

Case Report

Cervical ganglioneuroma: a case report

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ABSTRACT

Cervical ganglioneuroma (CGN) is a rare benign tumour that originates from the neurogenic soft tissue of the cervical sympathetic chain. It is part of the neuroblastic tumours that arise from the neural crest cells of the central or peripheral nervous system. The clinical presentation can range from a palpable mass in the neck, dysphagia, dysphonia, or features of Horner's syndrome. We present a case of a 65-year-old lady that was seen in the ENT outpatient clinic with complaints of left-sided neck swelling, dysphagia, and dysphonia. Complete surgical excision of the mass revealed the histological diagnosis of CGN. Treatment is considered complete when the entire specimen is excised.

Keywords: CGN, Neuroblastic tumour, Benign tumours, Ganglioneuroma

INTRODUCTION

Ganglioneuromas (GNs) are a type of benign tumours of the neuroblastic family that originate from the central or peripheral autonomic nervous system. The most common sites are the posterior mediastinum, retroperitoneum, adrenal glands and less frequently, the neck. CGN s account for less than 8% of all ganglioneuromas.¹ They are more common in children with variable ages at presentation. There are no known risk factors for the development of ganglioneuromas, however, isolated ganglioneuromas might be syndromic, specifically with multiple endocrine neoplasia type IIb and Neurofibromatosis type 1. CGN s are benign, slow-growing tumours with low malignant transformation potential, and the mainstay of management is complete surgical excision.

CASE REPORT

A 65-year-old lady attended the urgent ENT cancer-suspected pathway outpatient clinic with a 2-month history of left-sided level 6 neck swelling, dysphagia, and

dysphonia. She denied having B-type symptoms including fever, night sweats or unintentional weight loss. She had no previous head and neck diseases, and no previous irradiation therapy to the area. She was a non-smoker.

On examination, a 20×20 mm neck mass was palpable in left level 6, which was non-tender and immobile. Oral cavity, oropharynx, and flexible fiberoptic laryngoscopy were normal. The patient subsequently had a neck ultrasound which revealed a well-defined, hypoechoic lesion on the left side of the neck between the trachea, carotid artery, and thyroid gland, measuring 21×13×32 mm in size (Figure 1). The lesion showed some calcifications with no appreciable internal vascularity or cervical lymphadenopathy. The ultrasound differential diagnosis was initially of a parathyroid mass or enlarged lymph node origin. A fine needle aspiration cytology was performed which was inconclusive. Thyroid function testing, Serum calcium, parathyroid hormone and calcitonin were normal. A subsequent non-contrast magnetic resonance imaging of neck and chest revealed a well-circumscribed lesion situated adjacent to the lower

pole of the left lobe of the thyroid gland (Figure 2). The thyroid gland had a normal size and shape with no evidence of lymphadenopathy.

The case was discussed at the head and neck multidisciplinary team meeting as there was uncertainty regarding the diagnosis. The recommendation was for surgical excision of the lesion. Intraoperatively, the lesion was well-circumscribed and appeared benign, the decision was taken to perform a simple level 6 lumpectomy. The lesion was distinct from any thyroid and parathyroid tissue. The tumour was in the level 6 neck directly under the recurrent laryngeal nerve and extended posteriorly behind the trachea (Figure 3 and 4). Post-operative histopathology analysis showed an encapsulated, circumscribed soft tissue nodule composed of bland spindle cells with areas of degenerate change. No atypia or mitotic figures were noted. On immunohistochemistry, the tumour cells were strongly positive for S-100; weak positivity for synaptophysin, and negative for CD34, SMA and desmin. These features were consistent with ganglioneuroma. The patient was reviewed post-operatively at 6-month intervals for 3 consecutive years, with no evidence of recurrence and resolution of symptoms.

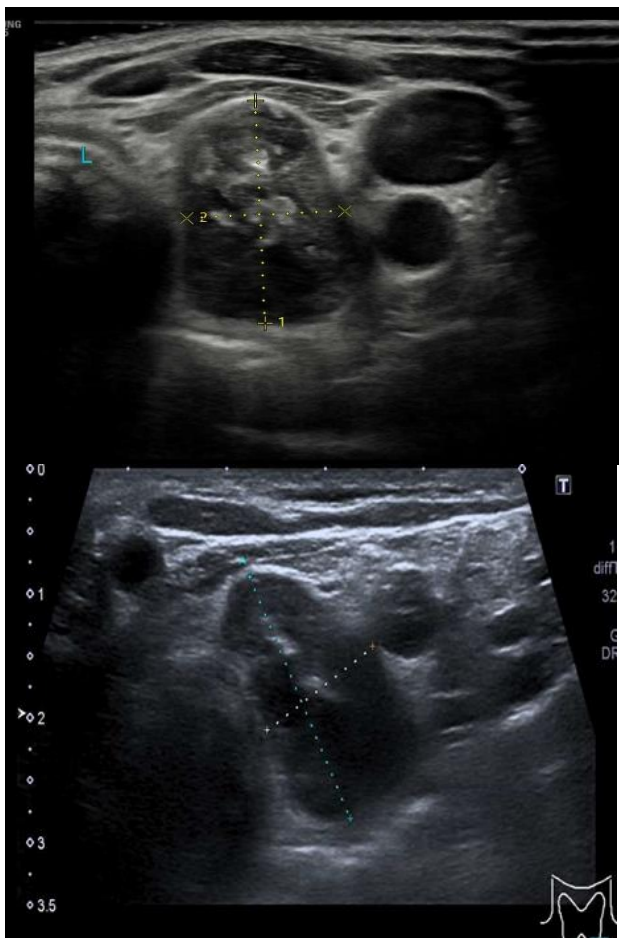


Figure 1: Ultrasonography showing a well-defined, hypoechoic lesion measuring 21×13×32 mm in size in the left side of the neck.

