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Laryngeal paraganglioma: an endoscopic diode-laser-assisted surgical approach: a case report

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Abstract. *Laryngeal paraganglioma: an endoscopic diode-laser-assisted surgical approach: a case report.* Laryngeal paraganglioma is a rare vascular neuroendocrine benign tumour. Surgery is the standard treatment. The pre-operative gold standard is Magnetic Resonance Imaging with Gadolinum contrast. Computed tomography scans and angiography are also useful in making the diagnosis of paraganglioma before surgical intervention.
A case of a large supraglottic paraganglioma is described. Complete excision of the tumour was achieved using micro-laryngoscopical-laser surgical resection. No severe complications were observed. The patient was discharged from hospital without any complaints on the fourth post-operative day and she was disease-free 36 months after surgery.
The effectiveness of this conservative endoscopic surgical procedure compared to the external approaches is discussed and it can be concluded that the endoscopic diode-laser resection of even extended laryngeal paraganglioma provides a good operative field and is a safe and effective technique.

Introduction

Laryngeal paragangliomas are unusual benign tumours arising from the superior or inferior laryngeal paraganglia. Laryngeal neuroendocrine tumours are usually seen in the supraglottic larynx[1,2,3] but have also been found in the glottis and subglottis.[2,4] They are more frequent from the fourth to the sixth decades of life with a female/male ratio[1,2,5] of 3-4/1.

Clinical symptoms depend on the location and size of the tumour[6]. Supraglottic paragangliomas can cause hoarseness, dysphagia, dyspnoea, stridor, and the sensation of a foreign body in the throat. Subglottic paragangliomas may result in wheezing and stridor as they grow. Haemoptysis, sore throat and coughing have also been reported.[1,7] Neuroendocrine neoplasms of the larynx have been divided into those of epithelial or neural origin[5]: the epithelial-origin group can be divided into the typical or atypical carcinoids and small-cell neuroendocrine carcinomas; the neural-origin group consists of paragangliomas.

The pre-operative gold standard is Magnetic Resonance Imaging (MRI) with Gadoliniun contrast[6] or ^{111}In-pentetreotide scanning, which make it possible to distinguish neuroendocrine tumours from other submucosal laryngeal lesions.[6] The correct pathological diagnosis of neuroendocrine tumours is made by conventional histology, immunohistochemistry and electron microscopy. It is of major importance for paragangliomas to be differentiated from other neuroendocrine tumours of the larynx because of different treatment modalities. Laryngeal paragangliomas are rarely malignant[8] so a conservative surgical procedure should suffice.

A case of a complete endoscopic diode-laser surgical resection of a laryngeal paraganglioma is presented. The effectiveness of this conservative surgical procedure is discussed by comparison with the external approaches. Judging from the cases reported in this study and those identified in the literature, it can be concluded that the endoscopical treatment is a minimally invasive surgical approach with the potential to become the treatment of choice for selected benign tumours such as the paraganglioma described here.

Case report

A 57-year-old female patient was referred to our ENT department complaining of moderate hoarseness for the previous 6 months, dyspnoea, and a sore throat, but no dysphagia. The patient was a non-smoker and non-drinker. Indirect laryngoscopy and examination with a flexible endoscope showed a smooth, well-delimited mass on the right-hand side of the supraglottic region, with intact laryngeal mucosa, extending from the

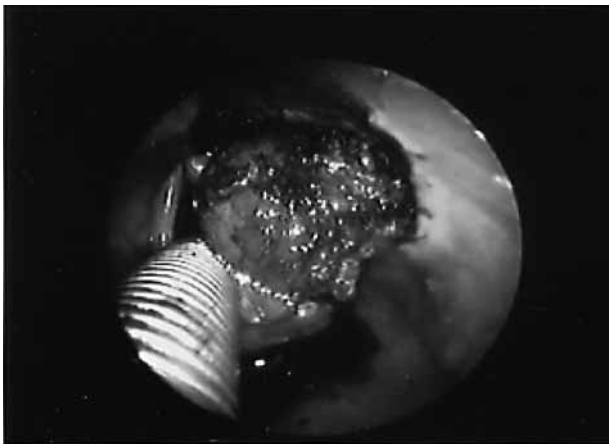

Figure 1
Smooth delimited mass on the right side of the supraglottic region, extending from the level of the aryepiglottic fold to the right true vocal cord.

