

SHORT REPORT

Renal cell carcinoma presenting as Garcin's syndrome

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Abstract

A 50 year old man presented with progressive unilateral cranial nerve palsies (Garcin's syndrome) for one year. Skull radiography and computed tomography (CT) showed intracranial extension of a soft tissue tumour from the right base of the skull. Necropsy revealed a papillary cell carcinoma of the right kidney and metastases in the base of the skull.

A 50 year old man presented with progressive unilateral cranial nerve involvement caused by a metastasis from a renal cell carcinoma. In 1926, Raymond Garcin described this "syndrom paralytique unilateral global des nerfs craniens".¹ The syndrome is now known as Garcin's syndrome, hemi basal syndrome, Guillain-Garcin syndrome or Bertolotti-Garcin syndrome and is characterised by: progressive involvement of the cranial nerves culminating in total unilateral paralysis of all the cranial nerves; absence of motor or sensory signs in the limbs, absence of raised intracranial pressure, and abnormal skull base radiographs.² Since then reported causes of this syndrome have been: nasopharyngeal, paranasal sinuses, or parotid tumours;^{3,4} sarcoma of the base of the skull or meninges;^{5,6} middle ear carcinoma;^{7,8} rhabdomyosarcoma;⁹ cylindroma;¹⁰ craniopharyngioma;¹¹ metastatic spread from cancer of the breast, lung, liver or uterus;¹² solitary chondroma;¹³ giant aneurysm of the internal carotid artery;¹⁴ chemodectoma of the glomus jugulare;¹⁵ luetic purulent tuberculous, carcinomatous or leukaemic meningitis¹⁶ and others. We report a patient with an identical syndrome seen in association with renal cell carcinoma.

Case Report

A 50 year old right handed man was admitted to our neurology unit with a one year history of headaches, double vision, numbness of the right side of the face, progressive loss of weight and general ill health.

He was well until eighteen months before admission when he was investigated for right sided loin pain. He was advised to have surgery but declined and was subsequently lost to follow up.

One year before his admission, he began to

have generalised headaches that were dull, continuous and increasing in severity, but they were not associated with nausea or vomiting. Nine months before admission, he developed double vision, numbness on the right side of his face and progressive weight loss. Over a period of months he experienced recurrent epistaxis from the right nostril. Two months before admission, he had difficulty in chewing and in talking.

On examination the patient appeared emaciated with a weight of 45 kg. No organomegaly, lymphadenopathy or clubbing were found. Other systems were normal. Neurological examination revealed loss of smell in the right nostril, complete ophthalmoplegia of the right eye with ptosis. He was blind in the right eye with optic atrophy; the left disc was normal. The right pupil showed no reaction to light. There was hypoaesthesia involving the right side of the face with weakness and wasting of the right masseter muscle.

The right corneal reflex was lost. The right nasolabial fold was decreased with weakness of the right frontalis muscle. He was deaf in the right ear. The palate deviated to the left and gag reflex was absent on the right side. There was severe wasting of the sternocleidomastoid and trapezius muscles on the right. The right side of the tongue was also wasted and showed fasciculation. Muscle tone, power, sensation and coordination were normal in all extremities. Tendon reflexes were present and plantar responses flexor. Gait was normal.

Laboratory findings, including cerebrospinal fluid (CSF), protein and cytology were normal, but the patient had a haemoglobin level of 9.3 G/dl and an ESR of 118 per hour, and 4-5 red blood cells per high-power field in the urine. The chest radiograph was normal, but the skull radiograph showed an eroded sella turcica with multiple lytic lesions in the skull vault, whilst basal views showed destruction of the right petrous bone. Contrast CT scan showed a huge osteolytic soft tissue tumour with marked enhancement in the right side of the base of the skull destroying the tip of the petrous, sphenoid and ethmoid bones. The medial orbital wall was destroyed. There was an intracranial extension of the tumour to the right temporal lobe, cerebellar hemisphere, pons and mid-brain. No hydrocephalus was seen.

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All other investigations including abdominal ultrasound, intravenous urography, kidney scan with Tc-DMSA and bone scan performed with Tc-MDP suggested bony metastasis with a primary lesion in the upper pole of the right kidney. Multiple biopsies taken from the right nostril, post-nasal space and pharynx showed no malignant change.

The patient was advised to have surgery to remove the primary lesion to confirm the histological diagnosis and to determine further management, but he refused and discharged himself. Two weeks later he returned with a three day history of severe pain in the right eye followed by a watery discharge.

An ophthalmic examination revealed a rupture of the globe. Tarsorrhaphy was performed. The patient's general condition deteriorated still further, he became drowsy and disorientated and eventually lapsed into coma and died. Necropsy revealed papillary cell carcinoma of the right kidney and confirmed the presence of metastases in the base of the skull, the spine and ribs.

Discussion

Garcin's syndrome is usually a late presentation of a disease process that is probably but not invariably malignant. The meninges and bones of the base of the skull are commonly involved. Direct extension from nasopharyngeal carcinoma or primary tumour of the base of the skull are probably more common than metastatic tumours or other conditions causing basal meningitis. Clinically the patient presents with progressive cranial nerve palsies.

The pathological process of direct extension of tumours can affect the cranial nerves at their exit from the skull or in their extracranial pathways. Alternatively, the bones may be involved. In this case the cranial nerves are probably involved at the foramina

by direct extension of bony metastases. If the cranial nerves are involved inside the skull by meningeal disease, it is more likely to be accompanied by raised intracranial pressure, signs of meningeal irritation, bilateral cranial nerve palsies and abnormal CSF.

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