Case Report

Spontaneous Intracranial Hypotension Presented as VI Cranial Nerve Palsy in a Breast Cancer Patient: A Case Report

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Abstract

We report a case of a 58-years old woman with previous history of breast cancer treated with nipple-sparing mastectomy with axillary lymphadenectomy and posterior adjuvant chemotherapy, radiotherapy and hormonal therapy. The patient consulted for horizontal diplopia and headache being diagnosed of right VI cranial nerve palsy. Cranial MRI revealed dural enhancement suggestive of meningeal carcinomatosis but without exclude intracranial hypotension. MRI myelography was made finding CSF collection in T1-T2 foramen conjunction, larger at left side, suggestive of point of CSF leakage. In lumbar puncture was detected low opening pressure of CSF, but without any malignant cell in cytology. The patient was diagnosed of SIH and treated successfully with conservative treatment. We have to take into account differential diagnosis with other benign diseases, as SIH, in cancer patients. In our case MRI finding of dural enhancement was suspicious of meningeal carcinomatosis, but this hypothesis was discarded after a complete study. In conclusion, SIH is a cause of orthostatic headache that we have to suspicious by symptomatology but we must confirm it by cranial MRI and MRI myelography. Conservative treatment is the first choice, but it also can be treated with epidural blood patch or surgery.

ABBREVIATIONS

MRI: Magnetic Resonance Imaging; CSF: Cerebrospinal Fluid; SIH: Spontaneous Intracranial Hypotension

INTRODUCTION

Neurological symptoms are relatively frequent in cancer patients due to secondary involvement of central nervous system (CNS). Spontaneous intracranial hypotension (SIH) syndrome is manifested mainly with orthostatic headache or with other neurological symptoms as cranial nerve palsies, parkinsonism, ataxia or coma. The magnetic resonance imaging (MRI) findings as pachymeningeal contrast enhancement can mimic meningeal carcinomatosis, thus is very important to reach the correct diagnosis in this type of patients [1]. We present a clinical case report of a woman with previous diagnosis of breast cancer and diplopia.

CASE PRESENTATION

A 58-years old woman, with history of pyrazolone allergy, asthma and former smoker since twenty years ago who works as psychologist, was diagnosed of invasive ductal breast cancer after screening mammography. The patient underwent a nipple-sparing mastectomy with axillary lymphadenectomy confirming the diagnosis of invasive ductal carcinoma and mixed invasive lobular-ductal carcinoma with estrogen-receptor positive, progesterone-receptor positive and positive expression of Her-2 by immunochemistry (3+). Two of the eleven lymph nodes isolated were positive for carcinoma, so she was diagnosed of breast cancer pT2 pN1a (2/11) cM0 (TNM 7th edition Stage IIB).

Adjuvant chemotherapy with doxorubicin 60mg/m2 day 1 intravenous (iv), cyclophosphamide 600mg/m2 day 1 iv every 21 days during 4 cycles followed by weekly paclitaxel 80mg/m2 iv with weekly trastuzumab (charge doses 4mg/m2, subsequent doses 2mg/m2) during 12 weeks was administered. Afterwards, the patient began the treatment with tamoxifen 20mg/day and she completed one year of treatment with trastuzumab 6mg/kg iv every 21 days. External radiotherapy was delivered along
tumor bed and axillar and supraclavicular lymph nodes to 50Gy total dose (2Gy/day).

After 17 months of cancer diagnosis, the patient went to the hospital for binocular horizontal diplopia that started 4 days ago associated with increasing intensity headache during the last 2 weeks without worsening in an upright position. The patient only remarked the acquisition of a new dog with frequent pulls in right arm during walks. No other symptomatology was reported by her.

She was diagnosed of right VI cranial nerve palsy by ophthalmologist. Physical examination did not reveal another alteration. The Cranial Computed Tomography (CT) scan without intravenous contrast showed multiple hypo-isodense laminar collections at bilateral convexities without mass effect suggestive of sub-acute-chronic subdural hemorrhages.

Cranial MRI was performed revealing dural thickening in the convexity of cerebral hemispheres, tentorium and cervical spinal cord along C1-C4 suggestive of meningeal carcinomatosis, but without discarding intracranial hypotension (Figure 1 & 2). Positron Emission Tomography (PET) excluded visceral tumor spread.

