

## Case Report

# Paediatric Granular Cell Tumour of the Larynx: A Case Report of Laryngo fissure Resection and Buccal Mucosal Graft

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### INTRODUCTION

We present a case of a granular cell tumour (GCT) in the larynx of a 9-year old boy. The literacy review confirmed that this would be the youngest patient reported in Sri Lanka. The histological diagnosis of GTC is emphasized as macroscopic appearance may confuse with viral papillomatosis of the larynx. In addition, we present a method of surgical treatment of the tumour, involving the hitherto unreported technique through laryngofissure excision and buccal mucosal graft to preserve the exact function of the larynx with complete excision of the tumour.

### CASE REPORT

A 9-year old boy, referred to our unit with a history of gradual onset of snoring while sleeping associated with sweating, dry cough, weight loss and difficulty in breathing followed by forced expiration with husky voice. He had been born by normal delivery at full term. There was no maternal history of genital warts.

He was repeatedly treated for recurrent attacks of lower/upper respiratory tract infection at nearby peripheral hospital. The condition progressed slowly without apparent improvement and his school teacher noticed a muffled voice. After one year later (2011) he was referred to the closest base hospital for ENT opinion. ENT examination had revealed no tonsillar enlargement, enlarged adenoid or any other common pathology to describe the upper airway obstructive symptoms. But the rigid laryngoscopy was, therefore performed to assess the upper respiratory tract which revealed a right side vocal cord growth. The patient was then transferred to the Paediatric ENT unit of SBSCH Peradeniya for further management.

At SBSCH, a growth on posterior one third of right vocal cord was confirmed with direct laryngoscope. The granular polypoidal appearance of the lesion was suggestive of viral papillomatosis. The left vocal cord was normal. A punch biopsy was taken for histology which revealed a papillomatous lesion lined by squamous epithelium. The underlying stroma contained diffuse sheets of large, polygonal cells with abundant eosinophilic granular cytoplasm. The nuclei were small, uniform and round. The tumour extended to the base of the polyp and involved the resection

margin. The histomorphological diagnosis of granular cell tumour was made which was later confirmed by immunopositivity of the tumour cells for S100 stain. MRI scan was performed to assess the extent of the tumor.

Literacy review suggested the best current treatment for this condition is laser resection. As at the moment laser facility is not available in Sri Lanka we opted for elective tracheotomy and Laryngo fissure access to vocal cord for complete tumor excision. The lesion was excised with a good margin and depth of the dissection was up to inner perichondrium of the thyroid cartilage. Therefore the defect created was large and primary closure without tension on the neighbouring mucosa was impossible. Therefore adequate mucosal epithelial graft was harvested from the buccal mucosa and primary grafting was done. Post operative period was uneventful and tracheotomy decannulation was achieved in one week time. The patient was kept on regular clinic follow up and so far he is with satisfactory normal voice. The indirect laryngeal examination showed no granulations or recurrence of the tumour.

### DISCUSSION

Only few cases of laryngeal GCT have been reported in paediatric patients. These tumours are usually sessile, painless and somewhat firm immovable polypoidal nodules. Grossly GCT may be confused with recurrent juvenile viral laryngeal papilloma except that the latter is often multiple and may grow to a considerable size obstructing the larynx and has a tendency to recur necessitating repeated excisions.

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Viral papilloma also has a tendency to spread through respiratory tract and exhibits a tendency for spontaneous remission. GCT on the other hand does not have tendency to spread beyond the respiratory tract and has less than 7 % recurrence after excision. Therefore though viral papilloma of the larynx is the commonest growth in the larynx of paediatric patient, GCT should be considered in the differential diagnosis.

Granular cell tumour, also called Abrikossoff tumors, is a benign, slowly growing neoplasm, presumably of Schwann cell origin. They may occur anywhere in the body, although 50% occur in the head and neck. The most common site is the tongue; the larynx is involved in approximately 10% of all cases. Granular cell tumours typically develop in the fourth and fifth decades of life and are quite rare in children. A slight female preponderance has been reported. About 10% of patients develop multifocal synchronous or metachronous tumours. Clinically, patients with laryngeal granular cell tumors present with hoarseness, dysphagia, and cough and, less frequently stridor. Most tumours are smaller than 2 cm and larger tumours can cause difficulty in breathing. The vast majority of tumours arise from the posterior aspect of the true vocal folds and approximately half of these extend into the sub glottis.

The granular cell tumour being a tumour of nerve sheath origin shows immuno positivity for S100 protein. The tumour cells are large and polygonal with distinct cell membranes. Faintly eosinophilic small granules are present in the cytoplasm. Nuclei are small and usually centrally located. Mitoses are generally not present. Pseudo epitheliomatous hyperplasia of the over lying epithelium is a usual finding.

Literature review suggested that the ideal treatment of this tumour is local excision with histologically negative margins. Proposed treatments for these lesions have included local 'cold steel' excision or laser excision, laryngofissure for large lesions, and, rarely, laryngectomy. Granular cell tumours are radio resistant, therefore radiation therapy is not indicated in the treatment of these benign lesions.

Our case highlights the challenge of preserving normal function of the larynx and complete tumour excision especially in a paediatric patient. The ideal is to have laser facility which will help to excise the lesion with a minimal scar. After thorough literature survey it was decided that the best option for this patient is the Laryngo fissure access. Elective tracheostomy and routine midline laryngofissure was performed. Having well exposed, the lesion was found to be attached to posterior end of the right true vocal fold spreading to subglottis measuring about 2 cm. As it was a sessile lesion, wide local excision without sacrificing significant amount of mucosa was inevitable. The exposed inner aspect of the laryngeal cartilage had to be lined with a mucosa to achieve primary intention healing with minimum scarring. As expected in the preoperative planning the locally available mucosal margins were not adequate for the primary closing without tension. A satisfactory mucosal graft was harvested from the buccal mucosa and sutured using monofilament absorbable suture material.

Meticulously Laryngo fissure was repaired paying attention to avoid any future web formation at the anterior commissure by suturing the inner lining perichondrium to outer one first and then two halves together.

In conclusions, though the juvenile laryngeal papilloma is the commonest childhood benign laryngeal lesion, laryngeal GCT should be considered in the differential diagnosis and the new surgical method adopted would be a good option in the absence of laser resection.

## REFERENCES

- Conley SF, Milbrath MM, Beste DJ. Paediatric laryngeal granular tumour. *Otolaryngol* 1992;21:45-3.
- Pransky SM, Seid AB. Tumours of the larynx, trachea and Bronchi. In: Bluestone CD, Stool SE (Eds). *Pediatric Otolaryngology*: Philadelphia: WB Saunders; 1990. p. 1215-8.
- Lazar RH, Younis RT, Kluka EA, Joyner RE, Storgion S. Granular cell tumor of the larynx: Report of two pediatric cases. *Ear Nose Throat J* 1992;71:440-3.
- Goldosky E, Hirschfield LS, Abramson AL. An unusual Laryngeal tumour in children: Granular cell tumour. *paediatrOtorhinolaryngol*. 1988;15:263-7.