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CASE REPORT

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Garcin syndrome in a case of acquired immunodeficiency syndrome

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Abstract

In this study, we report a parapharyngeal diffuse large B-cell lymphoma in a human immunodeficiency virus (HIV) infected patient which had caused the patient to suffer from Garcin syndrome.

KEYWORDS

acquired, immunodeficiency syndrome, Garcin syndrome, human immunodeficiency virus, immunodeficiency syndrome, parapharyngeal space tumors

1 | INTRODUCTION

Parapharyngeal space tumors account for only 0.5% of head and neck tumors.[1] However, they deserve to receive desire attention since with the tumor growth, important anatomical structures such as the carotid artery, jugular vein, sympathetic chain, and cranial nerves can be influenced.[2]

Patients with parapharyngeal space tumors (PPS) usually represent a neck or oropharyngeal mass without presence of detectable symptoms on physical examination. The reason for the absence of symptoms is because that only the inferior and medial boundaries of the PPS are distensible. PPS lesions enlargement can lead to cranial neuropathies. With compression of central nervous system (CNS) IX, X, XI, or XII, symptoms of hoarseness, dysarthria, and dysphagia appear.[3,4]

Garcin syndrome is characterized by a progressive ipsilateral involvement of cranial nerves, culminating in paralysis of at least seven of them, with no sensory or motor

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long-tract disturbance, no intracranial hypertension and with osteoclastic involvement in the skull base on radiographic computed tomography.[5] The underlying cause is usually a sarcoma or carcinoma of the skull base.[6,7] Horner syndrome results from an interruption of the sympathetic nerve supply to the eye and is characterized by the classic triad of miosis (i.e., constricted pupil), partial ptosis, and loss of hemifacial sweating (i.e., anhidrosis).[8] In benign lesions, pain is unusual but in cases with compression or hemorrhage into the lesion, it can be observed. Totally, pain and neurologic dysfunction are more often indicative of malignancy with infiltration of the skull base.[9]

In this study, we report a parapharyngeal diffuse large B-cell lymphoma in a human immunodeficiency virus (HIV) infected patient which had caused the patient to suffer from Garcin syndrome.

2 | CASE PRESENTATION

A 48-year-old man with acquired immunodeficiency syndrome (AIDS) was referred to our center with complaints of headache from 5 days ago, ear pain, dysphagia, diplopia, and vertigo. The patient also reported a 5 kg weight loss over the past 2 months. The diagnosis of AIDS was made 8 months ago, of which cluster of differentiation 4 (CD4) counted 62×10^6/l and he had poor compliance regrading antiretroviral treatment (ART) and prophylaxis including sulfamethoxazole-trimethoprim. The recent CD4 count for the patient was 162×10^6/l. He was methadone dependent (80 ml daily) and a cigarette smoker from several years ago.

On admission, the patient exhibited normal vital signs. Head and neck examination revealed miotic pupils, diplopia, limited abduction of the left eye, facial numbness, hearing loss in the left ear, absence of the gag reflex, weakness of the left trapezius muscle, voice changing, and right deviation of a protruded tongue with local palsy. Bilateral plantar reflex was down, and other neurologic examinations were unremarkable. In the abdominal examination, splenomegaly with no sign of hepatomegaly was noted.

The patient was examined using indirect laryngoscopy (IDL). The larynx was full of secretion, and the patient was unable to swallow. There was paralysis of left cord of larynx with no mass effect.

Brain and neck CT scan with contrast revealed a hypodense mass-like lesion with mild enhancement in the left parapharyngeal space which causes carotid artery deviation to anterior and left nasopharyngeal bulging. (Figure 1) Left jugular vein obstruction due to mass compressive effect and also an erosion in the occipital condyle and lateral atlas mass (C1) in this side were seen.

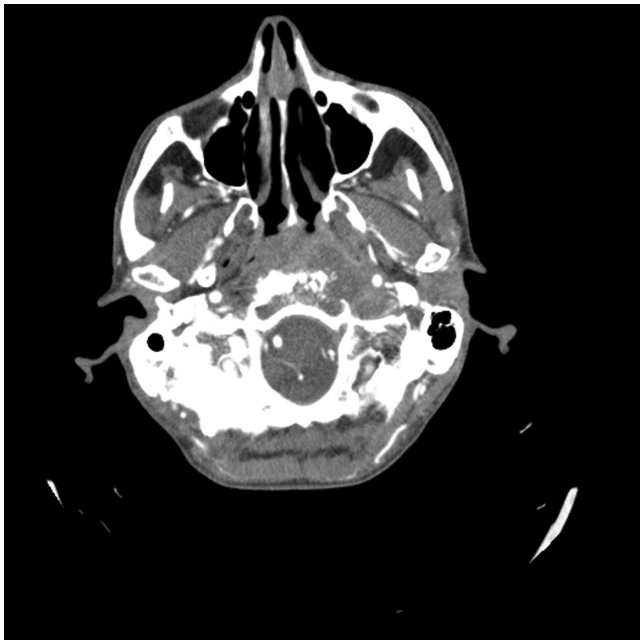

FIGURE 1 Brain and neck CT scan with contrast revealed a hypodense mass-like lesion with mild enhancement in the left parapharyngeal

