CASE REPORT

Laryngeal schwannoma as an acute airway presentation

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SUMMARY

A schwannoma is a neurogenic tumour arising from nerve sheaths. Between 25% and 45% of schwannomas occur in the head and neck region. Schwannomas of the larynx are extremely rare. They usually occur in women during the fourth and fifth decades of life. We present a case of a laryngeal schwannoma in a 76-year-old patient with acute stridor, hoarseness and dysphagia. Laryngeal conservation surgery was performed without the need for a tracheostomy. One year later, the patient remains symptom-free with no evidence of recurrence. Clinical presentation, diagnosis and management are discussed and the literature is reviewed.

BACKGROUND

A laryngeal schwannoma is an uncommon tumour that can rarely present as an acute airway.¹ Based on our experience, we believe it is of major importance to cooperate with the anaesthetic team in order to secure the airway and completely extirpate the lesion with conservative surgery, avoiding the need for tracheotomy and external approaches.

CASE PRESENTATION

A 76-year-old woman presented to the ear, nose and throat unit on call via the accident and emergency department, with a 2-day history of acute stridor. She had also reported of progressive hoarseness over a 10-year-period, exertional dyspnoea and dysphagia for 4 months previous to the consultation. She was a non-smoker and consumed alcohol occasionally. Medical history included total thyroidectomy for thyroid goitre.

INVESTIGATIONS

Neither palpable cervical lymph nodes nor café au lait skin patches were present. Fibreoptic laryngoscopy revealed a large mass with a shiny surface, arising from the left false vocal cord completely obstructing the view of the glottis, narrowing the airway and causing a ball-valve effect on inspiration (figure 1). Emergency contrast-enhanced CT demonstrated a 2 cm well-defined lesion in contact with the posterior pharyngeal wall and the lower part of the left aryepiglottic fold (figure 2). The mass was expanding inferiorly to the upper part of the cricoid cartilage. The left piriform recess was intact and there were signs of neither infiltrative growth nor cartilage destruction.

Figure 1 Laryngoscopic view of the mass, completely blocking the glottis.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis includes benign laryngeal lesions such as adenoma, chondroma, laryngeal cyst and internal laryngocoele.

TREATMENT

Written informed consent was obtained from the patient. Under general anaesthesia, bougie-assisted tracheal intubation via video laryngoscopy using a 5.5 endotracheal tube was performed. Microlaryngoscopic cold excision was performed

Figure 2 Contrast-enhanced CT demonstrates a 2 cm well-defined lesion in contact with the posterior pharyngeal wall and the lower part of the left aryepiglottic fold.

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and the mass removed with extreme care to preserve as much normal laryngeal mucosa as possible. The patient was extubated uneventfully without the need for a tracheostomy. She had an uneventful postoperative recovery and was discharged home symptom-free 2 days later.

OUTCOME AND FOLLOW-UP

Histopathological examination showed a tumour that exhibited packed elongated cells with nuclei lining up in palisades to form Verocay bodies and less cellular regions, consistent with schwannoma (figure 3). Postoperative laryngoscopy revealed normal vocal cord mobility. At 6-month and 1-year follow-up visits, the patient remained well with no evidence of recurrence and no residual mass. Her swallowing and breathing were back to normal and her voice was serviceable.

DISCUSSION

Schwannomas and neurofibromas are two types of neurogenic tumours representing less than 1.5% of all benign laryngeal tumours.² Verocay described the first neurogenic tumour of the larynx in 1908.³ Schwannomas are benign slow-growing tumours that arise from the Schwann cells of any nerve except for the olfactory and optic nerves—due to their lack of Schwann cell sheaths.⁴ They occur mostly in women with an increased incidence in the fourth and fifth decades of life. Schwannomas emanate from the Schwann cells and grow adjacent to the parental nerve but extrinsic to nerve fascicles. Neurofibromas derive from perineural fibrocytes, they are not encapsulated and are intertwined with the parental nerve fascicles.⁵ This characteristic is essential because surgical removal of neurofibromas is difficult. Neurofibromas are present most frequently in neurofibromatosis (type 1,2). Malignant transformation occurs in 10% of neurofibromas, while in schwannomas this is very uncommon.⁵ Laryngeal schwannomas are most commonly located in the aryepiglottic folds in the supraglottis (80%) and 20% of them arise from false or true vocal cords. The nerve of origin is the internal branch of the superior laryngeal nerve in most cases or smaller nerve fibres in the laryngeal submucosa.⁶

Patients with laryngeal schwannoma may present with dysphagia, odynophagia, dysphonia, hoarseness, globus sensation, lateral neck lump and dyspnoea. In cases with airway problems, variant types of dyspnoea have been described, such as progressive worsening dyspnoea, inspiratory dyspnoea, exertional dyspnoea, subacute dyspnoea and progressively worsening stridor, wheezing and nocturnal dyspnoea.^{7–13} It is therefore rare for them to present as an acute airway problem, as in our case.

Figure 3 A tumour exhibiting packed elongated cells with nuclei lining up in palisades to form Verocay bodies and less cellular regions, consistent with schwannoma.

There has also been a case of an asphyxial death reported from laryngeal schwannoma.¹⁴

High-definition video laryngoscopy is the examination of choice and usually reveals a submucosal mass. In many cases, a large supraglottic tumour can obstruct the view of the true vocal cords, affecting their mobility and causing a pseudofixation of the cricoarytenoid joint with subsequent symptoms and signs of unilateral vocal fold paresis.

