CASE REPORT

Novel surgical management of a laryngeal granular cell tumour

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SUMMARY

Granular cell tumour (GCT) is a rare benign tumour occurring, most commonly, in the head and neck. Multiple tumours occur in 5–16% of patients. These tumours are chemo-radio-resistant and have high recurrence rates despite their benign histopathological features. Traditional management, depending on access, involves total tumour resection with wide margins due to the high rates of recurrence with incomplete resection. We present a patient with two synchronous GCTs of their upper airway: in the larynx and the trachea. Complete excision of the supraglottic lesion would have rendered the patient's larynx incompetent. Therefore, after multidisciplinary team (MDT) discussion, and following a discussion with the patient regarding the risks and benefits, a novel surgical management approach was agreed. Subtotal CO₂ laser excision of the upper GCT was performed enabling functional organ preservation. A subsequent procedure was required when the patient became symptomatic due to tumour recurrence.

BACKGROUND

Granular cell tumour (GCT) is a benign subepithelial tumour; 50% of these tumours occur in the head and neck with 10% arising in the larynx.¹ They most commonly appear in the third–sixth decades of life and have a slight female preponderance.² They are typically slow growing, well-circumscribed masses and often present with symptoms of dysphonia, hoarseness, dysphagia or odynophagia. Multiple tumours occur in 5–16% of cases and, therefore, it is important to look for a second synchronous tumour.¹ The treatment of choice for this chemo-radio-resistant tumour is complete surgical excision with clear margins due to its high recurrence rate.^{3–5} Tumour recurrence after excision with clear margins is reported to be around 8%, however, when the margins are positive, the recurrence rate rises to 21–50%.⁶ Malignant GCTs are rare, accounting for 1–2% of cases, but should be considered when presented with large tumours >4 cm in size.⁷

CASE PRESENTATION

A 48-year-old woman presented with progressive dysphonia and progressive difficulty in breathing particularly on exertion. Her symptoms had begun 12 months previously but only became troublesome over the past 3 months. She denied having a cough or any gastro-oesophageal reflux symptoms. There was no significant medical history and she was not taking any regular medications. She had a

15-pack-year smoking history, and rarely drank alcohol. On examination, she had no palpable cervical lymph nodes. A flexible laryngoscopy was performed in clinic, which revealed a large (apparently 2×3 cm), well-circumscribed, submucosal supraglottic mass arising from the right arytaenoid complex and aryepiglottic fold. Both vocal cords were functioning normally.

INVESTIGATIONS

The patient underwent urgent contrast-enhanced CT of the neck and thorax, and was discussed in the head and neck multidisciplinary team (MDT) meeting. The CT scan showed a 7×1.4×3.3 cm soft tissue mass in the hypopharyngeal region at the level of the larynx with supraglottic as well as subglottic components. There was no invasion of surrounding tissues or bony destruction. There were no enlarged or suspicious looking lymph nodes in the neck.

A pan-endoscopy and microlaryngoscopy was performed in order to inspect the mass macroscopically, take biopsies for histological diagnosis and look for a second primary. This revealed that there were, in fact, two separate masses in the airway; a supraglottic mass overlying the right arytaenoid (figure 1) and a tracheal mass 3 cm proximal to the carina. Both were well-circumscribed submucosal soft tissue masses. Both masses were biopsied using cold steel, and histology confirmed both to be GCTs; mucosa covered by mildly hyperplastic squamous epithelium in which there was a well circumscribed lesion composed of spindle cells with granular cytoplasm was noted. These cells diffusely express S100 protein on immunohistochemistry and do not express smooth muscle actin or desmin. The proliferation index with Ki-67 is less than 5%. These appearances are consistent with a GCT.

Figure 1 Microlaryngoscopic view of the laryngeal granular cell tumour.

