CASE REPORT

Oropharyngeal paraganglioma presenting with stridor: an unusual presentation

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SUMMARY

Head and neck paraganglioma is a rare disease to encounter in clinical practice and involvement of the base of the tongue in the oropharynx is yet to be described in the current literature. Although various surgical approaches have been described in the literature, transcervical mandibular swing is an effective technique for adequate exposure and complete excision of the oropharyngeal paraganglioma with adequate control on bleeding especially when it is assisted by coblation.

BACKGROUND

Paraganglioma is a rare benign lesion that accounts for 0.6% of the all the benign tumours affecting the head and neck region.¹ They originate from the paraganglionic cells from neuroendocrine tissues extending from the skull base to the pelvic floor and the most common sites affected are the head and neck region in association of the lower cranial nerves.^{2,3} In the head and neck region, the vagus nerve, jugulotympanic complex and superior-inferior laryngeal paraganglia are the predominant sites to be affected by the disease. Looking into the literature, total 77 cases of laryngeal paraganglioma have been documented to date and most of them are found to involve the supraglottis larynx.^{4,5} The oropharynx is very rarely affected by the disease and to the best of our knowledge, only two cases have been reported in the literature affecting the tonsil^{6,7} and no clinical report has been documented to date affecting the base of the tongue. In the larynx, both the superior and inferior laryngeal nerves are the most common sites of origin for the paragangliomas.^{8,9} In contrast, no definite site of the origin has been confirmed in the case of oropharyngeal paraganglioma which could be due to the rare occurrence of the disease at the concerned site. Women in fourth to sixth decade of life are predominantly get affected by the disease. Clinical presentation depends on the site and the extension of the tumour which includes haemoptysis, haematemesis, change in voice, dysphasia and very rarely stridor. Flexible fibre-optic laryngoscopy is always carried out as the first investigation in cases of oropharyngeal lesion to look for the nature, the extent of the lesion and for the assessment of the upper airway. Radiological evaluation (contrast-enhanced MRI with MR angiography) is most vital in assessing the vascularity, site of the origin and the predominant feeding vessels and later may be required for preoperative embolisation or controlled ligation during

surgery. Histopathology is always confirmatory for the final diagnosis demonstrating a vascularised tumour consisting of two types of cells, the chief cells (polygonal) and the subtentacular cells.

Chief cells are polygonal cells with inconspicuous nuclei and eosinophilic cytoplasm with characteristics of 'Zellballen' pattern. Subtentacular cells contain basophilic cytoplasm, are argyrophil positive and argentaffin negative.¹⁰ Again, chief cells are immunopositive to synaptophysin and negative for chromogranin. The subtentacular cells show immunopositivity to the S-100 protein.¹¹ Management is mostly by surgical excision with removal of the regional draining lymph nodes to rule out the underlying malignancy. Although recurrence is very uncommon, patients need a close follow-up in the postoperative period.

CASE PRESENTATION

A 42-year-old male patient presented to the emergency department with stridor for 15 days, dysphagia and change of voice for 1 month. Flexible nasopharyngoscopy revealed a large irregular mass almost occluding the lumen of the oropharynx. The endolarynx was found normal with bilateral mobile vocal cords. General physical examination and systemic examination were found to be normal. Emergency tracheostomy was performed as a life-saving measure.

INVESTIGATIONS

Contrast-enhanced MRI (T1-weighted) revealed a heterogeneously enhancing mass, almost occluding the whole lumen with abroad fibrovascular attachment at the tongue base (figure 1) with multiple cervical lymph nodes (level II, III on right side). MR angiography revealed the predominant blood supply coming from the right lingual artery (figure 2). Routine haematological and biochemical tests were found to be normal. The direct laryngoscopic biopsy was taken under local anaesthesia and the biopsy report was confirmed to be a paraganglioma. Biochemical screening was done for 24 hours urinary catecholamine and it was found to be negative. Histopathology revealed tumour cells arranged in nests of clusters separated by subtentacular cells (Zellballen pattern). The cells are large, polygonal with abundant eosinophilic granular cytoplasm, centrally placed round to oval nucleus with vesicular chromatin and conspicuous nucleolus (figure 3). The tumour cells are immunopositive to the S100 as demonstrated in figure 4. After

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Figure 1 Contrast-enhanced MRI (T1-weighted) revealed a heterogeneously enhancing mass occluding the lumen of the oropharynx.

confirmation of the biopsy, the patient was counselled for excision of the mass under general anaesthesia.

