CASE REPORT

Bulge in the tonsillar fossa. Is it a quinsy?

Ananth Vijendren,¹ Iman Eladawy,² Prasad Kothari²¹East and North Herts NHS Trust, Lister Hospital, Stevenage, UK²Luton and Dunstable University Hospital NHS Trust, Luton, UK**Correspondence to**
Ananth Vijendren,
shivan5@yahoo.com**SUMMARY**

A young girl presented to the ENT acute clinic with a persistent cough and a bulge in the left oropharynx. As there were no clinical signs suggestive of a quinsy, an MRI was performed that showed a large mass extending from the base of the skull to the epiglottis medialising the lateral and posterior pharyngeal walls. The patient subsequently underwent an excision biopsy which demonstrated a ganglioneuroma, however developed a left-sided Horner's syndrome and mild vocal cord palsy after.

BACKGROUND

Parapharyngeal space tumours are rare, comprising less than 1% of all head and neck neoplasms. The vast majority of them are benign (80%) and usually of salivary or neurogenic origin. Neurogenic tumours account for 30% of these infrequent masses, typically arising from the post-styloid space.¹ A ganglioneuroma is an uncommon type of neurogenic tumour, which in our patient presented in an unusual way.

CASE PRESENTATION

A 15-year-old girl, presented to her general practitioner (GP) with a non-productive persistent cough which had been ongoing for a few weeks. There was no associated dyspnoea, dysphagia or odynophagia and she was otherwise well in herself. On examination, the GP noticed a bulge in the left lateral oropharyngeal wall. Wanting to rule out a quinsy, the patient was referred to the ENT acute clinic on the same day. The girl was otherwise fit and well with no relevant medical problems, drug allergies or medications. She was a student, non-smoker, teetotaler and lived with her parents.

Further examination by ENT revealed a well-looking girl with no stridor, change of voice or trismus. The bulge noted by the GP was seen but the oropharynx otherwise appeared healthy. A flexible nasoendoscopy (FNE) examination revealed a smooth mass on the left side of the pharynx, extending from the base of the skull to the epiglottis and causing a rightward shift of the oropharyngeal structures.

INVESTIGATIONS

An MRI of the neck (figure 1) was organised for the outpatient, and it showed a large mass measuring 5.9 cm×2.2 cm×8.6 cm in the left retropharyngeal/parapharyngeal space. The lesion demonstrated atrophy and enhancement and had medialised the lateral and posterior pharyngeal walls while lateralising the internal carotid artery and jugular vein.

Figure 1 Coronal MRI scans ((A) T1-weighted and (B) T2-weighted postgadolinium contrast) showing a large mass (5.9 cm×2.2 cm×8.6 cm—green arrows) in the left retropharyngeal/parapharyngeal space. The lesion demonstrates atrophy and enhancement and has medialised the lateral and posterior pharyngeal wall while lateralising the internal carotid artery and jugular vein. Coronal MRI scan ((B) T2-weighted) showing a large mass (5.9 cm×2.2 cm×8.6 cm—green arrows) in the left retropharyngeal/parapharyngeal space. The lesion demonstrates atrophy and enhancement and has medialised the lateral and posterior pharyngeal wall while lateralising the internal carotid artery and jugular vein.

TREATMENT

After an extensive discussion with the patient and her mother, the decision for a surgical excision was made. This was to alleviate the patient's symptoms of persistent cough and to obtain a histological diagnosis. Eighty per cent of the mass was removed via transoral excision, as its close proximity to the base of skull and the great vessels of the neck made it difficult for a complete resection. The histology (figure 2) revealed a spindle cells matrix with interspersed mature ganglionic cells, consistent with a ganglioneuroma.

OUTCOME AND FOLLOW-UP

Postoperatively, the patient developed a left-sided Horner's syndrome which was treated conservatively. She was fed via a nasogastric tube for the first 48 h and then slowly started on a liquid and oral diet. She was discharged 3 days after her surgery.

A follow-up 3 months later revealed the persistent Horner's syndrome along with a weak voice. This was confirmed on FNE to have arisen from a mild left vocal cord palsy, which may have been damaged intraoperatively as it was a site of attachment of the tumour. A repeat MRI (figure 3) demonstrated a smaller ganglioneuroma, measuring

To cite: Vijendren A, Eladawy I, Kothari P. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2012-008147

Figure 2 Histopathology of lesion postsurgical excision revealing a spindle cells matrix with interspersed mature ganglionic cells (red arrow), consistent with a ganglioneuroma.

2.6×2×5 cm in the left carotid space, which was partially encasing the left internal carotid and displacing the left internal jugular vein laterally. There was no evidence of invasion into other head and neck structures.

The patient will be followed up every 3–6 months with repeat MRIs to assess the size, extent and location of her tumour.

