Journal of Medical Imaging & Case Reports

https://doi.org/10.17756/micr.2019-032

Case Report

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Retroperitoneal Ganglioneuroma: An Unusual Cause for Hoarseness

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Received: July 29, 2019
Accepted: October 14, 2019
Published: October 15, 2019

Citation: Hmidi M, Sinaa M, Bellasri S. 2019. Retroperitoneal Ganglioneuroma: An Unusual Cause for Hoarseness. *J Med Imaging Case Rep* 3(2): 36-37.

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Abstract

Retroperitoneal ganglioneuroma (RGN) is a very rare entity, especially in children and adolescents. Certain clinical, histopathologic, radiologic, and genetic parameters can help distinguish ganglioneuroma from more aggressive tumors. We report the case of a 22-year-old man who was presented to Department of Otorhinolaryngology-Head and Neck Surgery, Military Hospital with a retroperitoneal ganglioneuroma, that presented as symptomatic left cervical mass and vocal cord paralysis.

Keywords

Ganglioneuroma, Retroperitoneum, Hypertrophic neuropathy

Case

Ganglioneuroma (GN) is a rare benign, slow-growing tumor arising from the immature cells of the sympathetic nervous system [1]. Most ganglioneuromas are poorly symptomatic or completely asymptomatic and discovered on routine screening [2]. The reported case is a classic example of incidentally found tumor, where the imaging did not allow a definitive diagnosis. Preoperative diagnosis is usually based on histopathological findings after complete surgical excision [3, 4].

A 22-year-old male was presented to the otorhinolaryngology department with a 6-month history of progressive hoarseness. The past medical history and family history were unremarkable. Physical examination revealed a palpable left cervical mass. Fiber optic laryngoscopy showed a paralytic left vocal cord. The mucosa was intact without tumoral lesions.

Neck ultrasound identified a level-II cervical lymph node (measuring 2 cm x 1.5 cm) with a hypoechoic, regular and diffuse enlargement of vagus and hyoglossus the nerves (Figure 1).

An exploratory cervicotomy-adenectomy was realized. At exploration, we noted the presence of a large hypertrophy of vagus, hypoglossus and greater ear nerves (Figure 2). The decision of greater ear nerve biopsy was made. Pathologic examination of both lymph node and nerve biopsy revealed no abnormalities.

Then, computed tomography (CT) (Panel A) and magnetic resonance imaging (MRI) (Panel B) of neck, chest and abdomen were performed, which determined a retroperitoneal mass (Figure 3). The tumor had infiltrated the celiac plexus and sympathetic nodes (Figure 4).

The patient underwent CT-guided core needle biopsy which revealed ganglioneuroma. There were mature ganglion cells in a schwannian stroma with

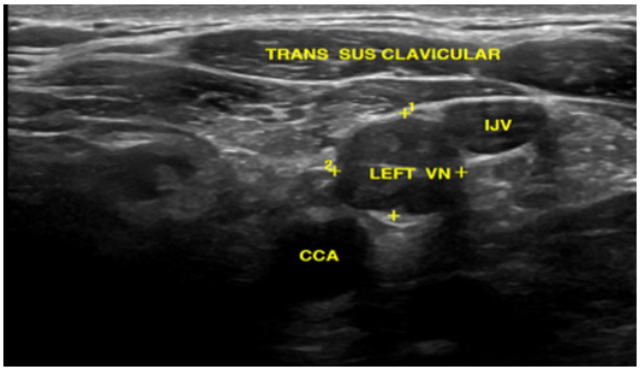


Figure 1: Neck ultrasound identified a hypoechoic, regular and diffuse enlargement of vagus nerve.

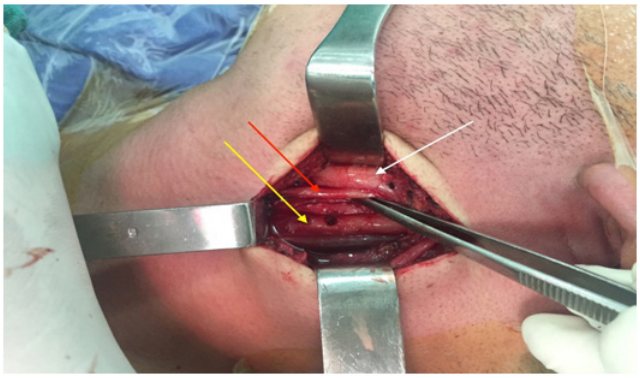


Figure 2: Exploratory cervicotomy-adenectomy noted the presence of a large hypertrophy of vagus (yellow arrow), hypoglossal (white arrow), descending branch of the hypoglossal nerve (red arrow), and greater ear nerves.

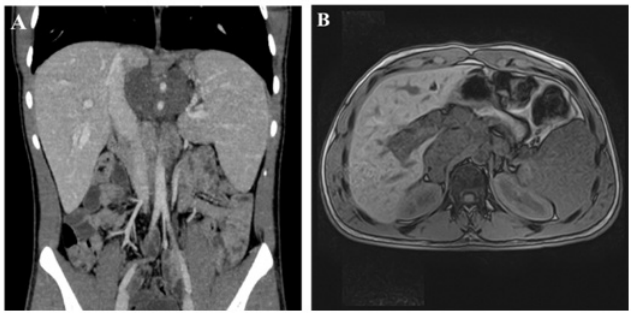


Figure 3: Computed tomography (Panel A) and magnetic resonance imaging (Panel B) of chest and abdomen illustrating a retroperitoneal ganglioneuroma.

no other histological signs of malignancy. Complete surgical resection of the tumor was performed as shown in video 1. Histopathological analysis was consistent with the diagnosis of ganglioneuroma.

In our particular case and considering imaging and exploratory cervicotomy findings, the lesion (RGN) was extending inferiorly to the celiac plexus and superiorly into the sympathetic cervical ganglia with infiltration of the vagus nerve, thus explaining laryngeal paralysis and consequently dysphonia.

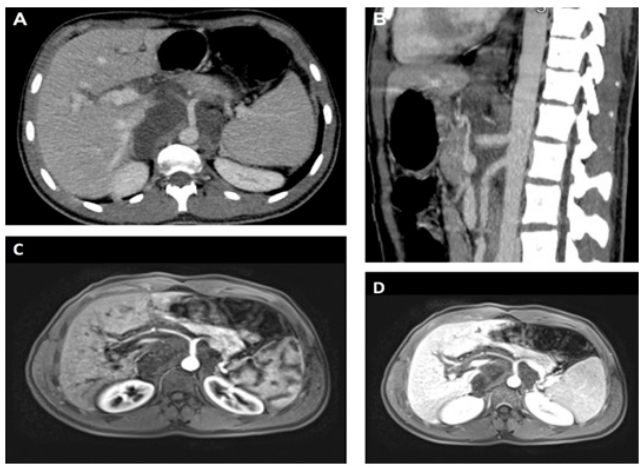


Figure 4: Computed tomography (CT) (Panel A, B) and Magnetic resonance images (MRI) (Panel C, D) show infiltration of the celiac plexus and sympathetic nodes by the retroperitoneal mass.



Video 1: Performing surgical resection for removing the tumor.

Conflict of Interest

The authors declared no conflict of interest.

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