Case Report

Castleman Disease Mimicking Neurogenic Tumor Radiologically: A Case Report

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[3,4,6] *Critical Review*
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Abstract

Introduction: Castleman disease which is also called giant lymph node hyperplasia is an uncommon benign B cell lymphoproliferative disease.[1] Paragangliomas on the other hand are rare tumors that develop from neural crest cells. They can arise from sympathetic or from parasympathetic ganglions.[3] There are only a few case reports of enlarged lymph nodes affecting the carotid artery and mimicking neurogenic tumors radiologically. We have one such report to present

Case presentation: We present a case of a young male with painless, slow-growing mass in the left upper part of the neck mimicking paraganglioma on CT scan and MRA, because of homogenous intense enhancement on computer tomography and rich blood supply from large-sized vessels respectively. The patient also had moderate bradycardia preoperatively, which is usually seen in vagal tumors. A postoperatively histopathology sample was sent which was identified as a case of Castleman disease. This case has been described in detail in our report.

Conclusion: Mass that was initially misunderstood as a case of neurogenic tumor and prepared accordingly turned out to be a case of Castleman lymphadenopathy. Thus the diagnosis of such a unique presentation was made Due to Castleman disease rarity and mostly asymptomatic presentation, it was difficult to diagnose. Its similarity to lymphoma and paraganglioma radiologically further added to its difficulty in diagnosis. Thus it was important to consider Castleman disease as a differential diagnosis of such mass.

Keywords: Paraganglioma, Castleman disease, neck mass, carotid body tumor, Castleman lymphadenopathy, hyaline type, Plasma type.

Introduction

Castleman disease, also called giant lymph node hyperplasia, is an uncommon benign B cell lymphoproliferative disease.[1] It usually occurs in young patients from 15-35 years with no gender predominance.[12] There are two subtypes of this disease, 1. Unicentric Castleman disease (localized with minimal symptoms) 2. Multicentric Castleman disease (systemic spread with anemia and splenomegaly). Based on histology it can be divided into many subtypes. a. hyaline vascular b. plasma cell c. HHV-8 associated Castleman disease d. multicentric Castleman disease not otherwise specified.[2]

Paragangliomas on the other hand are rare tumors that develop from neural crest cells. They can arise from sympathetic or from parasympathetic ganglions.[3] Parasympathetic ganglia-derived tumors are almost always found in the head and neck and can be of 5 types according to the site of origin. Jugulare, tympanicum, carotid body, vagale or laryngeal. These are usually benign and nonfunctional.[4]

Castleman disease has an unknown etiology, but the most widely accepted theory is that Castleman disease is a chronic low-grade inflammatory process. *I*nterleukin 6 plays important role in unicentric Castleman disease and both interleukin-6 and HHV-8 are considered to be involved in multicentric Castleman disease.[6]

In our patient, we were suspecting a nonfunctional paraganglioma most likely glomus jugulare based on a CT scan and surgical treatment was planned accordingly. Pre-operatively Computed tomography imaging confirmed the presence of a solid, homogenous, hypervascular, and well-delineated mass with homogenous intense enhancement after contrast. It was when the post-operative histopathological examination was done, we came to know that it was Castleman lymphadenopathy with hyaline type.

Materials and Methods

We report a case of a 21-year-old male patient presented in the outpatient department of Holy Family Hospital, Rawalpindi complaining of an indolent swelling on the left upper neck for the past 2 and a half years with little to no pain over the involved area. There were no other associated complaints. No history of fever, cough, dysphagia, dysphonia, or dyspnea. The patient had no complaints of headache, palpitations, or sweating. The patient didn't have to take any medications for the swelling.

Examination revealed 5*4 cm swelling below the left submandibular area, non-tender and firm inconsistency. The swelling was non-adherent to overlying skin and was mobile in the horizontal plane but fixed in the vertical plane. Mass was non pulsatile and bruit was absent.

His blood pressure, respiratory rate, and temperature were normal but his pulse showed bradycardia (Ranging from 43-55 rpm).

Systemic examination was unremarkable and he didn't have any such swelling on the rest of the body.

Ultrasound was performed which showed echo mixed swelling. FNAC was done later on and it revealed atypical cells. CT scan neck showed a heterogenous mass with post-contrast enhancement surrounded by multiple tortuous vessels arising from the thyrocervical trunk. MRA is suggestive of a nerve sheath tumor with blood supply from the external carotid artery. Urine and blood cathecolamines were done and came out normal

A Presumptive diagnosis of nonfunctional paranganglioma, most probably glomus vagalae, was made.

