal cord paresis eight months post-operatively and vocal cord augmentation is planned.

Conclusion: Schwannomas of the RLN are extremely rare. This is the first case reported in the English otolaryngology literature. Complete surgical excision is the mainstay of diagnosis and treatment for these tumors. Despite nerve preservation with careful surgical technique and intraoperative nerve monitoring, vocal cord paralysis remains a potential complication of tumor resection.

INTRODUCTION

Recurrent laryngeal nerve (RLN) Schwannomas are extremely rare benign tumors of the nerve. We present the first case report of a RLN Schwannoma in the English otolaryngology literature.

Schwannomas of the RLN are difficult to diagnose primarily due to their slow and insidious course. Their size and location make them hard to biopsy and ultimately a complete surgical excision is usually the diagnostic and therapeutic treatment of choice for these tumors. Vocal cord paralysis remains a feared potential complication of such an approach.

CASE REPORT

A 56-year-old woman presented with elevated serum calcium (11.5) and parathyroid hormone level (167) consistent with primary hyperparathyroidism. A sestamibi scan was equivocal. Ultrasound revealed a left inferior parathyroid adenoma, as well as a second left neck mass. Multiplanar T1 and T2 weighted magnetic resonance images with and without contrast revealed a 7.5 x 3.5 x 3.5 cm highly vascular mass located in the deep lower left neck, posterior/inferior to the lower pole of the left thyroid lobe, and extending into the superior mediastinum. It was clearly separate from the thyroid gland, compressing and shifting the trachea to the right. It demonstrated relatively homogeneous low-intermediate T1/high T2 signal, contained multiple small flow voids, and enhanced avidly and homogeneously (Fig.1). Fiberoptic nasolaryngoscopy showed normal vocal cord function. She underwent a successful excision of a left inferior parathyroid adenoma. Further transcervical exploration revealed a mass arising from the left RLN. Careful dissection with RLN monitoring allowed for delivery of the mass from the mediastinum and complete resection, while preserving the nerve. However, at the termination of dissection, only distal stimulation of the RLN resulted in vocal cord movement. Histopathology it was consistent with a schwannoma (Fig.2). The patient has persistent vocal cord paresis eight months post-operatively and vocal cord augmentation is planned.

Figure 1 MRI, Coronal and Axial views. T1, T1-post, T2 scans showing a 7.5x3.5x3.5cm highly vascular mass located in the deep lower left neck, posterior/inferior to the lower pole of the left thyroid lobe, and extending into the superior mediastinum.

DISCUSSION

- Schwannomas (also called neurinomas or neurilemomas) are benign peripheral nerve-sheath tumors that occur in the head and neck region in 25-45% of cases. In the neck, they most frequently arise from the vagus nerve.
- They are associated with Neurofibromatosis (NF) Type 1 in almost 1 in 5 cases. Malignant degeneration of a schwannoma is rather uncommon, yet it is associated with NF in over 50% of cases.
- The greatest incidence occurs between the third and fifth decades of life with no gender discrimination.
- In the head and neck they generally present as single or multiple (3-4%) slow-growing lesions, resembling painless lymphadenopathies, or patients can be completely asymptomatic until other structures are compressed. Symptoms include unilateral vocal fold paralysis with associated dysphonia, dysphagia and progressive laterial cervical enlargement. On physical examination, these tumors are painless, well demarcated and tense.
- The diagnosis is often not suspected until other more common causes of neck masses have been excluded.
- Ultrasonic reveals a well-defined, homogenous, hypoechogenic nodule with hypervascularity under Doppler guidance. CT shows a homogenous mass with a clear border. MRI shows low density in T1W1 and high and diffuse low density in T2W1.
- FNA may be useful to differentiate a nerve neoplasm from a lymph node. The procedure can trigger an excruciating pain which has been considered diagnostic. However, it has a high rate of inconclusive results. CT-guided FNA biopsy of has been reported and proven reliable, but care must be taken when the mass is in close proximity to vital structures.
- If a preoperative diagnosis is made and the patient doesn’t have compressive symptoms, a radiological follow-up could be considered as an alternative to surgery.
- Grossly they appear as a white encapsulated smooth surface tumor with irregular, yellow areas corresponding to foamy macrophages.
- Histologically, there are two types of tissue: Antoni type A and Antoni type B. Other typical features include necrosis, hemorrhage and cystic degeneration.
- Bening Schwannomas are universally S-100 positive. Malignant Schwannomas (neurofibrosarcomas) differ in that they have a higher mitotic rate, necrosis, infiltrative appearance and irregular positivity for S-100.
- The treatment of preference is a conservative surgical excision, or enucleation, to prevent continued expansion and compression of adjacent structures.
- Although, there is an increasing trend towards radiation therapy for vestibular (acoustic) schwannomas, it hasn’t been described as a treatment option for RLN Schwannomas.
- Recurrences are uncommon and have been related to residual disease, as surgical treatment does not always eradicate the disease. Lagner et al. reported that almost 25% of the patients they studied developed new neurogenic tumors.
- If the tumor is malignant, the prognosis is poor with rare survival beyond 1 year after diagnosis.

CONCLUSIONS

- Schwannomas of the RLN are extremely rare benign tumors.
- NF1 appears to play a role as a risk factor for the development of multiple lesions and for malignancy.
- They primarily affect patients between the third and fifth decades with no predilection for genders.
- Their preoperative diagnosis is challenging, thus complete surgical excision remains the definitive diagnostic and therapeutic treatment of choice for these tumors.
- Despite nerve preservation with careful surgical technique and intraoperative nerve monitoring, vocal cord paralysis remains a potential complication of tumor resection.

REFERENCES


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