In cervical CT scan, findings were included a parapharyngeal mass with no specific limitations extending from skull base to oropharynx that makes obliteration in parapharyngeal fat and deviation of ICA to the lateral side. (Figure 2) Basiocciput erosion and anterolateral of C1, anterior wall of transverse foramen of C1 was seen. Compressive effect on the left jugular vein and complete obstruction was also observed. Asymmetrical uvula and vallecula showed cranial nerve palsy due to mass compression effect. Lymphadenopathy in the left side level 2 with short axis diameter = 12 mm was observed.

Abdominal CT scan showed a large spleen with 145 mm diameter and several hypodense masses with targeted view in the spleen with mean size 30*20 mm. (Figure 3).

A core needle biopsy was performed from cervical lymph node level 2, and the pathological report was high grade diffuse large cell lymphoma activated B-cell (ABC) type. Immunohistochemistry study revealed positive results for LCA, CD20, CD79a, MUM1, Bcl2, Bcl6, and negative results for CK, CD3, CD10, and Myc. Proliferation index was high (Ki67 = 70%).

Because of refractory and progressive decrease in platelet count ($8\times10^3/\mu L$) due to lymphomatous infiltration of spleen, the patient underwent total splenectomy and a few days later thrombocytopenia was resolved from ($307\times10^3/\mu L$).

Eventually, the patient deceased due to rapidly progressive respiratory distress probably opportunistic infection including Pneumocystis jiroveci despite of

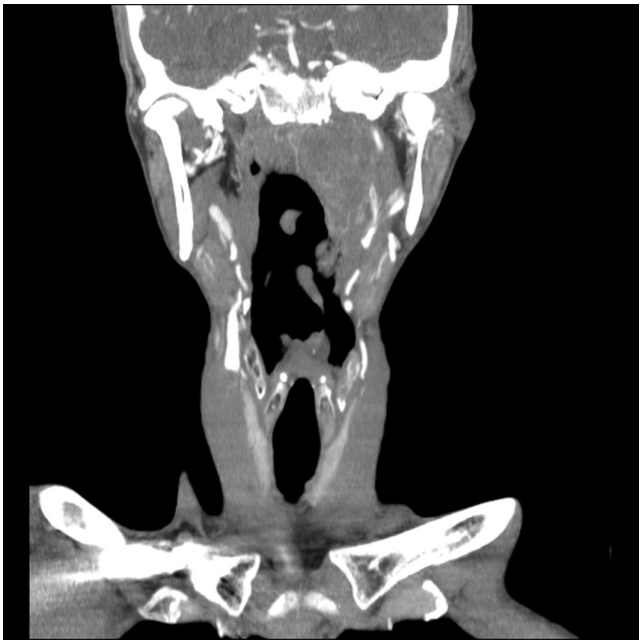

FIGURE 2 Cervical CT scan revealed a parapharyngeal mass with extending from skull base to oropharynx that makes obliteration in parapharyngeal fat and deviation of ICA to the lateral side

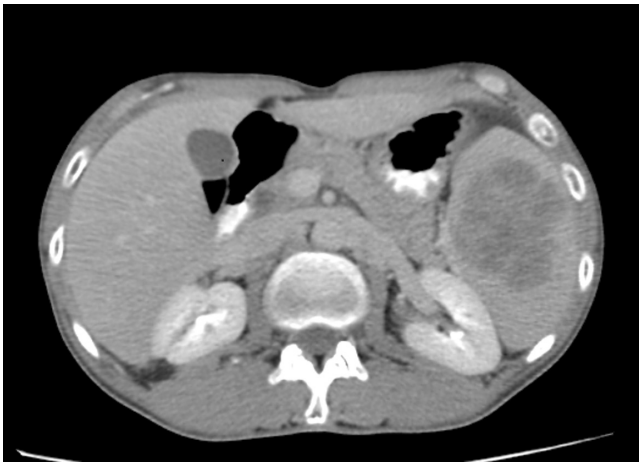

FIGURE 3 Abdominal CT scan showed a large spleen with 145 mm diameter and several hypodense masses

urgent therapeutic intervention and sulfamethoxazole-trimethoprim prophylaxis during recent admission. (Figure 4).