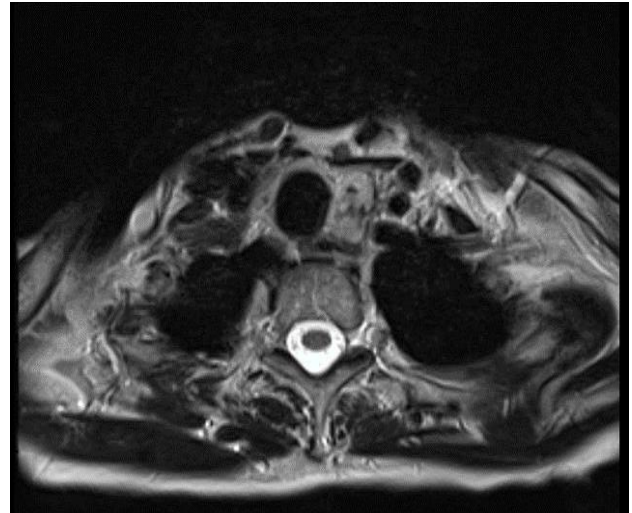


Figure 2: Non-contrast magnetic resonance imaging of the neck and chest with axial cut revealing a well-circumscribed lesion situated adjacent to the lower pole of the left lobe of the thyroid gland.

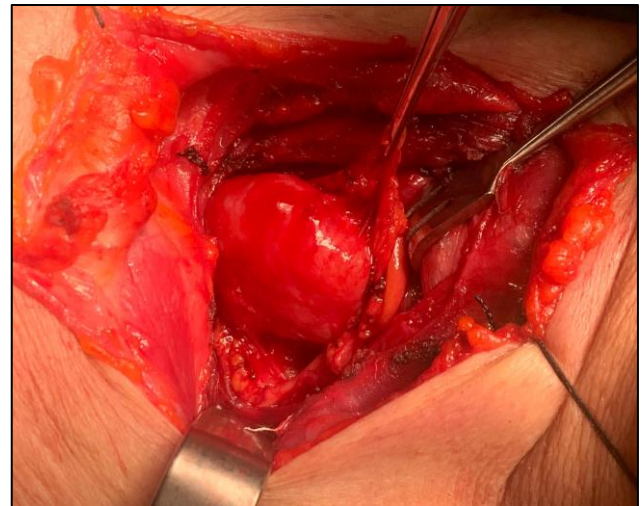


Figure 3: Intra-operative images revealing a well-circumscribed right-sided neck lesion.



Figure 4: Surgical specimen completely excised.

DISCUSSION

The most common location of CGNs is the cervical sympathetic chain, however, the larynx, pharynx, and ganglion nodosum of the vagus nerve have been reported.² They are often asymptomatic, but if they grow to a considerable size, they can begin to compress vital structures in the neck. If compression affects the sympathetic nervous system, then features of Horner's syndrome may be present. CGNs are more common in the paediatric population, with 60% of cases arising in patients under the age of 20.³

Definitive diagnosis of CGN is with complete excision of the mass allowing formal histological assessment. Complete surgical excision provides symptomatic improvement for the patient. Various imaging modalities such as computed tomography and magnetic resonance imaging can aid in the assessment of the location, size, and consistency of the mass. It can also aid surgical planning. Ultrasound-guided biopsies can lead to inconclusive tests, thus fine needle aspiration is not recommended as a primary diagnostic tool for CGN.⁴

A literature review identified 36 case reports for CGNs. Two reports showed tumours mimicking tumours of the thyroid gland (5.56%). Twenty-two of the reported cases (61.11%) were in the paediatrics population (below 18 years of age), while 14 patients (38.89%) were over 18 years old. Male to female preponderance was roughly equal with 20 cases in females (55.56%) and 16 patients were males (44.44%). Most of head and neck ganglioneuromas arise from the sympathetic chain. Ten cases reported postoperative Horner's syndrome of which was either partial or complete and temporary or permanent. One patient was reported to have a postoperative vocal cord paralysis.² In this case report, no post operative complications were reported.

There are different surgical approaches for the excision of CGN. The location and size of the ganglioneuroma are deciding factors for surgical technique. Simple excision of the mass via a transcervical incision is possible for small, well-circumscribed lesions. If complete surgical excision is not possible, debulking procedure is indicated, with regular follow-up to ensure no progression of symptoms or disease.⁵ Different approaches have been reported in the literature, including transoral approach without mandibulectomy.⁶

Management of CGN is the complete surgical excision of the neck mass, with serial follow-up to monitor symptoms, post-operative complications or recurrence of the disease.

CONCLUSION

CGNs are the least common type of ganglioneuromas that can occur in the body. They are benign tumours that are generally slow-growing. Definitive diagnosis is by complete surgical excision of the lesion with histopathological assessment. Treatment is considered complete with surgical excision. Follow-up is recommended to assess for any recurrence or post-operative complications.

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