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Case Report

Multiple unilateral craniopathies (Guillain-Alajouanine-Garcin syndrome) resulting from a malignancy of the external auditory canal: a case report

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ABSTRACT

Garcin syndrome (or Gullain-Alajouanine-Garcin syndrome) was first described in the year 1926, as a paralytic unilateral cranial syndrome consisting of multiple cranial nerve palsies, without findings pointing towards increased intracranial pressure or long tract involvement. A 54-year-old female presented to us initially with ptosis and lateral rectus palsy of the right eye, later on, followed by progressive hearing loss on the right ear, right facial hemianesthesia, and progressive ipsilateral facial paralysis. A mass at the external auditory canal was seen during otoscopy, and a biopsy revealed squamous cell carcinoma. Contrast-enhanced neuroimaging also showed an enhancing mass lesion at the right skull base.

Keywords: Garcin syndrome, Multiple craniopathies, External auditory canal malignancy

INTRODUCTION

Gullain-Alajouanine-Garcin syndrome is a rare neurologic clinical disorder that was first reported by Garcin in 1926, which was described as multiple unilateral cranial nerve palsies occurring in the absence of long tract signs and evidence of increased intracranial pressure.[1] Garcin reported involvement of almost all of the cranial nerves, but it was Desai et al who described is as paralysis of at least 7 ipsilateral cranial nerves back in 2013. He described it as a rare progressive neurologic condition that is not only secondary to compressive or osteolytic skull base lesions or malignancies, but also due to pachymeningitis that could be inflammatory, infectious, or or malignant.[2] What makes this case reportable is that the patient presented with clinical, radiologic, and pathologic findings that are infectious, neoplastic, and inflammatory.

CASE REPORT

A 54-year-old female patient was initially admitted to our institution due to complaints of ear pain and ear discharge, frontal headache, diplopia, and dizziness, with neurologic findings of right eye ptosis, bidirectional horizontal gaze palsy, sensorineural hearing loss on the right ear with ipsilateral ear discharge. The patient had no history of previous illness except for pulmonary tuberculosis, which was treated for 6 months. During this admission, she was initially managed as a case of Tolosa-Hunt syndrome and chronic otitis media and was treated with a 4-month course of steroids. The patient was discharged improved and was advised follow up consult. 2 months after discharge, the patient was readmitted due to progressive right facial hemianesthesia and paralysis, with worsening ear discharge and hearing loss. By this time, the patient was already in poor general health condition, unable to walk and maintain balance.

Neurologic examination showed intact cortical functions, with an MMSE score of 28. A detailed examination of the cranial nerves revealed an intact sense of smell (CN I); visual acuity of 20/30 on the left and 20/75 on the right without correction, no visual field cuts, with no evidence of papilledema on fundoscopy (CNII); with negative direct and consensual pupillary light reflex on the right eye (CN II and III) and complete ophthalmoplegia on the right (CN III, IV, VI).

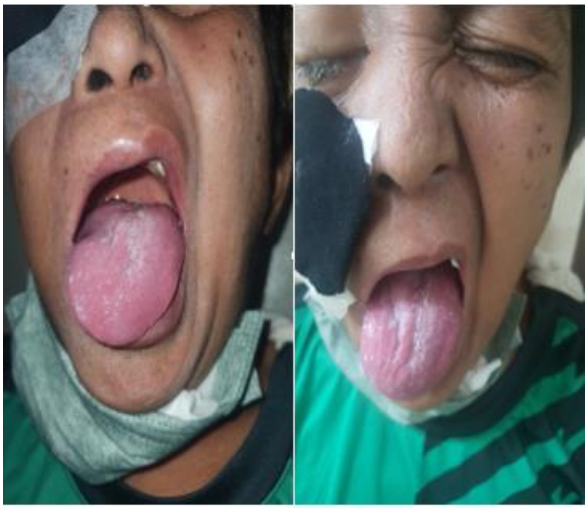

Figure 1: Complete right CN VII paralysis with tongue deviation and hemiatrophy.

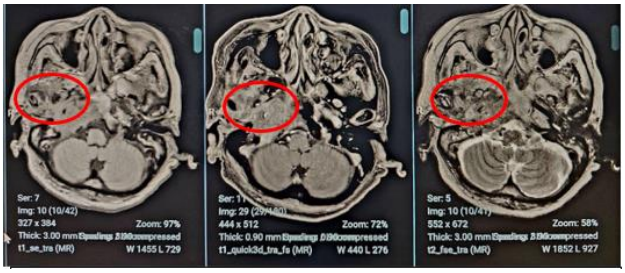

Figure 2: Contrast-enhanced MRI showing a lytic mass lesion on the right external auditory canal extending to the right middle cranial fossa.

There was complete hemianesthesia to pinprick, temperature, and light touch on the right side of the face, with gross and disfiguring facial weakness on the right as well (CN VII). The patient had a House-Brackmann score of VI. There was sensorineural hearing loss in the ipsilateral ear, with a noted severe inability to maintain balance (CN VIII). The patient had hoarseness of voice with weak gag reflex, and on the oropharyngeal exam, there was asymmetric palatal elevation on phonation (CN IX, X). Shoulder and cervical muscle strength was normal (CN XI). On tongue exam, there was significant hemilingual atrophy with deviation towards the right side (XII).

Motor examination did not show significant weakness during individual muscle group testing, while sensory testing on the extremities was also unremarkable. Cerebellar testing was also normal, while gait testing was not performed due to a profound inability to stand and maintain balance. The patient was normoreflexic on bilateral upper and lower extremities and was negative on extensor toe signs. The neck was supple and there were no signs of meningeal irritation elicited.

