Garcin syndrome caused by parotid gland adenoid cystic carcinoma
A case report

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Abstract

Rationale: Garcin syndrome is characterized by the gradual involvement, and ultimately, unilateral paralysis of at least 7 and sometimes all cranial nerves, without intracranial hypertension or any long tract signs.

Patient concerns: We report the case of a 59-year-old woman who presented with Garcin syndrome, which gradually progressed over a period of 2 years.

Diagnosis: A left parotid gland biopsy revealed parotid gland adenoid cystic carcinoma (PGACC) with perineural invasion of a peripheral nerve bundle and lymph node metastasis.

Interventions: The patient was treated 3 times with local-field palliative radiotherapy.

Outcomes: She died after several months.

Lessons: To the best of our knowledge, this is the first report of PGACC presenting as Garcin syndrome. PGACC is a rare tumor with a high propensity for perineural spread, and it should be considered as a possible cause of Garcin syndrome.

Abbreviations: ACC = adenoid cystic carcinoma, CT = computed tomography, MRI = magnetic resonance imaging, PGACC = parotid gland adenoid cystic carcinoma.

Keywords: adenoid cystic carcinoma, cranial nerves palsy, Garcin syndrome, parotid gland, PGACC

1. Introduction

Garcin syndrome is a rare disorder with progressive unilateral involvement of all or at least 7 cranial nerves.[1] The most common cause is carcinoma of the nasopharyngeal region with skull base invasion. Adenoid cystic carcinoma (ACC) is a slow-growing malignant epithelial neoplasm, characterized by wide local infiltration and a high incidence of perineural spread, local recurrence, and late distant metastasis.[2] ACC accounts for 7.5% to 10% of all salivary gland malignancies and approximately 1% of all malignant tumors of the head and neck region.[3] To our knowledge, only 2 cases of Garcin syndrome due to ACC have been previously reported.[4,5] Herein, we report a rare case of Garcin syndrome caused by parotid gland adenoid cystic carcinoma (PGACC).

2. Case report

A 59-year-old woman was admitted with left facial lancinating pain, facial palsy, dysarthria, and dysphagia, all of which had gradually worsened over a period of 2 years. Two years before admission, she had presented with severe left facial pain, in the areas supplied by the third branch of the ipsilateral trigeminal nerve, and was diagnosed with and treated for trigeminal neuralgia at the local hospital without positive cranial computed tomography (CT) findings. However, the treatment did not relieve her symptoms. One year after onset, the patient began to gradually develop left facial palsy with swelling, dysarthria, and dysphagia. Subsequently, 6 months before admission, the patient was treated at another institution using radiofrequency thermo-coagulation of the gasserian ganglion, again for suspected trigeminal neuralgia, but the treatment was effective for only 9 days. In order to conduct further investigations, she was referred and admitted to the First Affiliate Hospital of Wenzhou Medical University in September 2011.

The neurological examination on admission revealed left side trigeminal hypesthesia, facial palsy, hearing impairment, bulbar palsy, shoulder weakness, and tongue fasciculation and weakness, suggesting involvement of the fifth and the seventh to twelfth cranial nerves (V, VII, VIII, IX, X, XI, and XII), which met the criteria of Garcin syndrome. Her muscle strength, sensation,
and coordination were normal in all 4 extremities. Laboratory test results, including serum and cerebrospinal fluid analyses, were all within normal ranges.

Plain and gadolinium-enhanced magnetic resonance imaging (MRI) revealed an extensive abnormal signal in the left parapharyngeal space, root of the tongue, and parotid gland (Fig. 1). Pathological analysis of a left parotid tissue biopsy led to a diagnosis of PGACC with perineural invasion of a peripheral nerve bundle and lymph node metastasis (Fig. 2). Given the wide invasion and metastasis of the tumor, the patient was treated 3
case report was treated using local-field palliative radiotherapy because of widespread tumor invasion and lymph node metastasis.

In summary, to the best of our knowledge, this is the first reported case of PGACC presenting as Garcin syndrome. This case illustrates the need to consider ACC as a possible cause when examining patients with unilateral/multiple cranial nerve palsies resulting in the progressive slowing of facial movements.

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