Clinical Note

Peritoneal carcinomatosis by pineoblastoma secondary to ventriculoperitoneal shunt

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Summary

The primitive neuroectodermal tumors (PNETs) produce metastases in the basicranial axis. Extracranial metastases are infrequent and occur by dissemination through the cerebrospinal fluid (CSP). Sixty two cases of peritoneal carcinomatosis have been reported to date as due to a ventriculo-peritoneal shunt (VPS), four of them by pineoblastoma. We present the case of a pineoblastoma showing this extremely rare evolution and its response to platinum compounds and taxanes. We reviewed the literature which relates to this neoplasm and its treatment.

Key words: Carcinomatosis. Chemosensibility. Pineoblastoma. Ventriculoperito-neal shunt.

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Resumen

Los tumores primitivos neuroectodermicos normalmente producen metástasis en el eje craneoespinal. Las metástasis estracraneales por estos tumores son infrecuentes y son debidas a diseminación a través del líquido cefalorraquídeo. Hasta la fecha, han sido descritos en la literatura 62 casos que nos informan sobre carcinomatosis peritoneal secundaria a diseminación a través de un shunt ventriculoperitoneal, cuatro de los cuales se han debido a pineoblastomas. Presentamos el caso de un pineoblastoma que evoluciona de esta forma tan poco frecuente y como responde a quimioterapia con platinos y taxanos. Después haremos un repaso de la literatura descrita hasta el momento actual sobre esta patología.

Palabras clave: Carcinomatosis peritoneal. Quimiosensibilidad. Pineoblastoma. Shunt ventriculoperitoneal.

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Introduction

Parenchymal pineal tumors are uncommon in adults account for 3%-8% of pediatric intracranial neoplasm. Extracranial metastasis is rare and is disseminated by CSF. Malignant Germinoma (27%), Medulloblastoma (19%) and endodermic sinus tumors are the most frequent causes of extracranial dissemination through VP-S The literature contains 62 reported cases of peritoneal carcinomatosis by dissemination through VP-S, four of these by pineoblastoma.

The time-lapse between placement of VP-S and the appearance of abdominal metastasis is about 17 months and depends on brain tumoral histology. Mortality of 62% has been reported 9 years after VP-S as a consequence of both first (*de novo)* disease and abdominal metastasis. Survival time varies from 1 month with Medulloblastoma to 9 years with Germinoma.

Clinical case

In January 1999 a nineteen-year-old woman was admitted with tonic-clonic seizure. Her family reported headache, vomiting, diplopia and vertigo two months before. Medical examination found giddiness, inability to spontaneously open her eyes, mumbled response to questions; she located pain and had isocoric and normoreagent pupils. Blood tests showed leukocytes 13900/mm^3 (86,8% Neutrophyles). CSF analysis found glucose 72 mg/dl, proteins 62 mg/dl, leukocytes 80/mm^3 (70% Lymphocytes), red blood cells 300/mm^3, with beta-HCG and alfa-fetoprotein negative. CSF cytology did not show neoplasm cells. Brain computer tomography (CT) showed an intracranial expansive lesion of 1.5 cms near the IV ventricle with homogeneous contrast enhancement and moderate ventricular dilation. MRI with homogeneous gadolinium enhancement confirmed the presence of a pineal tumor. An external ventricle shutting device was introduced to counter hydrocephalia; it was removed a week later when the patient improved. During planning of radiotherapy, neurologic impairment occurred due to another hydrocephalia, and a VP-S was introduced. Forty-five days of clinical debut radiotherapy treatment was started with the following doses: holocraneal radiotherapy with 24 Grays (Gys), pyneal and hypophyseal overloading with 47 Gys and spinal radiotherapy with 25.5 Gys. Complete remission (CR) with this treatment was achieved until April 2000, when she reported hypogastric colic pain of three days duration and vulvar edema. Medical examination showed perineal lymphedema and an irregular hard formation, fixed to the posterior face of the vagina, approximately 10 X 8 cms in size. Blood tests found haemoglobin 8.1 gr/dl, hematocrit 24%, BUN 48 mg/dl and creatinine 2.6 mg/dl. Abdominal echography showed a right adnexal mass of 7 cms. Abdominal CT showed peritoneal and omental implanting with free abdominal liquid, retroperitoneal adenomegalies and left obstructive uropathy. Craneospinal MRI did not show tumor. The new CSF analysis was negative. Diagnostic laparotomy showed a right adnexial mass and a transverse mesocolon mass. Biopsy of these was taken. Pathological anatomy diagnosis was ovarian and mesocolonic metastasis from grade IV pineoblastoma.