Neuroimaging revealed a heterogeneously enhancing lesion measuring about 6.0×6.7×3.9 cm (L×W×AP) involving the right external auditory canal and extending to the right side of the skull base which involves the bony temporal fossa and condylar process of the right mandible. Anteromedially, it further extends to the cavernous sinus and the jugular foramen. It also extends anteriorly to compress the lesser wing of the sphenoid and the optic canal and superiorly, it extends to the middle cranial fossa and abuts the temporal lobe. However, there were no abnormal signals noted within the brain parenchyma. There was also noted enhancement of the pachymeninges lining the cavernous sinus. There was no evidence of increased intracranial pressure with respect to neuroimaging. The mass was visualized via otoscopy wherein biopsy was also performed which revealed well-differentiated squamous cell carcinoma.

Contrast tomography of the chest showed sub-centimeter pretracheal lymph nodes, with pulmonary fibrosis on both upper lung apices. Contrast-enhanced CT of the Whole Abdomen only showed bile sludge, with incidental findings of degenerative changes in the lumbar spine.

CSF analysis was also done to rule out central nervous system infection. An opening pressure of 16 Cm H_2O was obtained. 36 ml of clear, colorless cerebrospinal fluid was subjected to pathologic analysis, wherein atypical cells that are suggestive of a malignant etiology were seen. CSF cultures were negative, while the CSF protein, glucose, and cell counts were all within normal limits. Culture studies of the right ear discharge were also done, which showed moderate growth of *Morganella morgagnii*.

The patient was referred to co-managing services (ENT, infectious diseases, medical and radiation oncology) for management of head malignancy and infection. During the course of admission, the patient developed nosocomial pneumonia and stress-related mucosal ulcer leading to gastrointestinal bleeding. Ultimately, the patient succumbed to severe sepsis and pulmonary embolism.

DISCUSSION

Gullain-Alajouanine-Garcin syndrome or Garcin's syndrome, was described by Garcin in 1926 as progressive hemifacial paralysis involving multiple ipsilateral cranial nerves.[3] The criteria that define the syndrome include the following: ipsilateral multiple cranial nerve palsies-at least seven cranial nerves must be

affected; absence of motor or sensory long-tract involvement; absence of evidence pointing towards increased intracranial pressure; and lastly, presence of osteoclastic skull base lesion.[2-4] The diagnostic challenge lies in the ability of the clinician to recognize such a syndrome and distinguish it from other benign conditions with the same clinical manifestations, as well as the patient's urgency to seek early consultation before the progression and spread of cranial nerve affectation. Syndromes or conditions that may present similarly include Collet-Sicard syndrome, cavernous sinus syndrome, Tolosa-Hunt syndrome, and Bell's palsy.[5] Physicians who have little clinical experience and exposure to neurologic diseases might easily misdiagnose Garcin syndrome as a benign condition like Bell's palsy, and this is the reason why appropriate diagnosis and treatment are delayed, leading to the progression of cranial nerve palsies.[6] It must be emphasized that prompt diagnosis and treatment of the etiology of Garcin syndrome is key to the patient's survival.

In the Philippines, there is no published case of Garcin syndrome caused by any pathologic entity. Fukai et al in 2018 reported 7 cases of Garcin syndrome secondary to metastatic and/or invasive skull base lesions that are pulmonary in origin, while Yadav in 2021 likewise described 7 cases of Mucormycosis presenting with Garcin syndrome.[5,12] No literature has reported a case of Garcin syndrome secondary to a primary otologic malignancy. Most reports of Garcin syndrome attribute the neurological manifestations to an expanding skull base tumor or coming from adjacent structures including the nasopharynx. Out of the reported cases, lung cancer is the third most likely to develop bone metastasis following breast cancer and prostate cancer.[7] Presently, there is no published literature ascribing skull base metastasis to breast and/or prostatic malignancies presenting with multiple craniopathies. While the majority of literature point towards metastatic skull base lesions, multiple case reports also described multiple craniopathies resulting from primary extracranial lesions, including multiple reports of giant cell sphenoid tumors, a case of a skull base diffuse B cell lymphoma, and an autopsied case of epipharyngeal rhabdomyosarcoma.[8-12]

Clinical diagnosis of Garcin's syndrome remains a challenge because it commonly presents neurologic manifestations that are commonly attributed to other neurologic diseases like stroke, Bell's palsy, Tolosa-Hunt syndrome, especially in its early stage. Therefore, awareness of such syndrome and its associated pathologic etiologies is important because it is essential to establishing the diagnosis and the appropriate treatment for each case. Imaging, such as computerized tomography (CT) or magnetic resonance imaging (MRI) is central to diagnosis and in fulfilling the criteria set by Garcin himself. MRI, however, provides better visualization of the CNS, perineural invasion and vascular obstruction and is the preferred imaging tool for such cases.[12] The presence of a tumor in imaging may subsequently warrant tissue sampling for histopathologic diagnosis, considering that most cases of Garcin syndrome are metastatic or invasive.

CONCLUSION

Patients suspected to have Garcin syndrome should be seen and managed in a multi-disciplinary approach involving neurology, pathology, neuroradiology, and oncology. Cases that are managed early, especially with prompt diagnosis of the underlying pathological etiology result in improved survival. However, the disability remains likely to be permanent and is not limited to physical disability but also includes psychosocial and emotional factors due to disfigurement caused by facial paralysis. Therefore, rehabilitation and psychiatric counseling also plays an important role in surviving patients.

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