Case Report

Occipital condyle syndrome guiding diagnosis to metastatic prostate cancer

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Abstract Occipital condyle syndrome (OCS) results from a unilateral occipital pain associated with an ipsilateral paresis of the 12th cranial nerve (hypoglossal), and is typically caused by metastasis of the skull base. OCS diagnosis occurred, in all cases described in the published literature, when metastatic prostate cancer (MPC) was previously known. We present a case of a patient whose initial manifestation of MPC was OCS. The patient was treated with complete hormonal blockade and non-steroidal anti-inflammatory drugs as opposed to locoregional radiotherapy applied in other cases. After 18 month follow-up, the patient had a complete neurological and biochemical response.

Key words metastasis, occipital condyle syndrome, prostate cancer.

Introduction

Prostatic cancer is the leading cancer diagnosis and the second most common cause of cancer-related death in men in the USA. Reviewing the published literature, the most frequent presentation of prostate cancer (PC) affecting the central nervous system is spinal cord compression caused by bone metastasis. A study performed by Saitoh et al. in deceased patients as a result of prostatic cancer, concluded that 15% were affected with lumbar spine metastasis whereas only 2% had skull dissemination.1 Lynes et al. described that less than 1% of patients with PC presented with clinical brain metastasis.2 Occipital condyle syndrome (OCS) results from a unilateral occipital pain associated with an ipsilateral paresis of the 12th cranial nerve (hypoglossal) and is typically caused by metastasis of the skull base. This syndrome was first described by Greenberg in 1981.3–5 In a recent review of a retrospective case series of 11 patients only four patients suffered PC and their disease was discovered when nerve palsy occurred.4 OCS diagnosis occurred, in all cases described in the published literature, when metastatic prostate cancer (MPC) was previously known.

We present a case of a patient whose initial manifestation of metastatic prostate cancer was occipital condyle syndrome.

Case report

A 66-year-old male complained of a constant unilateral daily headache, which had begun in the previous month. The pain was located on the right temporal region and was predominantly retro auricular. It improved with NSAI and initially did not interfere with sleep or daily activities. However, the pain became increasingly intense and 3 weeks after the onset of the headache he noticed difficulty in pronouncing certain letters and moving the tongue while chewing and speaking. The patient denied other neurological or general symptoms and had no relevant past clinical history, except erectile dysfunction. As a result of this, he had a routine prostatic revision with serum prostate-specific antigen (PSA) of 3.9 ng/mL (free PSA 1.87 ng/mL) and a normal digital rectal examination 1 year previously. The neurological examination revealed tenderness of the mastoid region and right twelfth nerve palsy with ipsilateral tongue atrophy (Fig. 1).

A standardized CT scan was normal. Skull base CT showed an osteolytic lesion of the right occipital condyle (Fig. 2). Magnetic resonance imaging confirmed an abnormal signal in the mentioned region (Fig. 3). Bone scan identified multiple pathological depositions (osteoblastic) in skull base, axial and appendicular skeleton and soft tissue of left iliac crest. Urological investigations included PSA serum levels of 22.7 ng/mL and transrectal ultrasound guided prostatic biopsy revealed a prostatic adenocarcinoma Gleason grade 4+4 on the left lobe and high grade displasia (PIN III) on the right lobe. With a diagnosis of MPC and OCS caused by skull base metastasis, the patient began treatment with complete hormonal blockade (Bicalutamide 50 mg daily and Gosereline 3.6 mg every
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The pain was treated successfully with NSAIs (Acetaminophen and only initially with dipirone). Eighteen months after the onset of treatment the patient was neurologically asymptomatic. He had recovered tongue mobility and trophism, his PSA serum level was undetectable (<0.04 ng/mL), and bone scan only showed a minimal captation on the left iliac crest.

Discussion

The incidence of prostate cancer has increased substantially since the 1970s. These trends are characteristic of the introduction of early detection methods. Since the use of PSA testing, the diagnosis of local-regional disease has increased whereas the incidence of metastatic disease has decreased.

Most authors concluded that the rise and fall in PC incidence and migration from advanced to early stages of disease could largely be attributed to early detection. After comparing prevalence figures in autopsies and PC clinical incidence, the conclusion is that the average duration of asymptomatic disease is 10–12 years. The behavior of Prostatic cancer varies, showing a wide clinical spectrum.

This tumor rarely causes symptoms early in the course of the disease because the majority of adenocarcinomas arise in the periphery of the gland distant from the urethra. The presence of symptoms as a result of prostate cancer suggests locally advanced or metastatic disease. Schaberg et al. referred that more than one third of patients with metastatic disease are asymptomatic at diagnosis.

Metastatic disease involving the axial or appendicular skeleton can cause bone pain or anaemia from replacement of the bone marrow. Patients with symptoms caused by neurological involvement are not infrequent in MPC. Nevertheless, cases in which disease begins with neurological symptoms are rare. Approximately 20% of all PC cases will develop neurological symptoms. The term ‘hidden carcinoma’ is applied to cases in which the diagnosis is based on the symptoms derived from metastasis. Among PC neurological manifestations we can include those secondary to bone affection and subsequent nervous compression and, less frequently, spinal or intracranial parenchymal lesions.
Of the five clinical syndromes of skull base metastasis described by Greenberg et al., OCS has the most constant findings. The cardinal sign is isolated palsy of the twelfth nerve, either preceded or associated with a characteristic ipsilateral headache. The first complaint is generally dull increasingly intense occipital pain, which is spontaneous and exacerbated by head rotation. Weeks after the onset of the headache, the patient notices difficulty pronouncing and swallowing. Neurological examination reveals tenderness in the suboccipital region and a peripheral hypoglossal nerve paresis with tongue deviation towards the side of the pain and hemilinguatrophy.

In the OCS, the twelfth nerve is extrinsically entrapped as it exits the skull through the anterior condylar foramina or hypoglossal canal. A wide variety of lesions along the nerve’s tract at other points can cause peripheral hypoglossal palsy: tumoral (Schwannoma), infectious, carotid dissection or endarterectomy, cervical trauma, among the most reported. The clinical picture in these cases may result in isolated XII palsy or multiple bulbar cranial nerve paresis.

What is unique to the OCS is the typical headache associated with the palsy, which narrows the differential diagnosis to tumoral or inflammatory affections in the occipital condyle region. The most common cause is metastasis of the breast or prostate cancer, followed by colorectal, renal cell, nasopharyngeal, primary bone tumors, meningiomas and lymphomas.

There is remarkable data in the present patient that distinguishes the case from others previously reported. The occipital region pain followed by the hypoglossal palsy were the guiding symptoms to diagnosis, not having encountered any reference in the literature of OCS being the initial presentation symptom of metastatic prostatic adenocarcinoma. Both CT and MRI findings were pathological and radionuclide bone scan disclosed multifocal bone metastasis.

Evolution in PSA values, from located to disseminated disease, was extremely quick (less than 1 year), and clearly related to tumoral undifferentiation (Gleason 8). The patient had a complete neurological and biochemical response to complete hormonal blockade after an 18 month follow-up, as opposed to locoregional radiotherapy applied in other cases.

References