We had a patient with peritoneal carcinomatosis by dissemination of pineoblastoma through VP-S fifteen months after the first diagnosis of tumour. Chemotherapy was started with cisplatine 100 mg/m^2 day 1, etoposide 100 mg/m^2 day 1-3 and vincristine 1.5 mg/m^2 day 1 for six cycles, with doses of 80% because of previous radiotherapy. She achieved complete remission of abdominal lesions. After five months, peritoneal carcinomatosis reappeared and a second line of chemotherapy was started with Taxol 175 mg/m^2 and carboplatin (AUC 5). The patient had important hematologic toxicity grade IV, therefore the second and third cycles were administered with doses reduction of 80% and G-SCF. She achieved partial remission although hematologic toxicity persisted. Chemotherapy intolerance prompted a change to a third line of treatment with 60 mg/m^2 Taxol weekly, which she received in eight cycles. She had disease-free progression for six months. Then, progression followed with bilateral pleural effusion, ascities, severe undernutrition and paraneoplasm hypercalcemia. At this point we started a fourth line of chemotherapy with Topotecan. Treatment was suspended because her clinical condition worsened. The patient died three years after diagnosis.

Discussion

Pineoblastoma is a parenchymal pyneal tumor included in primitive neuroectodermal tumors (PNETs). It accounts for 12% of pyneal tumors and is more frequent in young people[1]. Pineoblastoma is an aggressive tumor with 25% survival to 3 years after surgery and radiotherapy. Its histology is poorly differentiated neuroepithelium cells. It produces metastasis in the basicranial axis through spontaneous or iatrogenic way. Thus, MRI examination of basicranial axis and CSF cytology are important. The clinical syndrome consists in symptoms of acute intracranial hypertension. Leptomenix dissemination is frequent at the time of diagnosis[2]. Diagnostic procedures include CT and MRI, CSF cytology and pathological anatomy confirmation. Because the latter is technically difficult, it is usual to start with an empirical treatment according to an approximate diagnosis. Later, depending on response to treatment, final diagnosis is made[3].

Pineoblastoma treatment is complete surgical resection, difficult to perform because of its location and its infiltrated nature, so surgery mostly consists of performing an internal or external derivation[4]. Radiotherapy is mandatory in the treatment of both parenchymal and germinal pineal tumors[5]. Survival to 5 years after treatment ranges from 44% to 78% and depends on tumoral histology[6], disseminated disease, age at diagnosis, irradiation volume and doses given to the first tumor location[1]. Standard treatment is holocraneal radiotherapy with 24 Gys, overloading tumoral radiotherapy up to 50-60 Gys and craniospinal radiotherapy with 34 Gys on spinal cord. This one only in cases of tumor with a high risk of spinal cord dissemination, such as this case, positive cytology in CSF or signs of radiologic lesion. Prognosis in children with pineoblastoma treated solely with chemotherapy is poor; thus many studies on radiosensitive chemotherapy are being carried out[7,8]. One study reports using cisplatin 100 mg/m^2 day 1, etoposide 100 mg/m^2 days 1-3 and vincristine 1,5 mg/m^2 day 1 for four cycles and subsequent craniospinal radiotherapy, which was given to three patients. In this study, one patient achieved complete remission for two years, another complete remission for five years, and the third had progression after five months. This study found disease-free progression about 24-60 months after diagnosis with moderate hematologic toxicity[7]. Another study used high doses of cyclophosphamide 2 gr/m^2, days 1-2, monthly during 4 cycles. This study included seven patients, two with recurrent disease. After treatment these six patients achieved complete remission[9]. Only one patient with recurrent disease had progression disease. Another study compared a combination of chemotherapy (CCNU, vincristine and prednisone) and radiotherapy stand up to the same chemotherapy alone. Combined radiotherapy and chemotherapy achieved disease free progression of 61% for three years in seventeen patients, of whom twelve had partial remission at the end of treatment and remained with stable disease five years later. Four patients had progression disease. Chemotherapy alone achieved fourteen months of disease free progression in seven patients[10]. In this study the most important toxicity was secondary to radiotherapy with neurotoxicity. Chemotherapy alone for PNETs was unsuccessful; nevertheless chemotherapy with radiotherapy and surgery can increase disease free time, although global survival is similar[10]. In our case, we found chemosensibility of pineoblastoma first to cisplatin and etoposide with a complete remission and then to carboplatin and taxol, with partial remission and clinical improvement for some months.

However, optimal therapeutic approach to these patients remains unclear and requires re-evaluation.

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