

Occipital Condyle Syndrome as an Initial Presentation of Lung Cancer: A Case Report

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Abstract-

Purpose: Occipital condyle syndrome (OCS) is a rare cause of headache. This study herein reports a case in which a unique headache and tongue deviation appear as symptoms of the first presentation of a malignant tumor.

Case Report: A healthy 67-year-old male presented with a unilateral shooting pain in the occipital region, accompanied by slurred speech and difficulty swallowing. Neurological examinations later revealed atrophy and mild fasciculation of the tongue.

The clinical symptoms and MRI results suggested OCS. Screening for tumor markers showed an elevated CEA. The chest CT revealed a lobulated soft-tissue mass in the lower left lobe, and a CT-guided biopsy confirmed the diagnosis of adenocarcinoma. A whole body bone scan found multiple foci. The adenocarcinoma was graded pT2bN3M1b, stage IV.

The headache improved with a prescription of prednisone, 60 mg to be taken daily. With three months of treatment, clinical examinations showed that the patient was free of pain and that there had been no progression of the atrophy or deviation of the tongue.

Conclusion: The possible etiology of OCS includes a primary tumor or metastatic lesion that directly invades the base of the skull. Determining the underlying causes of OCS can be challenging, but MR imaging is currently the diagnostic tool of choice. An awareness of the features of OCS in healthy adults may be able to lead to earlier diagnosis of the underlying etiology and efficient relief of the symptoms.

Key Words: Occipital condyle syndrome, Skull base metastasis, Hypoglossal nerve palsy

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INTRODUCTION

Occipital condyle syndrome (OCS), a rare cause

of pain in the head, consists of a severe and persistent unilateral sub-occipital headache with ipsilateral 12th cranial nerve palsy.

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Its symptoms are usually worsened when the head is rotated to the non-painful side, causing patients to hold their heads with their hands and is simultaneous with or antedated by, from a few days to a few months, paralysis of the ipsilateral hypoglossal nerve. Persistent unilateral occipital pain with specific trigger factors of OCS differs from neuralgia and myalgia.

The possible etiology of OCS includes inflammation, a primary tumor or a metastatic lesion that has directly invaded the base of the skull. This study herein reports a case of a unique headache and tongue deviation as the first signs of a malignant tumor.

CASE REPORT

In December 2010, a previously healthy 67-year-old male arrived at our neurological department complaining of a continuous, severe, unilateral shooting headache (his score on the visual analogue scale was 9) over the occipital region and radiating to the ipsilateral temporal and forehead that he has been experiencing for 2-3 weeks. The precipitating factors were neck flexion and head rotation to the non-painful side. The headache was not relieved by NSAID or muscle relaxants. He developed mildly slurred speech and difficulty swallowing four days before admission. A CT scan of the brain at the local hospital showed unremarkable findings. The patient had no history of trauma, weight loss, night sweats or recent fever. Upon admission, neurological examinations revealed atrophy and mild fasciculation of the tongue with deviation to the

right when protruding, findings that indicated hypoglossal nerve palsy.

Magnetic resonance (MR) imaging of the brain revealed a marrow-replacing lesion in the clivus and right occipital condyle (Figure A) with encasement of the right hypoglossal canal (Figure B). The clinical symptoms and MR imaging results pointed towards occipital condyle syndrome (OCS)⁽¹⁾.

The subsequent screening for tumor markers revealed an elevated CEA (499 ng/ml). Furthermore, a chest radiograph found a patchy opacity in the left retro-cardiac region, and a CT scan of the lung revealed a lobulated soft-tissue mass measuring 5.5 cm in the medial aspect of the lower left lobe (Figure C). A CT-guided biopsy of the lung confirmed the diagnosis of moderately differentiated adenocarcinoma. A whole body bone scan revealed multiple foci of increased uptake in the skull base, spine, ribs, pelvic bones, and both scapulae. The adenocarcinoma was graded pT2bN3M1b, stage IV.

The headache improved with a prescription of prednisone, 60 mg to be taken daily. The dose was then tapered following the start of concomitant chemotherapy (Cisplatin and Gemcitabine HCL) and radiotherapy (200 cGy x 15 fractions of the whole brain and then 1.86 cGy x 13 fractions for the lesion). After three months of the treatments, clinical examinations showed that the patient was free of pain and that there had been no progression of the atrophy or deviation of the tongue. Nonetheless, the patient died due to respiratory failure six months later.

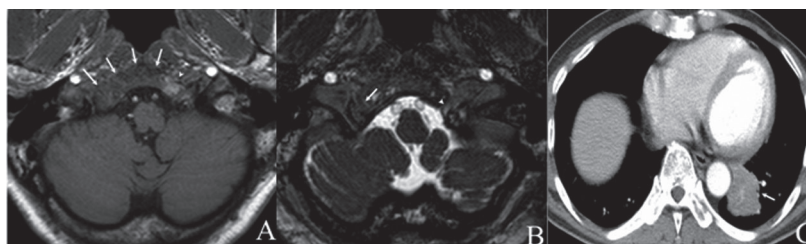


Figure 1. MRI of the brain and CT of the chest.