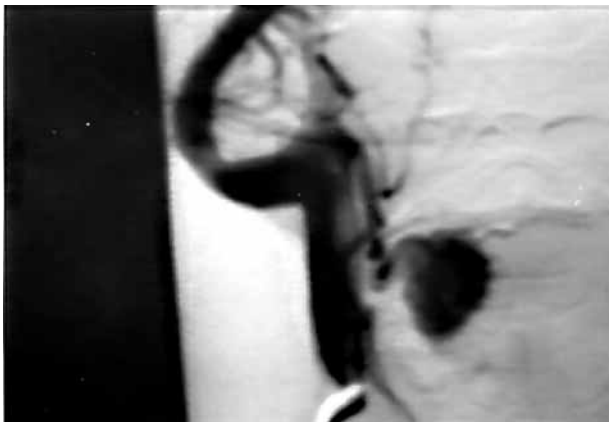

Figure 2
Arteriography demonstrating an evident vascular supply arising from the superior laryngeal artery.

level of the aryepiglottic fold to the right true vocal cord (Figure 1). Both vocal cords were normally mobile upon examination with indirect laryngoscopy and a flexible endoscope. There were no palpable cervical lymph nodes or masses.

Pre-operative radiographic diagnostics included: 1) high resolution computed tomography (HRCT scan), demonstrating an enhancing mass in the right laryngeal region consistent with a vascular mass; 2) MRI with Gadolinium showed an enhancing well-delimited tumour-like lesion extending from the right aryepiglottic fold to the level of the right true vocal cord; 3) arteriography demonstrated an evident vascular supply arising from the superior laryngeal artery (Figure 2). On the basis of the radiological findings the differential diagnosis included paraganglioma, hemangioma and schwannoma. Two days before surgery, given the decision not to adopt an open surgical approach, a nearly complete pre-operative devascularisation of the neoplasia by selective embolisation under local anaesthesia was obtained without any complications.

Surgical technique

The operation was performed under general anaesthesia using endotracheal intubation. A microlaryngoscopical procedure was performed. After a diode-laser mucosal incision was made over the mass, the tumour was excised stepwise keeping the thin fibrous capsule intact, allowing it to be safely dissected from the surrounding musculature of the larynx. A complete transoral removal of the tumour was obtained using a diode-laser-assisted technique –DIOMED 30 contact fibre FLD-D 5241 – utilising 5 watts in continous delivery for about 5000 joules of total energy emission. Intraoperative blood loss was minimal.

The complete removal of the tumour resulted in a large endolaryngeal wound. Fibrin glue was used to prevent post-operative bleeding, and a nasogastric feeding tube was placed. After the operation, the patient was nursed in the ENT department and received i.v. antibiotics and prednisolone for 3 days. The further clinical course was uneventful; there were no severe complications such as haemorrhage, dyspnoea, dysphagia or laryngeal oedema. The patient was discharged from hospital without any complaints on the fourth post-operative day. A three-year follow-up has showed no recurrence.

Histopathological findings

Macroscopically the surgical tumour specimen was 20 × 17 × 12 mm in size and of soft consistency. It was circumscribed by a thin capsule of connective tissue. The cut surface had a lobulated, grey-reddish, elastic appearance. **Microscopically** the tumour was composed of epithelial-trabecular tissue with some solid areas and thin fibrous tissue strands. The well-defined nests of epithelioid cells were arranged in distinct clusters (“Zellballen”) (Figure 3). Vascular channels containing erythrocytes in an organoid pattern separated the tumour cells. There

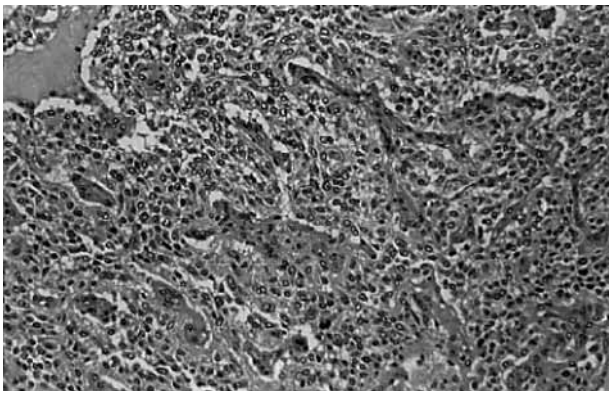

Figure 3
Clusters and nests of uniform cells surrounded by capillaries in an organoid pattern (H & E, × 100).

were uniform polygonal cells with indistinct borders and moderate amounts of slightly granular eosinophilic cytoplasm. Pleomorphic and giant nuclei with a lightly stipped chromatin pattern were also observed. Mitotic figures were rare.