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DIFFERENTIAL DIAGNOSIS

Differential diagnosis for a mass in the upper airway includes: granuloma, schwannoma, chondrosarcoma and squamous cell carcinoma. When a histological diagnosis is being sought, it is important to ensure that deep biopsies are taken as 50–65% of GCTs develop squamous ‘pseudoepitheliomatous hyperplasia’ of the overlying epithelium with the formation of keratin pearls.⁶ If superficial biopsies are obtained, the tumour can be misdiagnosed as squamous cell carcinoma, the management of which is, consequently, much more radical.

TREATMENT

Management of a GCT typically requires clear surgical margins to reduce the risk of recurrence. However, due to the upper airway location of the lesion, overlying the right arytaenoid in the supraglottis, complete surgical resection would have rendered the larynx incompetent. The patient would have had impaired vocal cord movement with incomplete closure of the glottis and would, consequently, develop an unsafe swallow and airway, weak cough and impeded phonation. After careful MDT discussion and gaining informed consent from the patient, a less radical approach was agreed on and the laryngeal GCT was debulked using microlaryngoscopy and CO₂ laser (figure 2). The tracheal GCT was cleared radically and showed no recurrence subsequently.

OUTCOME AND FOLLOW-UP

The decision to manage the GCTs more conservatively was made with the knowledge that close follow-up and repeat procedures to clear the tumour may be required. After a period of 12 months post the initial surgery, the patient became symptomatic, developed difficulty in breathing from the recurrent tumour, and had a further laser procedure. A total of 18 months have now passed since the second procedure and she remains asymptomatic. She is now followed up every 6 months with flexible laryngoscopy examination in the outpatient clinic.

DISCUSSION

The function of the larynx is, first and foremost, to protect the airway; during the process of swallowing the vocal cords close forming a seal. In addition, the epiglottis reflects over the vocal cords adding further protection of the airway. Without this protection, fluids and solids could be aspirated into the lower airway resulting in aspiration pneumonia. The functioning larynx also allows for phonation and Valsalva manoeuvre. The GCT in this patient involves the right arytaenoid cartilage and right aryepiglottic fold. Complete excision would prevent adequate adduction of the vocal cords and render the airway

unsafe. Incomplete excision is a novel approach due to the high recurrence rate of these tumours if not completely excised.

Similar cases in the literature, of GCTs of the upper airway, have been managed by complete surgical excision, however, the location and the size of the tumour has not rendered the larynx incompetent. Arevalo *et al* published two cases of patients with left vocal cord GCTs. One patient underwent complete excision with cold steel surgery, which resulted in negative margins and no recurrence after 4 years of follow-up. The other patient had complete excision using laser and no recurrence after 2 years of follow-up.³ Holton *et al* published a case of a paediatric patient with a GCT of the right vocal fold who underwent complete laser excision with CO₂ laser. They used intraoperative frozen sections to ensure that they achieved clear margins and 8 months after surgery, the patient had no recurrence of tumour. Although the patient’s voice quality had improved since presentation, she did have a slightly husky, breathy voice.⁸ We found one other case of subtotal excision of a laryngeal GCT in the literature. The patient had a GCT obscuring the posterior third of the left vocal cord extending inferiorly between the vocal folds. The patient underwent subtotal excision using cold steel, reserving the CO₂ laser for control of bleeding. The patient did not have any evidence of recurrence at 6 months, however, follow-up beyond this was not reported.⁹

Learning points

- ▶ Granular cell tumours (GCT) of the upper airway can be multifocal and, therefore, it is important to look for a second primary tumour.
- ▶ Deep biopsies must be obtained from GCT to avoid misdiagnosis of squamous cell carcinoma in the case of ‘pseudoepitheliomatous hyperplasia’ of the overlying epithelium.
- ▶ When considering surgical management of GCTs in the upper airway, the location of the tumour is crucial in planning total or subtotal excision in order to maintain a functioning larynx.
- ▶ Subtotal excision of GCTs have a high risk of recurrence, however, further procedures can be performed when the patient becomes symptomatic.

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Figure 2 Post laser subtotal excision of the laryngeal granular cell tumour.

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