Jayavelu *et al.*
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CASE REPORT Open Access

Massive ganglioneuroma of the parapharyngeal space in a pediatric patient: a case report

Swetha Jayavelu[1*], Raleigh Anderson[1], Garren Low[2], Karim Asi[1] and Soham Roy[1]

Abstract

Background: Neuroblastic tumors arise from primitive sympathetic ganglion cells and are the most common extracranial solid tumor of childhood. Ganglioneuroma is the most well-differentiated, benign subtype and comprises less than 1% of all soft tissue tumors. One to 5% of these occur in the head and neck, including the parapharyngeal space. It is uncommon to find a physically detectable, neoplastic parapharyngeal space mass in a child.

Case report: A 7-year-old boy presented with a painless left neck mass. Imaging revealed a 2.6 × 2.7 × 6.1 cm left parapharyngeal space mass. Fine-needle aspiration suggested a ganglioneuroma. The patient developed compressive symptoms, notably pain, thus the decision was made to surgically resect. The tumor was excised transcervically, preserving the great vessels, phrenic and cranial nerves. Postoperatively, the patient developed a mild ipsilateral Horner's syndrome, suggesting the mass to be a ganglioneuroma of the sympathetic chain.

Conclusions: Cervical ganglioneuromas typically present as slow-growing masses that cause compressive symptoms or are found incidentally. Resection is reserved for those with significant symptoms and is often complicated by Horner's syndrome. In the review of literature, there are 23 reported cases of ganglioneuroma in the head/neck with a median age of 17 years. Eight of these occurred in the para/retropharyngeal spaces. Of these, five presented as an asymptomatic neck mass, two presented with compressive symptoms, and one was discovered incidentally. It is important to consider ganglioneuroma in the workup of pediatric neck masses, even in young children. Asymptomatic masses should be monitored for symptomatic transformation, and excision should be considered.

Keywords: Pediatric neck mass, Benign cervical mass, Ganglioneuroma, Neuroblastic tumor, Neurogenic tumor, Case report

Background

Neuroblastic tumors arise from primitive sympathetic ganglion cells and are the most common extracranial solid tumor of childhood [1]. There are three recognized types—neuroblastomas, ganglioneuroblastoma, and ganglioneuromas. These tumors vary in their proportions of Schwann cells and neuroblasts. Classification is based on histological features including the degree of differentiation and cellular maturation. Neuroblastomas are aggressive and poorly differentiated, with decreased Schwann cells in the stroma. Ganglioneuroblastomas vary in the degree of Schwann cells but generally include an intermixed picture of Schwann cells and neuroblasts. The subtype, ganglioneuroma, is the most well-differentiated, with a dominant presence of Schwann cells in the stroma [1, 2]. This subtype is the most benign and comprises less than 1% of all soft tissue tumors. It most commonly occurs in the adrenal medulla, extra-adrenal retroperitoneum, and posterior mediastinum. One to 5% of these extremely uncommon tumors occur in the head and

*Correspondence: swetha.jayavelu@uth.tmc.edu

[1] Department of Otolaryngology, University of Texas Health Science Center at Houston McGovern Medical School, 6431 Fannin St, Houston, TX 77030, USA

Full list of author information is available at the end of the article

neck, with the parapharyngeal space (PPS) being a potential subsite [3, 4].

The PPS is a triangular compartment lateral to the upper pharynx composed primarily of fat, with the apex of the triangle at the hyoid and the base at the skull. Given its deep location, most PPS masses are not detectable on physical examination [5, 6]. Furthermore, while parapharyngeal masses are most commonly neoplastic in adults, these masses tend to have infectious causes in children [7]. It is quite uncommon to find a physically detectable, neoplastic parapharyngeal mass in a pediatric patient. We present the case of a 7-year-old boy with a large ganglioneuroma of the PPS and provide an updated review of the literature on the presentation of ganglioneuromas.

Case presentation

We report a 7-year-old boy with a history of recurrent otitis media who presented with 2 months of cervical lymphadenopathy. There has been no family history of this presenting condition. Ultrasound suggested reactive lymphadenopathy. Lymphadenopathy persisted after 10 days of clindamycin; thus, a CT was obtained. This revealed a 2.6 × 2.7 × 6.1 cm mass in the left PPS extending from the mandible to the thyroid, with a displacement of the internal jugular vein and common carotid artery (Figs. 1 and 2). A specimen obtained via fine-needle aspiration stained positive for S100 with negative SMA in the spindle cells, supporting the diagnosis of ganglioneuroma. MRI was performed for further characterization (Figs. 3 and 4). There were no diagnostic challenges to be reported. The patient began to experience neck pain and odynophagia; thus, we proceeded with surgical excision.