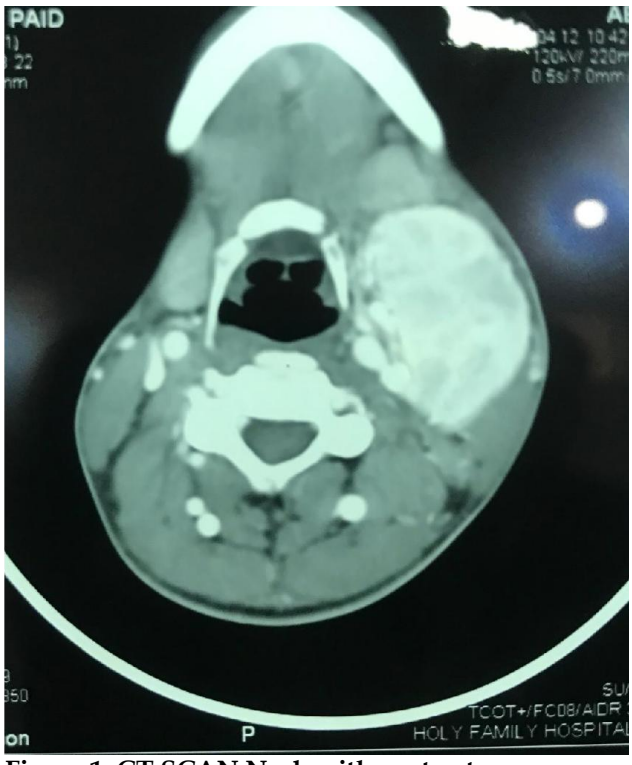

Figure 1: CT SCAN Neck with contrast
Well-defined 5.5 * 3.7*6.2cm oval avidly enhancing mass in left submandibular area with

lymphadenopathy. Dilated torturous vessels surrounding the lesion

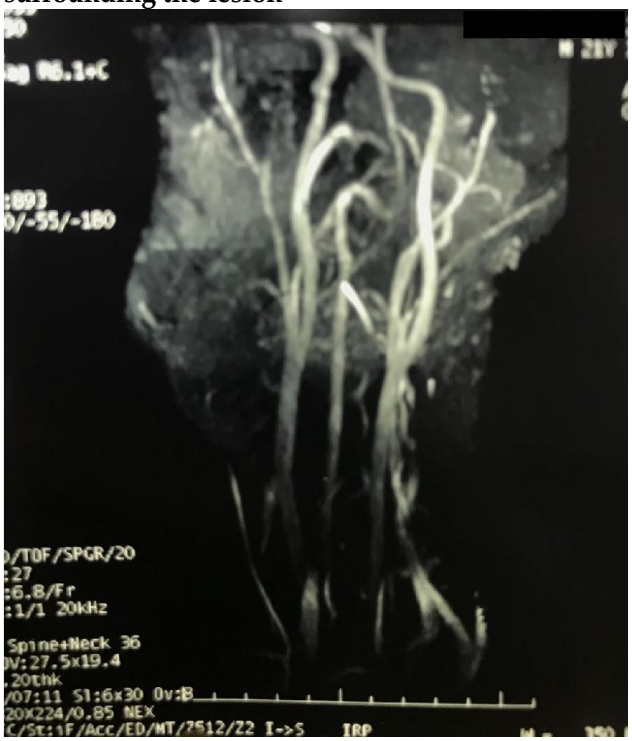

Figure 2: MRA

Baseline investigations were done before planning a surgery including complete blood count, prothrombin time, partial thromboplastin time, and Hepatitis B and C serology. All the investigations came out within normal range with Hepatitis B and C non-reactive.

