CASE REPORT

Dilemma in the Treatment of Paediatric Vagal Paraganglioma

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ABSTRACT

Vagal paraganglioma is a rare tumour of the head and neck region, particularly in the paediatric age groups. A 13-year-old girl presented at the Department of Otorhinolaryngology, Head & Neck Surgery, Universiti Kebangsaan Malaysia Medical Centre Malaysia with painless left neck swelling, associated with multiple cranial nerve palsies for 14 months. The neck swelling was diagnosed as a carotid body tumour initially, but it was deemed inoperable, and the child received radiotherapy instead. The treatment response was suboptimal. Upon re-evaluating the radiological images, a revised diagnosis of vagal paraganglioma was made. Her family refused surgical intervention, and the child was treated conservatively. Over the last four years, serial imaging has demonstrated a decrease in tumour size. Although vagal paraganglioma in paediatric may require surgical excision, there is still a role in the wait-and-see approach. Frequent follow-up with radiological surveillance can save patients from surgical morbidity.

INTRODUCTION

Vagal paraganglioma is a rare nonchromaffin, extra-adrenal paraganglioma that arises from the nodose ganglion of the vagus nerve.[1] It represents less than 5% of all head and neck paragangliomas, commonly seen in mid-adult life with average onset at 46 years old. Although the tumour is benign and slow-growing, it may later cause isolated vagus nerve palsy or multiple lower cranial nerve palsies, especially cranial nerve IX, XI and XII, as these nerves are close to the vagal body. The tumour can potentially extend into the skull base or intracranially, making surgical excision more challenging. Despite inevitable post-operative speech and swallowing deficit due to iatrogenic nerve injury or sacrifice, surgery is still the mainstay of treatment.[2] However, the timing of surgery may differ according to the patients.

The suitability of surgical excision in paediatric vagal paraganglioma remains uncertain.[2,3] We encountered a case of vagal paraganglioma in a child who presented with multiple cranial nerve palsies. To our knowledge, literature evidence in managing symptomatic paediatric vagal paraganglioma is non-existence, which has caused a dilemma in our clinical decision making.

CASE REPORT

A 13-year-old girl with no known medical illness presented at the Department of Otorhinolaryngology, Head & Neck Surgery, Universiti Kebangsaan Malaysia Medical Centre Malaysia for painless left neck swelling for 14 months. The neck swelling was progressively increasing in size, and she started to develop persistent dysphagia one month before her presentation. She denied any voice change and shortness of breath. There was no loss of appetite, unintentional weight loss or family history of malignancy. On examination, she had a soft and diffuse swelling over her left neck lymph node level II. The overlying skin was normal, with no carotid bruit heard on auscultation. Intraoral examination showed left-sided tongue fasciculation. On flexible

laryngopharyngoscopy, the left vocal fold was immobile and fixed at the paramedian position.

The initial magnetic resonance imaging (MRI) neck showed a large well-defined soft tissue mass encasing the common carotid at the carotid bifurcation level. The mass is insinuated between the bifurcation, causing splaying of the internal and external carotid arteries, measuring 3.2cm x 3.6cm x 7.1 cm (Figure 1-A). It extended superiorly just below the base of the skull. Post gadolinium contrast, the mass was enhanced (Figure 1-B). The computed tomography angiography (CTA) displayed an avid contrast enhancement mass with no abnormal aneurysm seen (Figure 1-C). Based on these investigations, she was diagnosed with a left carotid body tumour at that time.

The tumour was deemed inoperable, and she was treated with 50Gy over 25 fractions of radiotherapy. However, the repeat CTA showed no significant tumour size changes with increasing necrosis. Therefore, she was referred to our centre for a second opinion. A revised diagnosis of vagal paraganglioma was made after our radiologists reviewed the MRI.

The parents were not keen on surgical excision as yet, mainly because they have seen some reduction in tumour size. Hence the lesion was observed closely for the last four years with annual MRI surveillance. The latest MRI demonstrated a 48% volume reduction of the mass size (Figure 1-D). The dysphagia and neck swelling had resolved on recent follow-up, and she did not have any other new cranial nerve deficits.

