Occipital Condyle Syndrome, A Late Complication of Malignant Insulinoma

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Abstract

Occipital Condyle Syndrome (OCS) is a rare clinical syndrome caused by lesions at the base of the skull affecting the hypoglossal nerve. It is associated with various etiologies of tumoral and benign origin. Neurological symptoms associated with insulinomas have been only reported to hypoglycemia. Malignant insulinoma, defined by the presence of metastases, is rare. We describe a young woman with a malignant insulinoma with bone metastases who began to have severe unilateral occipital pain accompanied by dysarthria and dysphagia. An ipsilateral hypoglossal nerve paralysis was observed, and an MRI of the skull base disclosed an occipital condyle metastasis. With this case report, skull base metastasis of malignant insulinoma is added as a new cause of OCS. OCS is a diagnosis that clinicians should keep in mind in patients either with disseminated tumours or unknown cancers. Based on its stereotyped symptoms, OCS should be suspected promptly to undertake local radiotherapy.

Keywords: Insulinoma; Hypoglossal Nerve Diseases; Occipital Bone; Skull Base Neoplasms; Headache.

Introduction

Metastatic involvement of the skull base is a well-known complication of a variety of systemic cancers. In these cases, the clinical diagnosis is usually suspected by the presence of symptoms and signs secondary to the involvement of cranial nerves that exit through the basal foramina, together with persistent headache. The orbital, parasellar, middle fossa, jugular foramen and Occipital Condyle Syndrome (OCS) were the five clinical syndromes associated with skull base metastases described by Greenberg et al in 1981[1]. The OCS is characterized by unilateral occipital pain and unilateral tongue paralysis. Although a group of patients with OCS have a benign explanation, such as trauma, infection, stroke and Guillain-Barré syndrome, a wide variety of malignancies account for the remainder of cases of OCS. In fact, in these cases, the OCS is usually the first clinical manifestation of the neoplasm.

We report the first case of OCS associated with a malignant insulinoma.

Case report

This 57 year-old woman began in 2005 with recurrent neurological symptoms associated with hypoglycemia. A CT scan demonstrated a pancreatic mass. She underwent distal pancreatectomy and splenectomy, and the histopathological examination of the excised tissue revealed a pancreatic insulinoma. At that time, hepatic islet cell metastases were observed. The liver metastases had no change after two chemoembolizations with polyvinyl alcohol particles and treatment with octreotide and diazoxide was administered. In 2009, she underwent liver transplantation for the metastatic neuroendocrine tumour with no incidences. She was
Insulinoma is a rare neuroendocrine tumour with an annual incidence of 0.4 cases per 100,000 people. Insulinoma has malignant characteristics defined by metastases in only 10% of the cases [2,3]. Most patients with malignant insulinoma have lymph node or liver metastases and only rarely involve other organs, such as the skeletal system. The available treatments show only short-term benefits and the prognosis of these patients is relatively poor with a median survival period of approximately 2 years [2]. Insulinoma could be a feature of multiple endocrine neoplasia type 1 (MEN1) and approximately 4% of patients with insulinoma will have MEN1; in this case is very unlike a MEN1-associated insulinoma. An osteolytic lesion was found in the left clavicle by a thoracic CT examination. An MRI study of the skull base showed an enhancing soft tissue mass close to the left foramen magnum without intracranial invasion (Figure 1). A brain MRI was found to be normal. His general status deteriorated, and the patient died one week later.

**Discussion**

Insulinoma is a rare neuroendocrine tumour with an annual incidence of 0.4 cases per 100,000 people. Insulinoma has malignant characteristics defined by metastases in only 10% of the cases [2,3]. Most patients with malignant insulinoma have lymph node or liver metastases and only rarely involve other organs, such as the skeletal system. The available treatments show only short-term benefits and the prognosis of these patients is relatively poor with a median survival period of approximately 2 years [2]. Insulinoma could be a feature of multiple endocrine neoplasia type 1 (MEN1) and approximately 4% of patients with insulinoma will have MEN1; in this case is very unlike a MEN1-associated insulinoma. An osteolytic lesion was found in the left clavicle by a thoracic CT examination. An MRI study of the skull base showed an enhancing soft tissue mass close to the left foramen magnum without intracranial invasion (Figure 1). A brain MRI was found to be normal. His general status deteriorated, and the patient died one week later.

**References**