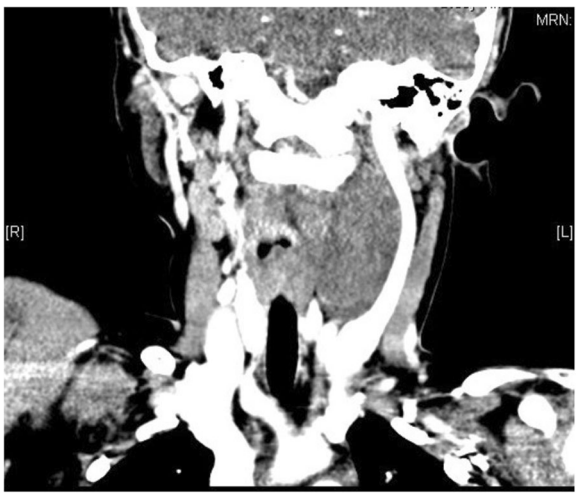

Fig. 2 CT coronal view

Complete excision of the tumor was performed via neck dissection. The tumor was beneath the vagus nerve, which was skeletonized, preserved, and remained intact on intraoperative nerve stimulation. The carotid artery and internal jugular vein were preserved along with their branches. The tumor was noted to be attached to cervical nerve rootlets. The nerve rootlets of cranial nerves XI and XII and phrenic nerves were stimulated and noted to be intact, which established that the tumor was not related to these nerve roots.

Postoperatively, the patient developed a mild left Horner's syndrome (HS). All other cranial nerves were intact. The final pathology confirmed the mass to be a

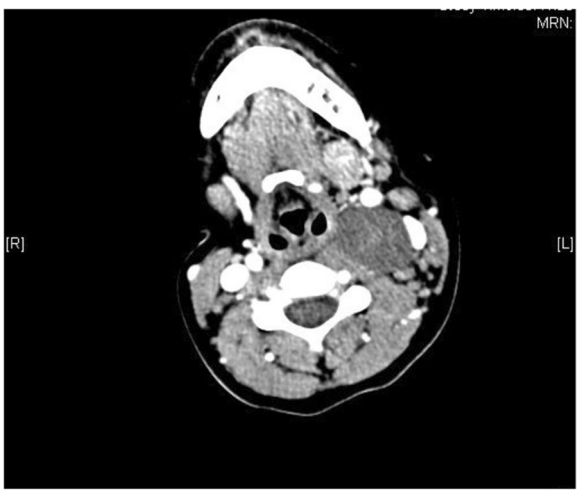

Fig. 1 CT axial view

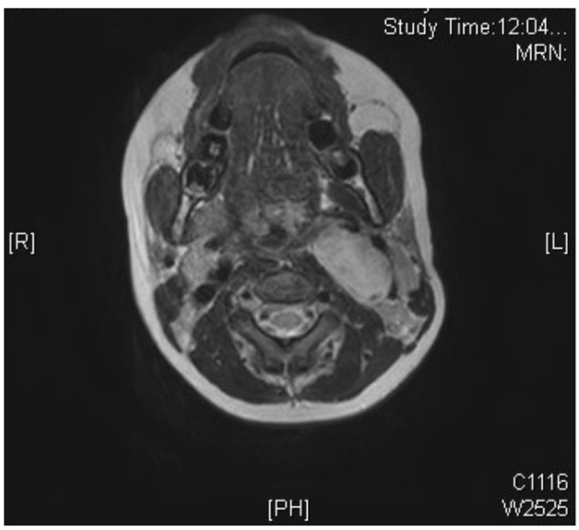

Fig. 3 MRI T2 axial view

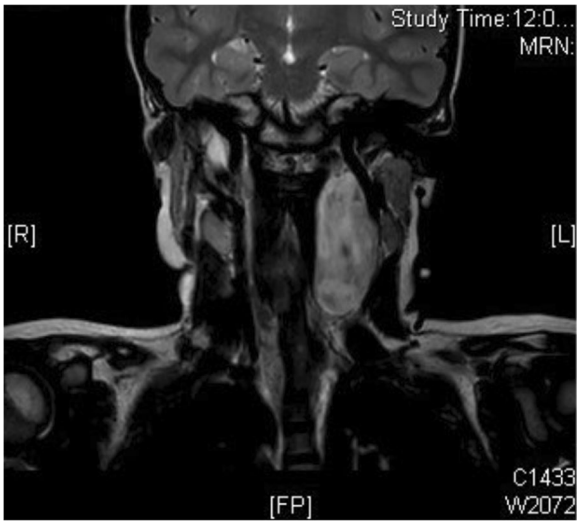

Fig. 4 MRI T2 coronal view

ganglioneuroma. The patient's postoperative HS without other neuropathies suggests the mass to be a ganglioneuroma of the cervical sympathetic chain.

