Journal of BUON **14:** 317-319, 2009

Leptomeningeal carcinomatosis presenting as bilateral sensorineural deafness and unilateral facial palsy

S. Mourgela[1], A. Sakellaropoulos[2], A. Ardavanis[3]

[1]Department of Neurosurgery, "Agios Savvas" Anticancer Institute, Athens; [2]Department of Pulmonary and Critical Care Medicine, "Neon Athineon" M.D. Hospital, Athens; [3]Department of Medical Oncology-A, "Agios Savvas" Anticancer Institute, Athens, Greece

Summary

This paper describes the case of a 56-year-old man with a history of small cell lung cancer under chemotherapy, who presented with left-sided peripheral facial palsy and progressive bilateral sensorineural deafness due to leptomeningeal carcinomatosis (LMC). Brain magnetic resonance imaging (MRI) of the petrosal bones and posterior cranial fossa revealed 2 solid lesions in the internal acoustic meatuses bilaterally and LMC of the skull base. Whole brain radiation therapy and methotrexate intrathecally were applied to the patient.

Key words: facial nerve palsy, meningeal carcinomatosis, lung cancer, sensorineural deafness

Introduction

LMC from solid tumors is a rare, clinically important neurological complication of systemic cancer [1]. Malignant cells infiltrate the subarachnoid space and cerebral pia mater causing meningeal irritation and cranial nerve symptoms. Gastric, breast and lung cancers are often causes of LMC [2,3]. Isolated bilateral deafness [4] is a rare manifestation of this entity, while facial paresis or plegia is a frequent finding [5]. Simultaneous presentation of facial palsy and sensorineural deafness is very rare.

We report herein the case of a patient with a small cell lung cancer presented with bilateral deafness and left-sided peripheral facial palsy due to LMC, in order to show the rarity of such a case and to denote that in cases of LMC, although brain MRI can be normal, MRI of skull base with thin slices and contrast material enhancement can be positive.

Case presentation

A 56-year-old man with previous history of small cell lung cancer under chemotherapy developed bilateral hearing problems and left-sided facial palsy for 2 months. Brain computed tomography (CT) and MRI revealed no abnormalities although the symptoms worsened, which were attributed to chemotherapeutic drugs toxicity.

MRI of petrosal bones and posterior cranial fossa with thin slices after contrast material injection revealed enhancement of both internal acoustic meatuses and leptomeninges (Figure 1). The solid lesions in the

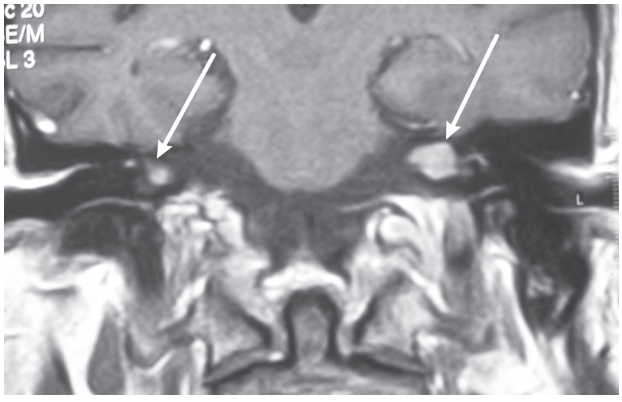

Figure 1. MRI showing lesions in internal acoustic meatuses bilaterally (arrows).

Correspondence to: Sofia Mourgela, MD. 12, Vikatou Street, 115 24 Athens, Greece. Tel / Fax: +30 210 69 25 520, E-mail: sofiamou@otenet.gr
Received 02-06-2008; Accepted 25-10-2008

internal acoustic meatuses bilaterally were in contact with the 7th and 8th cranial nerves, and measured 11 mm and 6 mm on the left and right, respectively. Cerebrospinal fluid (CSF) examination revealed increased protein and lactate dehydrogenase (LDH), decreased glucose and was cytologically positive. The optic fundi and pupils were normal. Since the diagnosis of meningeal carcinomatosis was established the patient underwent whole brain radiation therapy and intrathecal methotrexate administration via lumbar puncture.

Discussion

LMC, also known as meningeal carcinomatosis or neoplastic meningitis, represents a rare manifestation of metastatic malignancies and is an oncological emergency. Melanoma, breast and lung carcinoma are the most frequent malignancies behind LMC [6,7]. In lung carcinoma, meningeal carcinomatosis is far more common with small cell histology [8,9] and our patient had small cell lung cancer. Few reports exist of carcinomatous meningitis in non small cell lung carcinoma [10-13]. Other rare primary sites documented in literature are the uterine cervix, gallbladder and stomach [14-16].