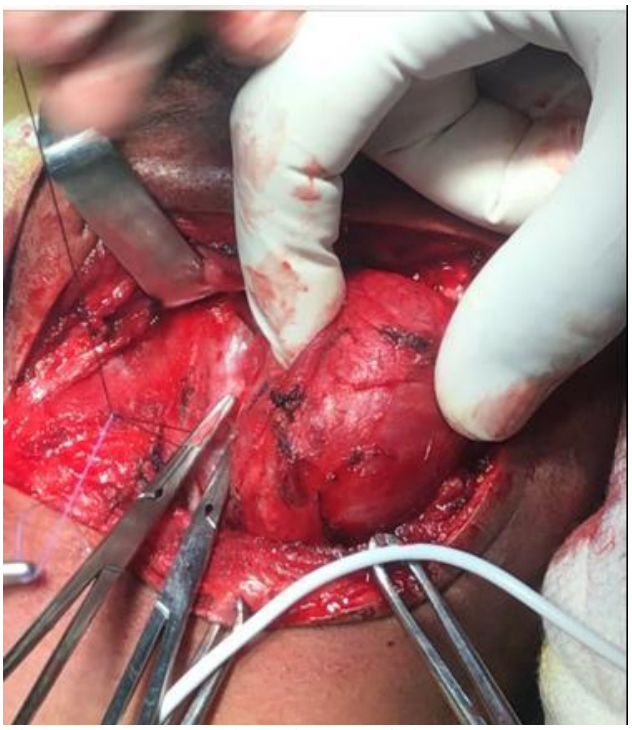

Figure 3: Peroperative picture of the mass

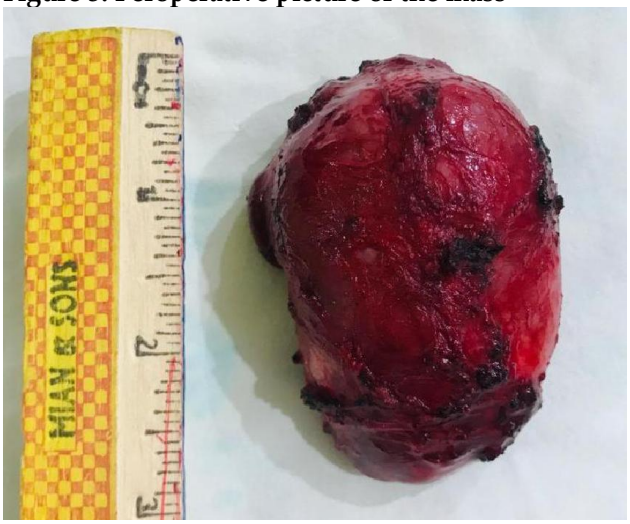

Figure 4: Mass delivered with dimensions 7.5 *4.5 *3.5cm

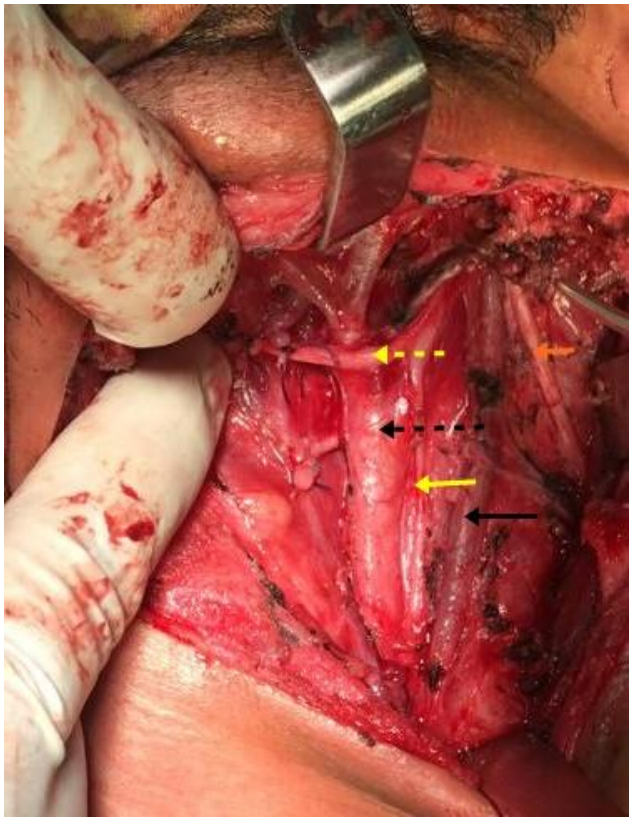

Figure 5 Black Arrow: IJV, Black dotted: carotid artery with Bifurcation, Yellow Arrow: vagus nerve, yellow dotted: hypoglossal nerve, Orange: spinal accessory nerve.