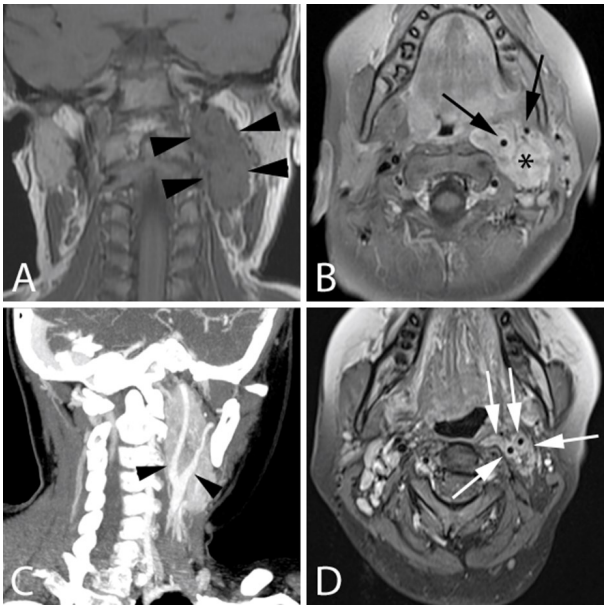

Figure 1: A represents a coronal T1 weighted neck MR image at the initial presentation. B is an axial fat suppressed T1-weighted post Gadolinium neck MR image, also at initial presentation. C is a CT angiographic image of the neck in maximum intensity projection (MIP) technique and parasagittal oblique reconstruction, showing the left carotid bifurcation in profile, 2 months post initial presentation. D represents a latest MR neck image in axial fat suppressed T1-weighted post Gadolinium sequence, 4 years after initial presentation. The well circumscribed mass located within the left carotid space is fusiform in shape and located high in the neck, just inferior to the skull base (black arrowheads in A). It causes partial filling of the left parapharyngeal space. Its superior margin does not extend into the ipsilateral jugular fossa. It is intensely enhancing (B and C). Several serpentine flow voids (asterisk in B) are identified within the mass (best seen in B, subtly appreciated in A). The mass does extend to the carotid bifurcation (and for a short distance below the bifurcation, C). Note the slight splaying apart of the left internal and external carotid arteries (black arrows in B, black arrowheads in C). The normal right internal and external carotid arterial distance in B may be used as a comparison .

DISCUSSION

Paraganglioma in the paediatric-age group is unusual. The incidence is estimated between 10% to 20% out of 0.3 cases per million per year.[4] Therefore, paediatric vagal paraganglioma is even rarer. The youngest patient reported previously was a 16-year-old, according to the most extensive vagal paraganglioma series in the United States of America by Netterville et al. However, the treatment and the outcome of the patient were not detailed in the study.[2] Jansen et al. described the wait-and-scan strategy for head and neck paragangliomas as feasible in patients younger than 50 years old. The study also found that vagal paragangliomas slowly grew at 10.9mm per year, and young age was not associated with higher tumour-induced complications.[5] Nevertheless, the short follow-up period inhibits the study from observing long-term complications such as malignant transformation. Vagal paraganglioma malignant transformation is reported between 16% to 19%, the highest of all head and neck paraganglioma.[6]

MRI is the most critical imaging tool in diagnosing head and neck paraganglioma as it can differentiate the tumour from its surrounding soft tissue and other vascular structures.[3] Paraganglioma vascularity causes a flowing void effect, typically illustrated as a "salt and paper" appearance in the MRI. Most vagal paraganglioma has an irregular shape, displaces the internal carotid artery anteriorly and usually, involves the jugular foramen.

Carotid body tumour has a more regular shape, causes splaying of the carotid bifurcation, and tends to displace the internal carotid artery posteriorly.[7] In our patient, the mass was located high in the neck, just inferior to the skull base. The carotid splaying is not sufficiently wide apart to form the classic lyre sign usually encountered in a carotid body tumour. This pattern of vascular splaying, the location and configuration of the mass, and the presence of flow voids and intense enhancement are considered compatible with a left vagal paraganglioma.

The management of vagal paraganglioma is not well established due to limited quality data. Sakthivel et al. suggested surgery for young patients as tumour growth is expected, but it is not without complications. Surgical morbidity such as lower cranial nerve palsies is high (23% to 61%) and often requires long-term post-operative speech and swallowing rehabilitation. Other complications include stroke, meningitis and cerebrospinal fluid leak.[8] These drawbacks might cause hesitance for surgeons and parents to opt for surgery in children. Watchful waiting in young patients should be altered with a shorter interval of follow-up and surveillance MRI to assess the tumour behaviour closely.[5] Other non-surgical approaches include radiotherapy, but its application in paediatrics has raised concerns about long-term side effects and tumour breakout growth. The Glomus cell itself is resistant to radiotherapy. Although the tumour size can regress, as demonstrated in this case, it does not eliminate it.[8]

CONCLUSION

Vagal paraganglioma in paediatric populations is more likely to grow and eventually require surgical excision, but this may take years. Hence, careful study of the imaging and discussion with parents are essential in the management. A non-surgical approach to vagal paraganglioma is age-specific, where paediatric cases require frequent follow-up and serial imaging compared to the elderly, but there is still a definite role. Therefore, an excellent multidisciplinary team is required while managing this condition conservatively.

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