Because treatments for primary malignant tumors have become more effective, patients live longer and, due to the lengthened survival, the possibility of LMC originated from any organ increases and becomes more common. LMC is often diagnosed after symptoms have progressed. Hence, the patient has a poor prognosis and lives no longer than 6 months. Median survival is 4-6 weeks if left untreated and 2-3 months if treated [3]. Our patient was under chemotherapy and at the same time he developed cranial nerves deficits.

The hallmark of the disease is variable and multifocal neurological symptoms and signs that involve different levels of the neuraxis appear [2,3]. The most common symptom is headache but other symptoms such as seizures and cranial nerves deficits also appear like in our patient. Patients typically have more abnormal findings than the symptoms, which is in contrast of what is typically observed in brain metastases.

The diagnosis can be verified with CSF cytology, which is highly accurate and reliable and very important to exclude meningitis [17]. False-positive findings are rare, but false-negative are common. CSF analysis can be aided by assaying the CSF for possible tumor markers [17]. Repeated CSF cytological examination may be necessary [1].

MRI with gadolinium contrast can demonstrate dura-arachnoid, pia-subarachnoid space, even subependymal enhancement, or hydrocephalus with or without associated meningeal enhancement.

Treatment options include radiotherapy and intrathecal or systemic chemotherapy. Radiotherapy is an option for symptomatic sites of disease and areas with bulky disease. Intrathecal chemotherapy options include methotrexate, thiotepa and cytarabine, but most solid tumors are relatively resistant to them. Intrathecal chemotherapy should always be administered via Omaya reservoir if possible, in order to enable repeated intraventricular administration. It is safer to administer intrathecal chemotherapy before the initiation of cranial irradiation. In our case both therapies were applied simultaneously.

Systemic administration of chemotherapeutic agents with high CSF penetration, such as high-dose methotrexate, may also provide a treatment option [8].

The destruction of the 7th and 8th cranial nerves in our case was probably due to direct infiltration by neoplastic cells as well as to ischemia through compression of the nerve-supplying vessels [5].

References

1. Wasserstrom WR, Glass JP, Posner JB. Diagnosis and treatment of leptomeningeal metastases from solid tumors: experience with 90 patients. Cancer 1982; 49: 759-772.
2. Newton HB. Neurologic complication of systemic cancer. Am Family Phys 1999; 15: 878-886.
3. Balm M, Hammak J. Leptomeningeal carcinomatosis. Arch Neurol 1996; 53: 626-632.
4. Testoni S, Pirodda A, Pastore Trossello M et al. Meningeal carcinomatosis causing isolated bilateral symmetric progressive hearing loss. Neurol Sci 2005; 25: 345-347.
5. Von Campe G, Regli F, Confavreux C et al. Bilateral deafness: an initial manifestation of meningeal carcinomatosis. Rev Neurol (Paris) 1992; 148: 133-138.
6. Strady C, Ricciarelli A, Nasca S et al. Carcinomatous meningitis and solid tumours. Oncol Rep 2000; 7: 203-207.
7. Dreyfus F, Le Chevalier T, Constans JP et al. Meningeal carcinomatosis in adults. Sem Hop 1982; 58: 19-23.
8. Janne PA, Janicek MJ, Brown F. Meningeal carcinomatosis in lung cancer. J Clin Oncol 2000; 18: 2927-2929.
9. Balducci Little DD, Khansur T, Steinberg MH. Carcinomatous meningitis in small cell lung cancer. Am J Med Sci 1984; 287: 31-33.
10. Rossi M, Morena M, Tognetti P. Carcinomatous meningitis. Presentation of 3 cases. Recenti Prog Med 1995; 86: 159-163.
11. Mateos Rodriguez F, Fuertes Martin A, Almeida Parra J et al. Carcinomatous meningitis as early manifestation of lung adenocarcinoma. An Med Interna 1993; 10: 517.
12. Csako G, Chandra P. Bronchioloalveolar carcinoma presenting with meningeal carcinomatosis. Cytological diagnosis in cerebrospinal fluid. Acta Cytol 1986; 30: 653-656.
13. Greco FA, Fer MF. Oat-cell carcinoma of the lung with carcinomatous meningitis. N Engl J Med 1978; 298: 1146.
14. Kumar S, Nair S, Alexander M. Carcinomatous meningitis occurring prior to a diagnosis of large cell neuroendocrine carcinoma of the uterine cervix. J Postgrad Med 2004; 50: 311-312.

15. Shen Y, Blumenthal DT, Digre K et al. Carcinomatous meningitis as the presenting manifestation of gallbladder carcinoma: case report and review of the literature. J Neurooncol 2004; 70: 67-71.
16. Delaunoit T, Boige V, Bello CJ et al. Gastric linitis adenocarcinoma and carcinomatous meningitis: an infrequent but aggressive association report of four cases. Ann Oncol 2001; 12: 869-871.
17. Glass P, Melamed M, Chernic KNL. Malignant cells in cerebrospinal fluid. The meaning of positive CSF cytology. Neurology 1979; 29: 1369-1375.