DISCUSSION

Ganglioneuromas are benign tumours of the sympathetic nerve fibres that arise from neural crest cells. They usually present between the ages of 10 and 40 and are typically found in the adrenal medulla. These tumours are slow-growing and can release hormones and chemicals as part of their neuroendocrine nature. Ganglioneuromas form part of the neuroblastic tumour group which also consist of ganglioneuroblastomas (generally benign but can turn malignant) and neuroblastomas (malignant and aggressive tumours).²

These lesions are usually asymptomatic; however, they can cause compressive symptoms depending on their size and

location. Lesions in the chest may result in chest pain and dyspnoea, whereas abdominal masses can present as abdominal bloating or pain. Head and neck lesions, on the other hand, usually present as a neck lump or as an incidental finding as in the patient.²

The imaging modalities of choice to investigate these masses very much depend on their location. CT scans are particularly useful for ganglioneuromas in the chest and abdomen to identify the tissue of origin and tumour size, whereas MRI scans tend to be performed for lesions in the head, neck and spine to assess tumour invasion and extension.¹ Blood and urine tests for catecholamines may be performed to determine if the tumour is actively producing hormones or chemicals. A biopsy is essential to confirm the diagnosis and to differentiate between tumours within the neuroblastic tumour group.²

The management of ganglioneuromas in the head and neck is a balance between watchful waiting and surgical excision, as most of these lesions are asymptomatic and surgery can carry significant morbidity. Song *et al* looked at six patients who had ganglioneuromas excised from the neck between 1995 and 2009 and were subsequently followed up for a median of 5.9 years. All patients had developed Horner's syndrome, most likely from sympathetic chain damage, but had no local recurrence or distant metastases.³ Retrosi *et al*, on the other hand, studied the general morbidity postganglioneuroma surgery in 23 children. All the children had biopsy-proven ganglioneuromas from multiple locations that were excised between 1998 and 2009. Nine children were asymptomatic on presentation with the rest having respiratory symptoms or abdominal pain. Seven patients had developed significant complications which included three Horner's syndrome, one chylothorax, one pneumothorax, one arm pain and one adhesive intestinal obstruction.⁴ Hence, a biopsy beforehand may play an important role in identifying the nature of these lesions which can aid in the patient's decision for further management.

Although surgical excisions for these head and neck lesions have traditionally been performed transorally or via an external approach, new methods have recently surfaced that could improve postoperative outcomes. A recent paper in the *European Archives of Otolaryngology* demonstrated transoral robotic resection of three benign and malignant tumours from the parapharyngeal space where complete tumour excision was achieved without any complications.⁵ Similarly, Yokoi *et al* had shown a novel technique of a transoral ganglioneuroma excision using a Weerdal laryngoscope which was found to be hugely beneficial.⁶

Figure 3 Coronal MRI scans 3 months later ((A) T1-weighted and (B) T2-weighted postgadolinium contrast) showing a smaller soft tissue mass (2.6 cm×2 cm×5 cm—green arrows) in the left carotid space partially encasing the left internal carotid and displacing the left internal jugular vein laterally. There is no evidence of invasion into the parotid gland, vertebral body or the pre-vertebral musculature. Coronal MRI scans 3 months later (A T1-weighted and B T2-weighted postgadolinium contrast) showing a smaller soft tissue mass (2.6 cm×2 cm×5 cm—green arrows) in the left carotid space partially encasing the left internal carotid and displacing the left internal jugular vein laterally. There is no evidence of invasion into the parotid gland, vertebral body or the prevertebral musculature.

Learning points

- ▶ Ganglioneuromas are rare benign neuroendocrine tumours which can present in the parapharyngeal space.
- ▶ The majority of these tumours are asymptomatic.
- ▶ The decision for surgical excision in the head and neck should be carefully weighed and discussed with the patient as it can result in significant morbidity. A biopsy beforehand may be vital.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Medscape. Parapharyngeal Space Tumours. <http://www.emedicine.medscape.com/article/849385-overview> (accessed Sep 2012).
- 2 PubMed Health. Ganglioneuroma. <http://www.ncbi.nlm.nih.gov/pubmedhealth/PMH0002409> (accessed Sep 2012).
- 3 Song Y, Liu H. Management of ganglioneuroma in the neck. *J Clin Otorhinolaryngol Head Neck Surg* 2012;26:392–3.
- 4 Retrosi G, Bishav M, Kiely EM, *et al.* Morbidity after ganglioneuroma excision: is surgery necessary? *Eur J Paediatr Surg* 2011;21:33–7.
- 5 Arshad H, Durmus K, Ozer E. Transoral robotic resection of selected parapharyngeal space tumors. *Eur Arch Otorhinolaryngol* 2012 [Epub ahead of print]
- 6 Yokoi H, Arakawa A, Inoshita A, *et al.* Novel use of Weerda laryngoscope for transoral excision of a cervical ganglioneuroma. *J Med Case Rep* 2012;26:88.

Copyright 2013 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow