Retropharyngeal Ganglioneuroma Presenting with Neck Stiffness: Report of a Case and Review of Literature

Celeste Gary, B.Sc.,¹ Hugh Robertson, M.D.,² Bernardo Ruiz, M.D., Ph.D.,³ Vladimir Zuzukin, M.D.,¹ and Rohan R. Walvekar, M.D.¹

ABSTRACT

Ganglioneuromas rarely occur in the retropharynx with only three cases reported in the current literature. The most common symptom associated with retropharyngeal ganglioneuromas is dysphagia. We report a retropharyngeal ganglioneuroma with an unusual clinical presentation of neck stiffness and pain. A 42-year-old woman presented with incapacitating neck pain, neck stiffness, right upper extremity weakness, as well as dysphagia. Neurological workup was normal. Imaging revealed a hyperdense, ill-defined, diffuse right retropharyngeal mass suggestive of a possible nerve sheath tumor with no communication with the cervical spine. Surgical removal was uneventful and associated with a postoperative Horner's syndrome. In follow-up, dysphagia and neck symptoms improved. Retropharyngeal ganglioneuromas can occur in a wide age range of patients. Surgical excision via a cervical approach offers definitive therapy but may be associated with an iatrogenic Horner's syndrome for which the patients should be counseled prior to operative intervention. Neck pain is an atypical symptom that needs to be worked up to rule out a communication with the spinal column prior to surgical removal. Patients must be counseled that atypical symptoms may not completely resolve with surgical treatment.

KEYWORDS: Retropharyngeal, ganglioneuroma, neck pain, dysphagia

Ganglioneuromas are benign tumors that originate from primordial neural crest cells which migrate from the mantle layer of the developing spinal cord to the sympathetic ganglia, adrenal medulla, and other sites. These tumors are well-circumscribed and consist of ganglion cells and Schwann cells with few neuroblasts. Ganglioneuroblastomas are malignant variations of these tumors that are characterized by decreased differentiation within ganglion cells in the presence of Schwann cells and are also associated with neuroblasts. Ganglioneuromas occur most commonly in children over 10 years of age.

The most frequent sites of presentation include the posterior mediastinum and the retroperitoneum. In general, the neck is a rare site of presentation.¹ In a series of 88 patients with ganglioneuromas, only two occurred in the cervical region.² The retropharyngeal area of the neck is an extremely rare site of presentation with only three reported cases in the literature.^{3–7} The most common symptom of presentation was dysphagia. We are reporting a retropharyngeal ganglioneuroma presenting with an unusual symptom complex that consisted primarily of neck stiffness, neck pain, and dysphagia.

¹Department of Otolaryngology–Head Neck Surgery; ²Department of Radiology; ³Department of Pathology, LSU Health Sciences Center, New Orleans, Louisiana.

Address for correspondence and reprint requests: Rohan R. Walvekar, M.D., Assistant Professor, Department of Otolaryngology–Head Neck Surgery, LSU Health Sciences Center, 533 Bolivar Street, Suite 566, New Orleans, LA 70112 (e-mail: rwalve@lsuhsc.edu).

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Figure 1 (A) MRI T2 coronal image hyperintense to muscle with distinct margin. (B) T1 coronal image with tumor isointense to muscle. (C) T1 coronal image with fat saturation and intravenous gadolinium contrast medium with moderate tumor contrast enhancement. (D) T2 axial image with tumor hyperintensity (white and black arrows defining the extent of the tumor).

CASE REPORT

A 42-year-old Caucasian female presented to our service with incapacitating neck stiffness and pain. The patient also reported difficulty swallowing and a change in her voice. Physical examination included a complete neurological evaluation, assessment of cranial nerve function, as

well as head and neck evaluation with flexible fiber-optic laryngoscopy. To further investigate her symptoms, a CT and MRI of the neck and spine were performed. The MRI demonstrated a hyperdense lesion with T2 enhancement in the right retropharynx. This mass was diffuse and ill-defined which suggested the possibility of a slow-growing

Figure 2 (A) Ganglioneuroma composed of clusters of ganglion cells within a neurofibrous stroma with a tenuous pseudocapsule. (B) Clusters of ganglion cells within the ganglioneuroma.

nerve sheath tumor with no communication with the cervical spine (Fig. 1A–D). On presentation to our clinic, examination of the neck was unremarkable with no palpable neck mass. A flexible fiber-optic laryngoscopy did not reveal an abnormal posterior pharyngeal wall bulging of the right as compared with the left, pooling of saliva, or abnormal vocal cord appearance or function. The past medical, surgical, and social history was non-contributory. The tumor was excised via a cervical route for diagnosis and curative treatment after obtaining an informed consent. Possible complications that included neurological deficits associated with resection of the vagus nerve, sympathetic chain or their branches were explained to the patient in view of a possible neurogenic origin of the tumor based on imaging studies.

