Schwannomas are benign, slow-growing tumors originating from Schwann cells. Along with neurofibromas, they are considered neurogenic tumors which occur rarely in the larynx, comprising only 0.5%-1.5% of all benign laryngeal tumors. The most common anatomic subsites of occurrence are the aryepiglottic folds and the false vocal folds. Laryngeal schwannomas are suspected to originate most commonly from the internal branch of the superior laryngeal nerve. Definitive diagnosis can only be established histologically, although CT and MRI are helpful in delineating the extent of the lesion. Definitive treatment is surgical excision.

DISCUSSION (continued)

Definitive diagnosis can only be established through histopathologic examination. Schwannomas are composed almost entirely of spindle cells with long, ovoid nuclei and indistinct cell membranes in a stromal network. Two distinctive patterns are associated with this peripheral nerve sheath tumor: Antoni A areas, which are cellular; and Antoni B regions that are less cellular, composed of loosely arranged spindle cells within a mucoid matrix, and often characterized by cyst formation and inflammatory changes. Enzinger and Weiss established three histologic criteria for the diagnosis of schwannomas: (1) the presence of a capsule, (2) the presence of Antoni A and/or Antoni B stroma, and (3) positive stain with S-100 protein, which is a reliable marker for neural crest differentiation seen in schwannomas. Neurofibromas may be differentiated histologically by lack of capsule and mixture of Schwann cells and axons within a collagen matrix. Neurofibromas also stain moderately to strongly with S-100 protein. This, combined with the histologic appearance is typically enough to differentiate these tumors from schwannomas. Immunohistochemical stains for ubiquitin and neurofilament protein, as well as Bielschowsky’s and Bodian’s stains can also be performed. If these stains reveal axons within the body of the tumor, it is most likely a neurofibroma. In addition, neurofibromas are entrained within the nerve fascicles, as opposed to schwannomas, that grow extrinsic to the nerve fascicles.

Distinction between the two types of neurogenic tumors is important for several reasons. First, schwannomas are less likely to undergo malignant transformation, while 10% of neurofibromas have been reported to undergo sarcomatous degeneration. Second, chance of recurrence is greater with neurofibromas. Third, schwannomas, as encapsulated tumors that grow extrinsic to the nerve fascicle, are more likely to be completely excised, and may be excised in some cases without resection of the nerve origin, in contrast to neurofibromas.

Surgical excision is the treatment of choice for laryngeal schwannomas. Endoscopic approach with laser assistance is typically used for smaller tumors, as in our patient. For larger tumors, external approaches may offer superior visualization over purely endoscopic techniques. Several approaches have been reported, such as lateral pharyngotomy, midline thyrotomy, transhyoid approach, and laryngofissure approaches. Transoral robotic surgery may offer another less invasive alternative to open approaches for some tumors. The choice of technique largely depends on the size and location of the tumor and surgeon experience. The majority of patients with laryngeal schwannomas experience complete resolution of symptoms following complete surgical resection, regardless of the operative technique used.

CONCLUSION

Laryngeal schwannoma is a rare tumor of the larynx. Although typically benign and slow-growing, it can cause airway obstruction, dysphagia, or voice changes, as in our patient. Excision may be curative, and can be accomplished endoscopically in select cases.

REFERENCES


CASE REPORT

A 21 year-old otherwise healthy nonsmoking male presented with a 5 year history of worsening dysphonia, without dysphagia or dyspnea. On exam, his voice was breathy, raspy, and weak. Fiberoptic laryngoscopy demonstrated a smooth, lobulated exophytic mass of the left false vocal fold extending into the true vocal fold and anterior commissure. Pre-operative MRI demonstrated a T2-enhancing mass with considerable transglottic extension. The patient underwent microsuspension direct laryngoscopy with an end laryngoscope at the level of the cricothyroid membrane. Pathologic gross examination revealed a 1.9 x 1.8 x 0.9 cm encapsulated mass that revealed a smooth glistening cut surface with focal areas of hemorrhage. Microscopic sections revealed a well-circumscribed lesion within the submucosa, characterized by interfascicles of spindled cells with uniform nuclei that stained positive for S-100 protein. Necrosis was not present, and mitotic activity was low.

Figure 1: Antoni A region - densely arranged spindle cells with nuclear palisading. (20x; H&E stain)

Figure 2: Antoni B region - spindle cells in a loosely cellular arrangement within a mucoid matrix. (20x; H&E stain)

Figure 3: Well circumscribed, encapsulated tumor with diffuse S-100 positivity. Insert shows intense nuclear staining. (1x; S-100; insert 40x)