TREATMENT

Through a transcervical route, complete exposure of the tumour was achieved with mandibular swing by a paramedian mandibulotomy (figure 5). A controlled ligature was taken over the lingual artery on the right side of the neck that may be required for the excessive intraoperative bleeding. Complete excision of the mass was achieved with the help of the coblation. The final histopathological

Figure 2 MR angiography showing the major blood supply coming from the right lingual artery.

Figure 3 Histopathology showing tumour cells arranged in nests of clusters separated by subtentacular cells (Zellballen pattern).

report was the same as the preoperative findings and the dissected lymph nodes were found free of tumour metastasis.

OUTCOME AND FOLLOW-UP

The patient is on regular follow-up for the past 6 months with endoscopic evaluation and found asymptomatic without the recurrence of the tumour.

DISCUSSION

Paraganglioma is a rare benign tumour arising from neuroendocrine tissue extending from skull base to the pelvic floor. Head

Figure 4 The tumour cells show immunopositivity for the S-100 protein.

Figure 5 Tumour was exposed through mandibular swing by a paramedian mandibulotomy.

and neck region frequently get affected by the disease and rarely it affects the larynx and trachea as documented in the literature.^{12,13} To the best of our knowledge, only two cases have been reported in the literature affecting the tonsil^{6,7} and no clinical report has been documented to date affecting the base of the tongue. Women (male to female ratio: 1:3) with sixth decade of their life are predominantly affected by the disease, in contrast to the present case, where the patient was a 42-year-old male patient. Clinical presentation mainly depends on the site and extension of the lesion. Change in voice and dysphagia are the common symptoms encountered in clinical practice and very rarely patients can present with stridor as was found in our case which required emergency tracheostomy. Flexible fibre-optic laryngoscopy is the first investigation carried out in patients with oropharyngeal paraganglioma to assess the nature, the extent of extension of the lesion and the patency of the upper airway. Contrast CT/MRI are vital to assess the site of origin, the vascularity of the lesion and to differentiate the similar swelling in the oropharynx. CT/MR angiography is performed in selective cases of paraganglioma to evaluate the feeding vessels, especially whenever it is planned for preoperative embolisation. In our case, we did not have the facility of embolisation and intraoperative controlled ligation over the feeding artery was taken to decrease the intraoperative bleeding. There has been always a debate whether for prerogative i biopsy of the tumour keeping in mind the high vascularity of the tumour mass and it is often avoided due to the risk of massive bleeding^{2,14} and the clinical diagnosis mostly depends on the flexible fibre-optic laryngoscopy and radiology. Most of the extra-adrenal paraganglioma are non-secretory in nature and in very few cases (1%–3%), it secretes catecholamine.¹⁵ Hence it is a routine practice for biochemical screening for urinary catecholamine to exclude the functional paragangliomas in each case irrespective of the sites of origin. Surgery is considered as the treatment of choice for the management of head neck paragangliomas,² aimed at complete removal of the tumour without damaging the surrounding structure. Although various surgical approaches have been

highlighted in the previous literature^{10–19} for laryngeal paragangliomas, open transcervical approach can be an effectively adopted for the lesion in the oropharynx. Mandibular swing through a paramedian mandibulotomy is an ideal surgical technique for oropharyngeal paraganglioma ensuring for complete excision of the lesion and adequate haemostasis. Neck dissection is usually not indicated²⁰ in paragangliomas, but regional cervical lymph nodes should be included in the primary specimen to rule out the malignant transformation of the lesion.^{21,22} Although the recurrence rate after complete excision of the tumour is very low, the patient needs a close follow-up after the surgery.

Learning points

- ▶ Paraganglioma is a rare benign tumour arising from neuroendocrine tissue extending from skull base to the pelvic floor.
- ▶ Involvement of base of the tongue in oropharynx is very rare and is yet to be described in the current literature.
- ▶ Patient can be successfully managed by coblation-assisted transcervical mandibular swing approach ensuring complete removal of the tumour with minimal postoperative morbidities.

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