On CTscan, small schwannomas appear as homogenous-enhanced lesions. Larger tumours are heterogeneous with areas of low attenuation surrounded by a peripheral enhanced ring.^{1 15} On T1 and T2-weighted MRI images, schwannomas have a variable intensity with homogenous or heterogeneous presence and are well-enhanced after gadolinium injection.^{1 15} Moreover, they do not seem to destroy adjacent tissues.

Histopathological analysis is the diagnostic gold standard. Enzinger and Weiss established three histological criteria for the diagnosis of schwannoma: (1) encapsulation (2) Antoni A and B areas and (3) S-100 protein positivity.¹⁶ Antoni A areas are characterised by spindle-shaped Schwann cells compactly arranged with nuclei lining up in palisades to form Verocay bodies. Antoni B regions are less cellular with spindle Schwann cells within a myxoid matrix.

Surgical removal is the definite treatment of choice. The approach depends on the size and location of the lesion. Endoscopic excision with or without laser is recommended for small lesions.^{5 17} Larger tumours may require an external approach such as lateral thyrotomy, median thyrotomy or lateral pharyngotomy.⁴ In 2011, Kayhan *et al*¹⁸ reported the first case of a robotic-assisted excision of a laryngeal schwannoma. Sometimes, a tracheostomy may be required to secure the airway. Recurrence or malignancy is rare after total excision. However, recurrences have been described in long-term follow-up following conventional surgery excision. According to the literature, most laryngeal schwannomas with dyspnoea were removed endoscopically using cold steel instruments with or without tracheotomy, and the rest were excised with laser or via an external approach (vertical hemilaryngectomy, medial thyrotomy).^{7–13} In our case, wide removal via microlaryngoscopy using cold steel instruments without tracheotomy, ensured an uneventful postoperative course with no evidence of recurrence.

Learning points

- ▶ Although rare, neurogenic tumours of the larynx should be part of the differential diagnosis of any laryngeal mass.
- ▶ They can even present as an acute airway.
- ▶ The primary aim is, as always, securing the airway, careful planning, discussion with the anaesthetic team and surgical excision.
- ▶ The final diagnosis is made histologically and complete resection should be the definitive aim of surgery.

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REFERENCES

- 1 Wang B, Dong P, Shen B, *et al.* Laryngeal schwannoma excised under a microlaryngoscope without tracheotomy: a case report. *Exp Ther Med* 2014;7:1020–2.
- 2 Jones SR, Myers EN, Barnes L. Benign neoplasms of the larynx. *Otolaryngol Clin North Am* 1984;17:151–78.
- 3 Verocay J. Multiple Geschwülste als Systemerkrankung am nervösen Apparat. In: Festschrift, Hans Chiari aus Anlass seines 25-jährigen Professorenjubiläums gewidmet. Vienna, Leipzig: W. Braunmüller; 1908:378–415.
- 4 Rosen FS, Pou AM, Quinn FB Jr. Obstructive supraglottic schwannoma: a case report and review of the literature. *Laryngoscope* 2002;112:997–1002.
- 5 Zbären P, Markwalder R. Schwannoma of the true vocal cord. *Otolaryngol Head Neck Surg* 1999;121:837–9.
- 6 Phang WK, Raman R, Jayalaksmi E. Neurogenous tumour of the larynx (a case report). *J Laryngol Otol* 1987;101:1209–10.
- 7 Chiu CC, Chou SH, Wu CC, *et al.* Obstructive laryngeal schwannoma in a young female. *World J Surg Oncol* 2015;13:24.
- 8 Murata T, Shino M, Yasuoka Y, *et al.* Subglottic Schwannoma: a report of a rare case that was treated with medial thyrotomy. *Am J Otolaryngol* 2013;34:569–73.
- 9 Ishida Ueha R, Nishimura S, Nito T. Laryngeal schwannoma with extralaryngeal extension. *Auris Nasus Larynx* 2012;39:317–20.
- 10 Rognone E, Rossi A, Conte M, *et al.* Laryngeal schwannoma in an 8-year-old boy with inspiratory dyspnea. *Head Neck* 2007;29:972–5.
- 11 Ndiaye M, Rouleau V, Rame JP, *et al.* [Laryngeal schwannoma: a case report and review of the literature]. *Rev Laryngol Otol Rhinol (Bord)* 2006;127:243–6.
- 12 Iglesias Moreno MC, Bhathal Guede B, González Gimeno MJ, *et al.* [Neurilemmoma of the larynx: a case report]. *Acta Otorrinolaringol Esp* 2004;55:446–50.
- 13 Pulli RS, Coniglio JU. Subglottic nerve sheath tumor in a pediatric patient: case report and literature review. *Head Neck* 1997;19:440–4.
- 14 Gardner PM, Jentzen JM, Komorowski RA, *et al.* Asphyxial death caused by a laryngeal schwannoma: a case report. *J Laryngol Otol* 1997;111:1171–3.
- 15 Plantet MM, Hagay C, De Maulmont C, *et al.* Laryngeal schwannomas. *Eur J Radiol* 1995;21:61–6.
- 16 Enzinger FM, Weiss SW. Benign tumors of peripheral nerves. In: Enzinger FM, Weiss SW, eds. *Soft tissue tumors*, 2nd edition. St. Louis: Mosby; 1988: 725–35.
- 17 Kou W, Zhang C, Wei P. Laryngeal schwannoma treated with a CO₂ laser: a case report. *Oncol Lett* 2015;9:1467–9.
- 18 Kayhan FT, Kaya KH, Yilmazbayhan ED. Transoral robotic approach for schwannoma of the larynx. *J Craniofac Surg* 2011;22:1000–2.

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