Immunohistochemical investigations of the tumour showed a positive reaction for the neuroendocrine markers chromogramin, synaptophysin, neuron-specific enolase (NSE), and protein gene product (PGP) 9.5. The sustentacular cells stained positively with the S-100 protein. By contrast, the reaction with NB84, which is a marker for neuroblastomas, was negative. The absence of immunostaining for cytokeratin, calcitonin and carcinoembryonic antigen (CEA) was a feature that supported the diagnosis of paraganglioma, as these antigens are identified in atypical carcinoid tumours.

The histopathological findings mentioned above resulted in the diagnosis "Laryngeal paraganglioma".

Discussion

Paragangliomas are rare neuroendocrine tumours. They are typically located in the gastrointestinal tract and the lungs, but the clinical course differs from that of the extremely rare laryngeal paraganglioma.[9] In the head and neck region, they are most commonly associated with the carotid body, vagus nerve, jugulotympanic paraganglia, and occasionally the superior and inferior laryngeal paraganglia.[4] Paragangliomas of the larynx are seen as a vascular submucosal mass and they occur mostly in the supraglottic space.[2]

Pre-operative angiography to evaluate the size, extent and especially the vascular structure and blood supply of the lesion, possibly in combination with selective embolisation, may be extremely helpful.[2,8]

A review of the literature shows that the treatment of choice of laryngeal paraganglioma is surgical resection.[10-13] The surgical excision of benign submucosal lesions of the larynx can be performed using a variety of techniques including laryngofissure, lateral pharyngotomy, medial thyrotomy, supraglottic laryngectomy and endoscopic procedure. Judging from the case reported in this study and those identified in the literature, malignant biological behaviour associated with laryngeal paraganglioma[11] is extraordinarily rare (< 2%). Because this is a benign submucosal lesion, conservative surgery is recommended, elective neck dissection is not necessary and the prognosis is excellent.[8]

Rubin *et al.*[10] described a conservative approach to submucosal lesions of the supraglottic larinx including paraganglioma: ***supero-lateral thyrotomy*** consists of the subperichondrial resection of the superior half of the ipsilateral thyroid cartilage with preservation of the internal lining and superior laryngeal nerve. The lesion may then be enucleated or resected and the defect closed with overlying mucosa and the flap of preserved perichondrium.

Myssiorek *et al.*[13] suggested that small submucosal lesions of the larinx can be removed via an external approach without a tracheotomy.

Del Gaudio *et al.*[2] reported a midline laryngofissure with mucosal preservation and concluded that an external mucosa-sparing approach is the treatment of choice.

Radiation, either pre-operative or post-operative, and chemotherapy as an alternative to surgical treatment, have been reported to be ineffective in the management of paragangliomas of the larynx.[1]

In recent years, the endoscopical, laser-microsurgical resection of large laryngeal tumours has increased.[14] Sesterhenn *et al.*[6] first described the complete and successful transoral resection of a large paraganglioma of the larynx using CO_2 laser surgery. Endoscopic excision alone had already been rejected by many authors because of the reduced exposure in the endoscopic opera-

tion site[1,15] and the possibility that haemorrhage might obscure the visualisation of tissue planes.[2] This could be true in those cases where the localisation of the tumour is unfavourable, for example in the subglottic space. Konowitz *et al.*[16] reported a case of a patient with a paraganglioma of the larynx who was unsuccessfully treated by laser excision and required surgical resection.

Even though open surgical procedures have resulted in a superior disease control rate,[6] the present case report successfully demonstrates that extended (20 × 17 × 12 mm) supraglottic vascular lesions can be safely excided endoscopically. Diode-laser treatment has allowed the excision of the tumour with the use of a fibre in direct contact with the endolaryngeal mass, so that the control of surgical margins has been safe. Due to the pre-operative devascularisation of the neoplasia by selective embolisation, intraoperative blood loss was minimal. The patient did not undergo tracheotomy and she was discharged without any complaints on the fourth post-operative day.

Conclusions

In conclusion, the endoscopic diode-laser surgical resection of even extended laryngeal paragangliomas provides a good operative field and is a safe and effective technique. Is has some advantages over the open procedures such as reduced morbidity, length of stay and a superior cosmetic result. The endoscopic diode-laser resection may be an alternative to conventional surgical excision in selected cases, and it merits use as an alternative treatment modality.

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