BLINDNESS AS AN INITIAL MANIFESTATION OF MENINGEAL CARCINOMATOSIS

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SUMMARY :

The term meningeal carcinomatosis refers to diffuse metastasis of the leptomeninges by systemic cancer. The diagnosis is suggested by signs and symptoms of headache, dementia, meningeal irritation and involvement of multiple cranial or spinal nerve roots. Blindness as an initial manifestation of meningeal carcinomatosis is very rare without evidence of a primary carcinoma. We describe the clinical course of a patient with meningeal carcinomatosis whose initial symptom was blindness.

KEY WORDS :

Blindness, meningeal carcinomatosis, systemic cancer.

INTRODUCTION

Meningeal carcinomatosis(3) has been recognised since the 1900's. Symptoms and signs of diffuse leptomeningeal metastases from a primary carcinoma outside the central nervous system may be the first evidence of malignant disease(2,6). The clinical findings can be classified: Brain, cranial nerve, spinal nerve and spinal cord(7). Although cranial nerve involvement has been reported in up to 94 percent of patients during the course of the disease, involvement of the second cranial nerve appears to be less common(1,4,8). We present a case of meningeal carcinomatosis of which the initial manifestation was blindness.

CASE REPORT

A 57-year-old man consulted the ophthalmology department on February 10, 1988 for evaluation of sudden loos of vision in his right eye. At that time ophthalmoscopic examination showed advanced pallor of the right optic disc. No retinal or choroidal lesions were observed. Intraocular pressure was normal. Total blindness of the right eye was noted. Ocular motility, visual fields, anterior segments and intraocular pressure were normal. One month later he was admitted to the neurology department for nonspecific headache and severe pain in his right leg. On examination the patient was disorientated and restless. Blood pressure was 120/90 mmHg. Neurological examination showed involvement of spinal nerve roots with decreased Achilles reflex, positive Laseques sign and hypoaesthesia or L4-5 and S1 levels in the right leg. Shortly thereafter he developed peripheric facial palsy and decreased visual acuity also in this left eye. He underwent extensive studies. X-rays of the skull and chest, gastrointestinal tract series, abdominal ultrasonography and analyses of urine showed no abnormality. Erythrocyte sedimentation rate was 102 mm/hr. Other analyses of blood were normal. Electroencephalography showed diffuse slowing of the background activity. CT scan showed mild ventricular dilatation.

Two lumbar punctures were carried out at one week intervals. The first spinal fluid contained 410 mg/dl of protein and 40 mg/dl of glucose with 20 white cells per cubic millimeter (WBCs.) VDRL was nonreactive. Stains and cultures for bacteria and acid-fast bacilli, and cytological examination for malignant cells were negative.

The second CSF disclosed a high level of protein (510 mg/dl) and cytology was reported positive for malignant cells (Figure 1). Repeated laboratory and radiological studies including CT and whole lung tomograms were normal.

The patient was started on systemic I.V. chemotherapy and brain radiation. However, the vision in both eyes and his mental condition progressively deteriorated. In April 1988 he died of heart failure. Permission for autopsy was refused.

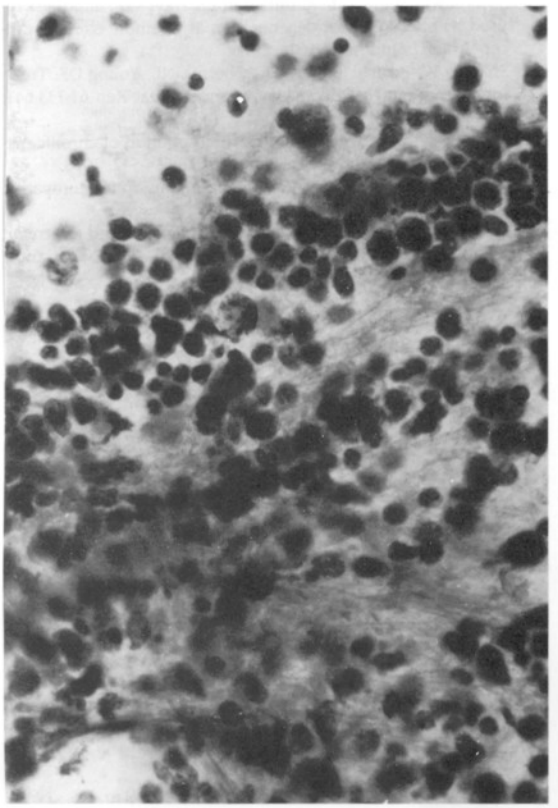

Fig. 1 : Cytological examination demonstrated malignant cells in cerebrospinal fluid.

DISCUSSION

Meningeal carcinomatosis is thought to be a rare condition, but according to reports published during the past ten years, it is likely to appear in up to 8 percent of patients with systemic cancer. Although early reports suggested that primary tumours of the gastrointestinal tract were the most frequent cause, today it is known that breast and lung are the most frequent sites and malignant melanoma is the third most common primary tumour(3,5,9).

The lack, as in our case, of a known primary malignancy does not preclude this diagnosis. And in many studies, 48-75% of patients with clinical manifestations of meningeal carcinomatosis had no previous history of cancer(2,8).

Clinical suspicion of the condition rests on the finding of neurological signs and symptoms at more than one level of the neurological signs and symptoms at more than one level of the neuroaxis. The most frequent symptoms and signs of cerebral and spinal involvement include headache, lethargy, seizures and radicular pain. Cranial nerve involvement has been reported in up to 95% of patients during the course of the disease(7). Loss of vision associated with meningeal carcinomatosis has been reported to occur in between 15-30% of cases(8). But blindness as the initial symptom is rare(3) and usually starts in one eye.

Our patient showed a full spectrum of manifestations characterized by headache, visual loss and radicular pain. The actual mechanism by which meningeal carcinomatosis produces blindness in not well defined(2,5,8). The implicated mechanisms can be summarized as follows: Direct infiltration of the optic nerve, tumour cuffing of the leptomeninges, compromised vascular supply, humoral toxin or retinal photoreceptor degeneration as a remote effect of cancer(3,6,9,10,13). We presume that the 2nd and 3rd mechanisms caused the visual loss in our patient. The abrupt onset of blindness and the normal appearance of the optic nerves at CT scan would seem to exclude the other possibilities.

The most important diagnostic test for leptomeningeal matastasis is examination of CSF. Repeated cytological studies, however, are frequently necessary to indentify malignant cells(5). 85% of patients were eventually found to have malignant cells in the cerebrospinal fluid. In our case meningeal carcinomatosis was confirmed on the second examination of CSF. Aside from malignant cells, increased protein content has been reported in up to 76% of cases. In recent reports increased levels of CSF ß-glucuronidase activity is throught to be a strong indicator for the presence of meningeal carcinomatosis(12,14).

Treatment of meningeal carcinomatosis must be directed toward the entire neuroaxis because of the diffuse nature of the condition(7,11,14). A combined approach with whole brain radiation and chemotherapy will produce temporary remission. It is reported that the latter appears to be most effective when the chemotherapeutic agent is injected directly into the CSF compartment(11,14). In the evaluation of acute total visual loss a physician should consider the possibility of meningeal carcinomatosis.

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