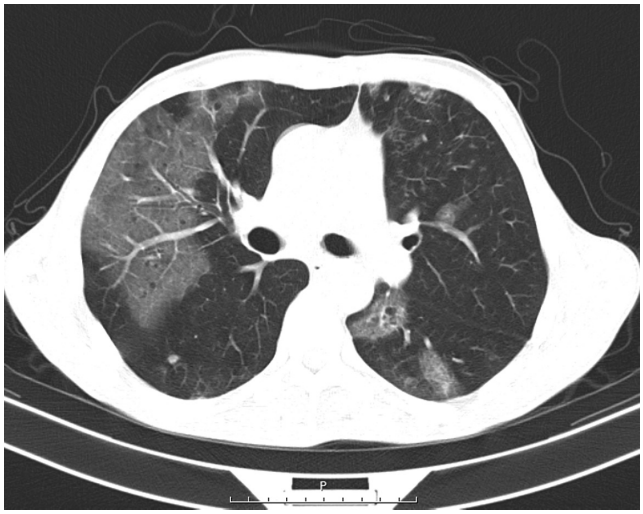

FIGURE 4 Lung CT scan showed bilateral multifocal ground glass opacities

3 | DISCUSSION AND CONCLUSIONS

Meeting the following four diagnostic criteria is used as definition of Garcin syndrome: 1. unilateral palsies of the cranial nerves, 2. neither sensory nor motor long-tract disturbance, 3. no intracranial hypertension, and 4. an osteoblastic lesion in the skull base.[10] Most cases with Garcin syndrome are susceptible to primary or secondary bone tumors and a few exhibits inflammatory disorders. In the majority of cases, not all twelve brain nerves are affected.[11]

Based on the reports, there have been various causes for Garcin Syndrome including tonsillar carcinoma, mucinous adenocarcinoma of the nasopharynx,[12] nasopharyngeal carcinoma,[13] adenoid cystic carcinoma,[14] parotid gland adenoid cystic carcinoma,[15] and rhinocerebral mucormycosis (in an aged with diabetic man),[16,17] skull base metastasis of ACDK-related RCC (which is a known unique carcinoma that occurs in patients with long-term HD).[18] The slow localized growth of these tumors is determination of the Garcin syndrome, rather than by their histology and primary localization.[12] There are also Collet-Sicard syndrome, cavernous sinus syndrome, Tolosa–Hunt syndrome which represent similar clinical findings with Garcin syndrome. These syndromes occur due to paralysis of a certain cranial nerve (e.g., Collet-Sicard is IX to XII).[7] For timely diagnosis of Garcin syndrome, findings on computed and magnetic resonance tomography play crucial role.[12] According to some reports, performing biopsy in cases with Garcin syndrome also provides beneficial information.[19]

The symptomatic duration of Garcin syndrome among patients varies from 2 months to 7 years.[20] The prognosis is as a rule unfavorable. Garcin syndrome usually is a late presentation of disorders. The skull basillar meninges and bones are the most common places that involved. Direct extension and invasion by nasopharyngeal carcinoma or primary tumor of the skull base are the main cause of basal meningitis. The clinical presentation is usually as

progressive cranial nerve palsies. The pathological process of direct extension of tumors may affect the cranial nerves at their exit sites from the skull or in their extracranial pathways.[21]

Tumors of the parapharyngeal space (PPS), a potential space lateral to the upper pharynx, occur less than 1% of all head and neck malignancies.[3,8,9] Among lymphoproliferative neoplasms, diffuse large B-cell lymphoma declare as the most common type in the United States.[22]

Diagnostic work-up for HIV infection is mandatory for patients with risk factors, especially those with large cell or small noncleaved-cell histopathologies.[23] Fluorodeoxyglucose (FDG) positron emission tomography with computed tomography (PET/CT) is the preferred imaging modality for staging FDG-avid nodal lymphomas, while CT alone is preferred for FDG-non-avid and variably FDG-avid histopathology.[8]

This patient had AIDS as a risk factor and his presentation was misleading abdominal, parapharyngeal, nervous signs, and symptoms. He needed an imaging study to find the cause of presentations and also surgery and histopathological study.

Lymphoma is one of the differential diagnoses for a patient presenting with PPS mass and Garcin syndrome. In our patient, cervical mass and several masses in the lung and spleen were seen. Lymphoma is one of the most important differential diagnosis that we should think about it in the same patients. Lymphoma can involve various organs such as lung, spleen, and parapharyngeal space. It is important that to take medical decisions fast and also medical invasive diagnostic and therapeutic workup perform as soon as possible.

AUTHOR CONTRIBUTIONS

All authors contributed to conception and design of study; MMR and IAD contributed to the acquisition of data; MMR, IAD, and OM contributed to the analysis of data; all authors contributed to the drafting of the article and/or critical revision; and all authors contributed to the final approval of manuscript.

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CONFLICT OF INTEREST

We declare no competing interests.

DATA AVAILABILITY STATEMENT

Not applicable.

ETHICAL APPROVAL

Not applicable.

CONSENT

Written informed consent for publication of patient's clinical details and/or clinical images was obtained from the relative of the patient. A copy of the consent form is available for review by the Editor of this journal.

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