A non-traumatic lumbar puncture was made with an opening pressure of 30mm H 2O and pressure with Valsalva maneuver of 60mm H 2O, compatible with hypotension of cerebrospinal fluid (CSF). CSF cell count was 7 leucocytes/mm³, glucose was 44mg/dl and proteins were 47mg/dl. The cytology of CSF was negative for malignant cells.

In cervical, thoracic and lumbosacral MRI-myelography we found an increased anterior epidural space in C1-C2 level with enhancement after the administration of gadolinium and a dural thickening along posterior wall of the spinal canal with maximum expressiveness in medium and low thoracic region.

Furthermore, a CSF collection over vertex area of left lung with asymmetry of T1-T2 foramen conjunction, larger at left side, suggestive of point of CSF leakage was detected. All of these findings were compatible with diagnosis of spontaneous intracranial hypotension.

The patient had to be off work and she received treatment with bed rest, oral caffeine, adequate fluid intake and close follow up. After three weeks, we observed minor or improvement of diplopia and persistence of non-orthostatic headache. Treatment with tamoxifen was stopped to facilitate the follow up because headache is an adverse event of this drug. Afterwards, treatment with letrozole was initiated.

A new MRI was made 3 months after the diagnosis without relevant changes. The main symptomatology, diplopia and headache, had disappeared. The patient currently does normal life and she has returned to her job. At this time, no evidence of cancer relapse has been detected.

**DISCUSSION**

The SIH syndrome is characterized by neurological symptoms, mainly orthostatic headache, low CSF pressure and dural enhancement on MRI. In most of cases the syndrome is caused by spontaneous CSF leak in the cervicothoracic junction or along the thoracic spine [1,2].

This entity has experienced a raising incidence during the last decades due to a more accurate evaluation of patients and also to the specific image findings that facilitate a correct diagnosis.

The most frequent symptom is orthostatic headache that worsening within 15 minutes after adopts an upright position and improves after lying down [1-3]. Nonetheless, it is not unusual to achieve the diagnosis in patients with atypical symptomatology as no headache or other types of headache, cranial nerve palsies, coma, parkinsonism, ataxia, chorea, unilateral hearing loss or cerebellar hemorrhage [1,2,4].

Image findings are relevant to suspect this entity or to confirm the diagnosis. In cranial MRI we can observe pachymeningeal enhancement, sagging of the brain, engagement of venous structures and subdural fluid collections. MRI mielography is the gold standard imaging test for the diagnosis of spontaneous intracranial hypotension due to ability to localize CSF leak with a sensitivity of approximately 89% [1,5].

In our case, the woman debuted with right VI cranial nerve palsy that produced horizontal diplopia and non-orthostatic
headache. Cranial nerve palsies can be explained by the displacement of the brain after CSF lost and subsequently the stretching of the nerves [1].

Dural enhancement at cranial MRI can mimic meningeal carcinomatosis as in this case. Therefore, it is especially important to consider other benign diagnosis before to attribute it to cancer dissemination.

Capecitabine has been related with intracranial hypotension in cancer patients in one case report [4]. In our case, we did not identify any causal relationship with the treatment received.

Conservative treatment consisting of bed rest, adequate fluid intake and oral caffeine, is the first choice of treatment. If this approach fails, epidural blood patch application is an alternative. Surgical repair of CSF leak is the last alternative of treatment only reserved for those patients in whom non-surgical treatments have failed [1,2,4].

In conclusion, SIH is a syndrome that we have to consider in patients with atypical headache or with typical image findings. Patients with previous history of HER2 positive breast cancer have a risk of leptomeningeal dissemination around 15% [6], and because of that we have to take it into account for the differential diagnosis. Nevertheless, we also have to consider other non neoplastic diagnostics due to the different prognosis and treatment of the entities.

CONFLICT OF INTEREST

All authors declare not to have potential conflicts of interest and not to have received reimbursements, fees, funding, or salary from an organization that may in any way gain or lose financially from the publication of this manuscript, either now or in the future.

REFERENCES