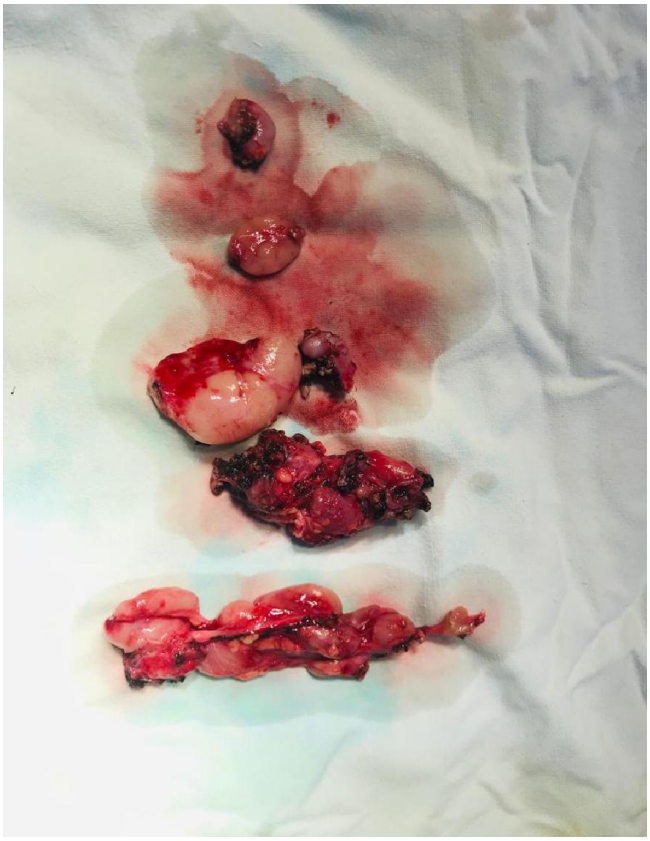

Figure 6: Lymph nodes removed from left level 2 and 3

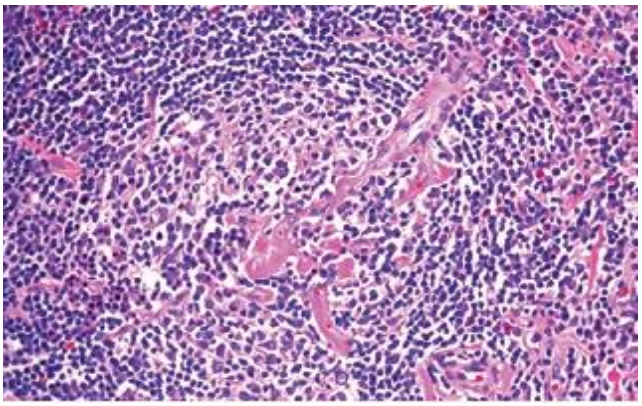

Figure 7: Hematoxylin and eosin (H and E) staining typical of Castleman disease, hyaline type.

Discussion

The classic CT appearance of Castleman disease with hyaline vascular type is that of localized nodal masses that demonstrate homogeneous intense enhancement after contrast administration.[5] CT scan findings in paraganglioma also show homogenous intense enhancement after contrast because of rich blood

supply. Due to castleman disease rarity and mostly asymptomatic presentation, it is difficult to diagnose. Its similarity to lymphoma and paraganglioma radiologically adds to the difficulty in its diagnosis.

Hyaline type is usually asymptomatic and unicentric. Histopathologically in hyaline vascular Castleman disease, lymph node follicles have widened mantle zones showing onion skin pattern which consists of concentric rings of lymphocytes. Lymphocytes are surrounded by small atrophic germinal centers.[7]

On the other hand, plasma cell type is associated with many signs and symptoms. Such patients can present with fever, malaise, sweating, weight loss, anemia, thrombocytosis, or hypergammaglobulinemia. They are usually associated with the multicentric disease. Histologically they show hyperplasia instead of regression and have vascular interfollicular regions.[8] The plasma cell type is most of the time inoperable and treatment includes radiotherapy, chemotherapy, or steroids.[11] Recurrence rate in hyaline type is low.

Our patient had unicentric disease with hyaline type. The prognosis for the unicentric hyaline vascular type after surgical removal is usually curative.[9] Radiation is another good option in this case.[10]

To conclude, this Castleman disease should always be among the differential diagnoses in a progressively growing mass in the neck. Thus patients should be examined and evaluated accordingly.

Conclusion

Mass that was initially misunderstood as a case of neurogenic tumor and prepared accordingly turned out to be a case of Castleman lymphadenopathy. Thus the diagnosis of such a unique presentation was made Due to Castleman disease rarity and mostly asymptomatic presentation, it was difficult to diagnose. Its similarity to lymphoma and paraganglioma radiologically further added to its difficulty in diagnosis. Thus it was important to consider Castleman disease as a differential diagnosis of such mass.

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