Discussion

Ganglioneuromas are benign, non-invasive tumors of the neural crest cells that give rise to the sympathetic nervous system. These tumors most commonly occur in the trunk; however, 1–5% occur in the head and neck [4]. Cervical ganglioneuromas commonly present as slow-growing masses that cause compressive symptoms or as incidental findings on imaging. It has been reported that these tumors can cause symptoms related to catecholamine release; however, this was not present in our case nor in any reviewed cases of the last 15 years. Patients most commonly present in childhood or early adulthood, with management usually consisting of regular monitoring. Surgical resection is reserved for symptomatic patients. Recurrence is rare, with one incidence recorded in the literature. Resection is commonly complicated by self-limited postoperative HS due to disruption of the sympathetic cervical chain [8–15].

Here, we present a 7-year-old male with an initially asymptomatic neck mass that subsequently became associated with neck pain and odynophagia. The patient was found to have a parapharyngeal ganglioneuroma that was surgically excised, with the only complication being mild ipsilateral HS. This presentation and treatment outcome is consistent with other sympathetic chain parapharyngeal ganglioneuromas presented in the literature [8–13, 15]. This presentation is not universal, however varying based on the location of the lesion and the age of the patient.

Table 1 Prevalence of presenting symptoms in the reviewed literature

Symptoms	Frequency
Neck mass	9
Dysphagia	3
Weakness	3
Odynophagia	2
Hearing loss	2
Incidental finding	2
Neck pain	2
Proptosis	1
Glaucoma	1
Cough	1
Neuralgia	1
Neck stiffness	1

In our review of the literature, there are 23 reported cases of ganglioneuroma affecting the head and neck. Sixteen of 23 occurred in patients under the age of 30 with a median age of 17, consistent with reported distributions of incidence [14]. Of the 16 cases in patients under 30, 9 presented with a chief complaint of a neck mass, making it the most common presentation in that group (Table 1). Most of these presented in the absence of other symptoms. There were no cases of patients over 20 years of age presenting with a discernible neck mass [8–30]. Of the 23 reported cases reviewed, 7 presented with postoperative Horner's syndrome (about 30%). Of those, 4/7 cases of postoperative Horner's syndrome resolved [8–13, 15].

Interestingly, patients with ganglioneuromas in the para- or retropharyngeal spaces more commonly presented with an identifiable neck mass. In our review, 8 patients were found to have para- or retropharyngeal masses, with 5 of the 8 initially presenting with a neck mass. Two of the other patients presented with symptoms of compression: cough, neck pain, and stiffness. The third was discovered incidentally on imaging. The second most common subsite was prevertebral and cervical spine with 6 cases, with other reported cases in the submandibular space, external and internal auditory canals, uveal tract, base of the tongue, superior orbit, inferior jugular region, and the glossopharyngeal tract [8–30].

Conclusions

Our case illustrates a key example of neurogenic tumor presentation, with a pediatric patient presenting with an initially asymptomatic neck mass that ultimately caused compressive symptoms, all in the absence of systemic signs or tissue invasion. Ganglioneuroma is an important consideration in the diagnosis of a neck mass in a child,

especially after more common etiologies, including infection, lymphoma, and benign congenital masses, have been considered. Children with a ganglioneuroma may commonly present with no signs of compression or systemic signs but still must continue to be monitored for symptomatic transformation with continued growth and advised on the risks and benefits of excision.

Abbreviations
PPS: Parapharyngeal space; HS: Horner's syndrome.

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Authors' contributions
SR and GL designed the concept of the work and substantially revised the writing. RA performed the literature review and designed Table 1. KA and SJ were major contributors to the writing of the manuscript. All authors read and approved the final manuscript.

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Not applicable

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Competing interests
The authors declare that they have no competing interests.

Author details
[1]Department of Otolaryngology, University of Texas Health Science Center at Houston McGovern Medical School, 6431 Fannin St, Houston, TX 77030, USA. [2]Present address: Department of Otolaryngology, Oregon Health and Science University, Portland, OR 97239, USA.

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