The mass was free of the carotid artery, internal jugular vein, and vagus nerve. However, the tumor seemed to be originating from the cervical sympathetic trunk. Gross examination of the specimen revealed a soft, oblong, $5.5 \times 3.2 \times 1$ cm, well-circumscribed mass surrounded by a tenuous pseudocapsule. Microscopically, ganglion cells were identified scattered on a neurofibrous stroma. A suggestion of immaturity was observed consisting of areas where the ganglion cells showed discrete variability in size and were surrounded by neuronal processes unassociated with Schwann cells. Rare scattered atypical cells and a single discrete aggregate of atypical cells were observed which may have corresponded to neuroblasts. The final pathology was reported as ganglioneuroma, maturing subtype (Fig. 2A, B). In follow-up, the patient did well with improvement in neck symptoms and dysphagia. The patient also developed an asymptomatic postoperative Horner's syndrome.

DISCUSSION

Ganglioneuromas are rare, benign, neurogenic tumors that originate from the sympathetic chain. Ganglioneuromas are most frequently diagnosed in patients between

the ages of 10 and 29 years and are most commonly located in the posterior mediastinum followed by the retroperitoneum.² Among the documented cases of cervical ganglioneuromas, retropharyngeal ganglioneuromas have only been reported in three patients in the current literature (Table 1).^{3–7} These tumors were reported in patients from ages 16 months to 57 years. Our patient presented to us at 42 years. The age range of presentation seems to differ for ganglioneuromas located in the retropharynx as opposed to other sites. Consequently, it would be reasonable to consider this differential diagnosis in patients that present with a retropharyngeal mass irrespective of age criteria.

The most common presenting symptom for patients with retropharyngeal ganglioneuromas is dysphagia as was seen in our patient.^{3–7} Our patient, however, primarily presented with an unusual symptom of neck stiffness and pain. The differential diagnosis of neck pain includes cervical arthritis, discogenic disorders, trauma, tumors or infection, cervical myositis, myofascial pain syndrome, torticollis, whiplash, scalenus syndrome, as well as spinal cord tumors.^{8,9} Although neurogenic tumors of the neck are relatively infrequent, they should be considered in the differential diagnosis of a neck pain as is evident from our case. In our case, etiology of the neck pain could be attributed to a radiculopathy associated with compression of cervical spine by the tumor or a spasm of the prevertebral muscles in reaction to the pressure caused by the retropharyngeal mass. This would also explain improvement in symptoms associated with surgical excision of the lesion. The most common differential diagnosis for retropharyngeal masses is likely to include abscess and lymphoma but does not often include ganglioneuroma. In addition, infectious causes, branchial cleft cyst, and some other malignancies can be possible etiologies.¹⁰

Surgical excision is the treatment of choice with cervical ganglioneuromas producing symptoms. Due to

Table 1 Retropharyngeal Ganglioneuroma: Updated Review of Literature

Author	Age/Sex	Presentation	Size (cm)	Complications
Müller et al ³	35 y/F	Retro-auricular headache, dysphagia, tonsillitis-like pain	3.5 × 3 × 1.5	Horner's syndrome
Preidler et al ⁴	57 y/M	Dysphagia and dyspnea	19 × 9 × 4.5	—
Papso and Bencat ⁶	16 m/M	—	—	—
Gary et al*	42 y/F	Neck pain and stiffness, right arm weakness and numbness, dysphagia, and change in voice	5.5 × 3.2 × 1	Horner's syndrome

*Current study.

this tumor's origin from the sympathetic chain, surgical removal results in a postoperative Horner's syndrome. Our patient developed a Horner's syndrome secondary to surgical excision of the retropharyngeal ganglioneuroma. Müller et al also reported a postoperative Horner's syndrome after excision of a retropharyngeal ganglioneuroma.³ Although the symptoms related to Horner's syndrome after surgical excision can be managed conservatively, it is important to counsel patients regarding this postoperative sequela.

CONCLUSION

Retropharyngeal ganglioneuromas can occur in a wide age group of patients and should be included in the differential diagnosis of dysphagia as well as neck pain. Neck pain and stiffness are atypical symptoms that need to be worked up to rule out a communication with the spinal column prior to surgical intervention. Surgical excision via a cervical approach offers definitive therapy but may be associated with an iatrogenic Horner's syndrome for which the patients should be counseled prior to operative intervention.

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