Non-Hodgkins Lymphoma presenting as Garcin's Syndrome

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ABSTRACT

An 80 year old man presented with a twelve month history of progressive multiple unilateral cranial nerve palsies. A gadolinium enhanced MRI scan of his head and neck was performed. This showed an extra-axial enhancing lesion, which enveloped the hemi-mandible at the level of the left inferior alveolar nerve, and a dumbbell shaped lesion within the cerebellopontine angle. An incisional biopsy was performed to gain a tissue diagnosis, which demonstrated a Non-Hodgkin's Lymphoma. The combination of signs, symptoms and radiological findings enabled a diagnosis of Garcin's Syndrome secondary to Non-Hodgkin's Lymphoma.

INTRODUCTION

Garcin's syndrome was first defined in 1926 by Raymond Garcin, who defined the "syndrome paralytique unilateral global des nerfs craniens" (1). Garcin's syndrome is characterized by (1):

(i) the presence of unilateral palsies of the cranial nerves,

(ii) no sensory or motor long tract disturbance,

(iii) no intracranial hypertension and

(iv) an osteoclastic lesion in the skull base

Patients reported to have Garcin's syndrome do not always have unilateral palsies of all cranial nerves; thus, this syndrome was redefined as the presence of at least seven ipsilateral cranial nerve palsies. (2)

CASE REPORT

An 80 year old man presented to his General Medical Practitioner. He noticed a mass in his left buccal sulcus, and both his General Medical Practitioner and General Dental Practitioner prescribed a total of three courses antibiotics. As there was no resolution he was referred to his local Oral and Maxillofacial Unit for further investigation. By this time he was also complaining of left sided deafness and diplopia.

On extra-oral examination he had an obvious soft tissue swelling emanating from his parotid, and extending buccally towards the mental foramen. An orthopantomogram showed a widening of the inferior dental nerve canal and signs of bony resorption. Left acoustic (VIII) and facial (VII) cranial nerve palsies occurred initially, followed by trigeminal (V), abducens (VI) glossopharyngeal (XI), vagal (X), accessory (XI), and hypoglossal (XII) left sided cranial nerve palsies. Seven of the 12 cranial nerves were involved in this patient. Nevertheless, his muscle tone, power, sensation, and coordination were normal in all four extremities.

Gadolinium-enhanced MRI of the brain showed an extra-axial enhancing lesion, which enveloped the hemi-mandible at the level of the left inferior alveolar nerve. The lesion was 3.8 cm at its maximal thickness, and eroded the cortex and the mandible. An examination under general anaesthetic was performed. This revealed a left sided parotid mass extending posteriorly to the mandibular angle , anteriorly beyond the mental foramen and postero-superiorly to the skull base. An incisional biopsy was taken which was histologically reported as Non-Hodgkin's Lymphoma. Following discussion at the Head and Neck MDT, the patient was referred to the Lymphoma team. A decision was made to treat the patient with primary Chemo-radiotherapy.

DISCUSSION

A rare case of Garcin's syndrome presented with progressive multiple unilateral cranial nerve palsies due to Non-Hodgkin's Lymphoma. To our knowledge, only one case of Garcin's syndrome due to Non-Hodgkin's lymphoma has been previously reported. (3) It is more frequently caused by malignant disease; however it can be caused by benign disease. Garcin's syndrome is reported widely as a result of metastatic spread from a primary tumour. However primary oropharyngeal tumours have rarely caused Garcin's syndrome. There are only three cases of Garcin's syndrome being caused by a primary lesion within the head and neck reported within the last forty years. These were caused by a chemodectoma (4) and a meningioma (5).

Benign causes of Garcin's syndrome include pachymeningitis secondary to otitis media (6) and a large internal carotid artery aneurysm (7).Our case is interesting as it was caused both by a primary lesion, and it is the only Non-Hodgkin's Lymphoma case to have been found pre-mortem and as a primary lesion intra orally. On admission to our hospital, the patient had palsies of seven left cranial nerves, which satisfied the clinical picture of Garcin's syndrome. The initial diagnosis in this case was Bell's palsy which is frequently encountered in clinical practice. Multiple cranial nerve palsies are rare; however the clinician must be aware of such cases to make sure early accurate diagnosis is achieved.

In conclusion, multiple cranial palsies are rare and often present initially as a single cranial nerve palsy. In our case of Garcin's Syndrome the patient presented with a facial nerve palsy and an accurate diagnosis of non-Hodgkins Lymphoma at this stage may have prevented the development of this syndrome. The diagnosis can only be a made with a high clinical suspicion and appropriate diagnostic imaging.

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