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

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Abstract

Objective: 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.

Study Design: Case report and review of literature.

Methods: A 42-year-old woman presented with incapacitating neck pain and neck stiffness as well as dysphagia. Neurological workup was normal. Imaging revealed a hyper-dense, 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 post-operative Horner’s syndrome. In follow-up, dysphagia and neck symptoms improved.

Conclusion: 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 recognized to rule out communication with the spinal column prior to surgical removal. Patients must be counseled that atypical symptoms may not completely resolve with surgical treatment.

Case Report

A 42-year-old Caucasian female presented to our service with incapsicating neck stiffness and pain. The patient also reported headache, dysphagia, tonsillitis-like pain, neck pain and stiffness. 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 x 3.2 x 1 cm, well-circumscribed mass surrounded by a tenuous pseudocapsule. Microscopically, ganglion cells were identified scattered on a neurofibrous stroma. The final pathology was reported as ganglioneuroma, maturing subtype. In follow-up, the patient did well with improvement in neck symptoms and dysphagia. The patient also developed an asymptomatic post-operative Horner’s syndrome.

Discussion and Conclusion

Discussion: Ganglioneuromas are most frequently diagnosed between the ages of 10 and 29 years and are most commonly located in the posterior mediastinum followed by the retroperitoneum. Among the documented cases of cervical ganglioneuromas, retropharyngeal ganglioneuromas have only been reported in three patients in the current literature. These patients ranged from 16 months to 57 years. Our patient presented with the unusual symptoms of neck stiffness and pain. The differential diagnosis of neck pain includes cervical arthritis, discogenic disorders, trauma, infection, as well as spinal cord tumors. Although neurogenic tumors of the neck are relatively infrequent, they should be included in the differential diagnosis of a neck pain as is evident from our case. Commonly the differential diagnosis for retropharyngeal masses will include abscess, branchial cleft cyst, lymphoma and other malignancies but does not often include ganglioneuroma. Surgical excision is the treatment of choice with cervical ganglioneuromas producing symptoms. Due to this tumor’s origin from the sympathetic chain, surgical removal results in a post-operative Horner’s syndrome as occurred in our patient. Although the symptoms related to Horner’s syndrome after surgical excision can be managed conservatively, it is important to counsel patients regarding this post-operative sequela.

Conclusion: Retropharyngeal ganglioneuromas can occur in a wide age range of patients and should be included in the differential diagnosis of both dysphagia and neck pain. Neck pain and stiffness are atypical symptoms that need to be worked up to rule out 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 patient should be counseled prior to operative intervention.

References