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Case Report

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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

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on tacrolimus, which was changed to everolimus one year later. Two years after liver transplantation, a CT scan of the abdomen disclosed peritoneal, lymph node and hepatic metastases and capecitabine was added. Five days before his admission in February 2010, the patient began to have dysarthria, dysphagia, and progressive left occipital pain. He complained of severe, continuous, left occipital pain that became unbearable with left suboccipital palpation or on neck rotation to the right. Routine analgesics were of no help and morphine was administered to control pain. There were no cervical masses on palpation. Neurological abnormalities were limited to left hypoglossal nerve paralysis. 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.

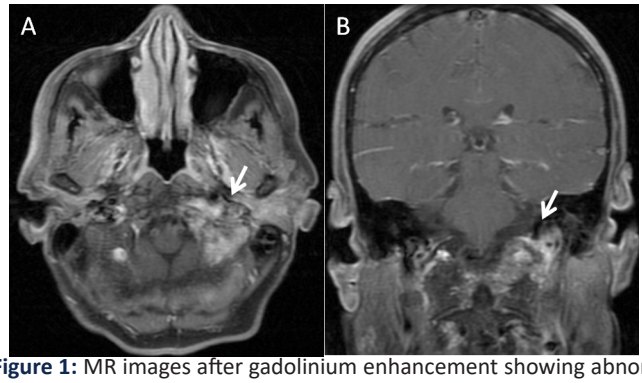

Figure 1: MR images after gadolinium enhancement showing abnormal signal in the left occipital condyle (white arrow) compatible with local metastases. A: Axial; B: Coronal postcontrast T1-weight.

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 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 based on the absence of a family history of MEN 1 and the absence of other clinical, biochemical or radiological characteristics of MEN 1 [4]. Due to the small number of patients with malignant insulinoma, there are little data regarding its neurological complications. Neurological symptoms are frequent in the initial phases of insulinoma. Hypoglycemia due to insulinomas mimics a great variety of neurological conditions and most patients with present with neurological or psychiatric manifestations that often lead to misdiagnosis. Symptoms from central nervous system glucose deprivation could persist for years since the diagnosis due to unregulated secretion of insulin and proinsulin-related products from malignant insulinoma [5,6]. In addition, peripheral neuropathy is unusually reported as a neurological complication in the course of insulinoma probably due to maintained hypoglycemia rather than secondary to hyperinsulinaemia [7]. To our knowledge, no other neurological features of insulinoma have been reported except symptoms due to hypoglycemia of either the central or peripheral nervous system.

The hypoglossal nerve arises from the motor nucleus located beneath the floor of the fourth ventricle, passing in front of the vertebral and posterior inferior cerebellar arteries. It exits the base of the skull through the hypoglossal canal in the occipital bone; it then traverses the neck and curves back, divides and innervates the tongue muscles. Metastatic involvement of the occipital condyle originates the OCS, which is characterized by unbearable unilateral occipital pain and ipsilateral tongue paralysis. Several malignant causes have been described as a cause of OCS, including metastases from different types of tumours [1,8-15]. but we have found no reference in the English language literature to isolated hypoglossal neuropathy caused by insulinoma metastases to the base of the skull.

When OCS is suspected based on its stereotyped symptoms, CT and MRI are the diagnostic steps. The MRI is better for delineating soft tissues. The sections of CT and MRI must be low enough to show the foramen magnum area to detect the suspected lesion. In patients with skull base metastases known to have untreatable cancer, local palliative radiation therapy should be undertaken as soon as possible [16]. When this treatment is delivered early, symptomatic relief can be expected in most patients. In patients without known systemic cancer, skull base metastases can be the first manifestation of neoplasia and a search for a primary source is indicated in patients with OCS.

Conclusion

In conclusion, we added malignant insulinoma as a cause of OCS; clinicians should keep in mind the diagnosis of OCS even in tumours for which OCS has not been reported in the literature. In addition, this description of malignant insulinoma contributes to a better understanding of its clinical course to clarify the role of therapeutic procedures in the disease.

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