(A) T1-weighted image demonstrates a marrow-replacing tumor in the middle and right aspect of the clivus (arrows). In the left aspect of the clivus shows a small normal-appearing fatty marrow (arrowhead). (B) On high-resolution T2-weighted image, the right hypoglossal canal (arrow) is obliterated while the left one (arrowhead) preserved. (C) Post-contrast CT of the chest shows a poorly-enhancing mass, 5.5 cm, in the medial aspect of the left lower lobe (arrow).

DISCUSSION

The hypoglossal nerve stems from the motor nucleus located beneath the floor of the fourth ventricle, passes in front of the vertebral and posterior inferior cerebellar arteries, exits the base of the skull through the hypoglossal canal in the occipital bone, and then traverses the neck. Finally, the nerve splits and innervates the tongue muscles. A wide variety of lesions along the nerve's tract can cause peripheral hypoglossal nerve palsy.

The occipital condyles are small, bilateral inferior extensions of the occipital bones that form part of the lateral aspect of the foramen magnum. Therefore, the abnormalities involving an occipital condyle such as trauma, a primary tumor, a metastatic lesion or inflammation that directly invades the skull base may result in lower cranial nerves neuropathy.

In 1981, *Greenberg et al.* evaluated 43 patients with skull base metastases and described the following five distinct clinical syndromes: the orbital, parasellar, middle fossa, jugular foramen, and occipital condyle syndromes⁽²⁾. Of these, the occipital condyle syndrome (OCS) was most clearly associated with the stereotypical clinical presentation consisting of unilateral occipital region pain and ipsilateral 12th cranial nerve (hypoglossal nerve) palsy.

Since then, several cases of OCS have been reported among medical literature. In 1998, *Moris et al.* reported four cases of OSC as the first clear clinical manifestation of a malignant tumor⁽³⁾. In 2002, *Capobianco et al.* reported 11 cases of OCS⁽⁴⁾. In 2006, *Salamanca et al.* presented a study on another OCS patient⁽⁵⁾. In 2007, *Moeller et al.* also reported two cases of OCS as the first symptom of metastatic cancer⁽⁶⁾. Furthermore, in 2010, *Bahl et al.* published a study about a case of small cell lung carcinoma presenting as OCS⁽⁷⁾. Of these patients, 10 cases had no prior history of cancer. Furthermore, only two of the aforementioned patients had lung carcinoma (one small cell lung carcinoma and the other squamous cell carcinoma) that was causing OCS^(6,7). As of now, this study features the third patient whose lung cancer presented with OCS as an initial symptom.

The patient experienced a unique pattern of a severe, continuous, unilateral pain in the occipital region, whose symptoms improved by turning the head toward the

painful side while the pain worsened with contralateral head-turning in our patient, as well as in almost all of the 28 previously reported cases of OCS. Physical findings were usually limited to an isolated unilateral hypoglossal paresis and mastoid tenderness with no other abnormalities found in the neurological examination. In most of these cases, headache often preceded tongue paresis by days to weeks.

In our patient and in others, diagnosing the underlying cause of occipital condyle syndrome can be a challenge. The utility of cytological examination of the cerebrospinal fluid (CSF) can be difficult to establish. Of the reported cases, only three patients underwent a lumbar puncture for CSF analysis. However, in all three cases, it was negative. Imaging techniques such as standard computed tomography (CT) of the skull base often do not reveal abnormalities in the occipital condyle. In our patient, as in previously reported cases, magnetic resonance imaging (MRI) was more sensitive for detecting abnormalities in the occipital condyle⁽⁸⁾. Said imaging often revealed a marrow-replacing lesion in the clivus and a right occipital condyle with encasement of the right hypoglossal canal of the skull base, pointing to the possibility of OCS. The whole body bone scan of our case also showed multiple foci of increased uptake in the skull base, spine, ribs, pelvic bones, and both scapulae, which indicated multiple bony metastases. This finding suggests that a radionuclide bone scan or a high-quality CT may complement an MRI for future diagnosis^(9,10).

According to the existing research, a wide variety of malignancies can cause OCS, including metastases from solid tumors of the breast⁽¹¹⁾, lung⁽⁷⁾, prostate⁽¹²⁾ and gastrointestinal tract⁽¹³⁾, as well as primary pharyngeal tumors and lymphoma⁽⁸⁾. Only a small fraction of patients with OCS have a benign cause for their symptoms. In a series of 100 patients with twelfth nerve palsy, over half of the patients with non-traumatic tongue paresis had a malignancy⁽¹⁴⁾. However, the diagnosis of a malignant tumor is often delayed by several months. The diagnosis of OCS can be elusive, and diagnostic imaging of the skull base may appear normal on initial presentation. An awareness of the characteristics of OCS in healthy adults may be able to lead to earlier diagnosis of the underlying etiology and the efficient relief of symptoms.

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