Case Report

MULTIPLE CONCOMITANT PRIMARY CNS TUMORS OF DIFFERENT HISTOLOGY: REPORT OF TWO CASES

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ABSTRACT

Two cases of multiple concomitant CNS tumors of different histologic types are presented. One was a case of neurofibromatosis type 2 (NF2) with 6 CNS tumors, and the other a case of 4 CNS tumors without any evidence of NF. To our knowledge, there is no report of concomitant occurrence of 6 CNS tumors in the medical literature. Neuroradiological and operative findings are illustrated and discussed.

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Key Words • Neurofibromatoses • central nervous system • meningioma •ependymoma

Introduction

Multiple concomitant primary CNS tumors are uncommon and can represent neurofibromatosis (NF), and more rarely, without neurofibromatosis.[1,2]

Herein, two patients with multiple concomitant primary CNS tumors are presented. One was a known case of NF2 with 6 concomitant tumors, while the second case harboring 4 CNS tumors, had no evidence of von Recklinghausen's disease.

Case 1:

18 months prior to admission, a 58 year-old man presented with numbness of the right hand and foot followed by numbness in the left hand associated with quadriparesis. On physical examination, left sided hearing loss, bilateral hyperreflexia associated with clonus and positive Babinski and Hoffman signs were detected. There were interosseous muscle atrophy, decreased power of the upper and lower extremities more prominent on the right side, disturbed ipsilateral cerebellar tests, and together with decreased pinprick sensation at C4 sensory level were detected.

MRI revealed a tumor with heterogenous signal intensity in the cervical spinal cord at the level of C3-C6, a left CP angle tumor, and a right frontotemporal meningioma (Fig 1).

The operative management comprised 3 stages. The first operative findings were:

1-An intradural extramedullary tumor, which was a schwannoma (tumor 1).

2-An intradural intramedullary tumor; ependymoma (tumor 2).

Total resection of schwannoma and radical resection of ependymoma were performed.

The second operative findings were: Three separate masses in the left CP angle:

1-An acoustic schwannoma (tumor 3).

2-A small neurofibroma of the fifth nerve (tumor 4).

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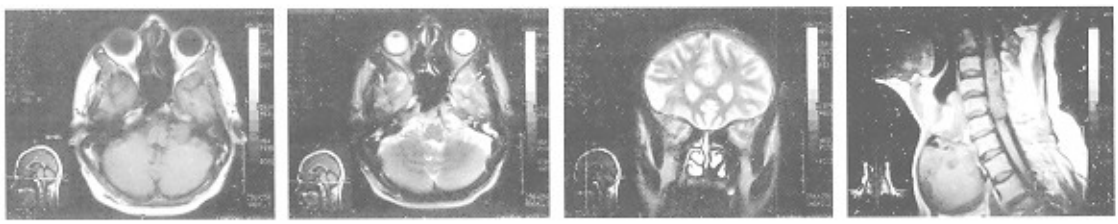

Figure 1: A Left CP angle tumor in axial T1 **(Left)** and T2 **(Middle left)** weighted MR image, and a right frontotemporal extraaxial mass **(Middle right)** in coronal T2WI, and a tumor in cervical spinal cord and a heterogenous neck mass in sagittal T1WI **(Right)**.

3- A small jugular foramen tumor (tumor 5), suggesting a tumor of IXth, or Xth or XIth cranial nerve.

Total resection of schwannoma was carried out, however, because of the presence of normal gag reflex and the normal fifth nerve function, excision of the 2 small tumors was not attempted.

Three months later, the third operation revealed a right frontotemporal meningioma.

A large heterogenous neck mass was also visualized on the cervical MRI (Fig. 1, right), however, ultrasound-guided biopsy showed degenerated cystic goiter with no malignant cells.

Case 2:

A 27-year old woman was referred with a history of headache for one year duration, which was exacerbated in the last 4 months, and associated with decreased hearing for the last 2 months.

On neurological examination, there was bilateral papilledema and right sided hearing loss. A brain CT showed, a right frontal meningioma, a right CP angle schwannoma and a small size tumor in the trigone of left lateral ventricle suggestive of either a meningioma or a choroid plexus papilloma (Fig 2). Here again the operation was performed in 3 stages.

The first operative finding was a vascular right frontal tumor attached to falx, which proved to be a meningioma.

The second operation was performed after 5 months and revealed a hard right CP angle tumor which proved to be an acoustic

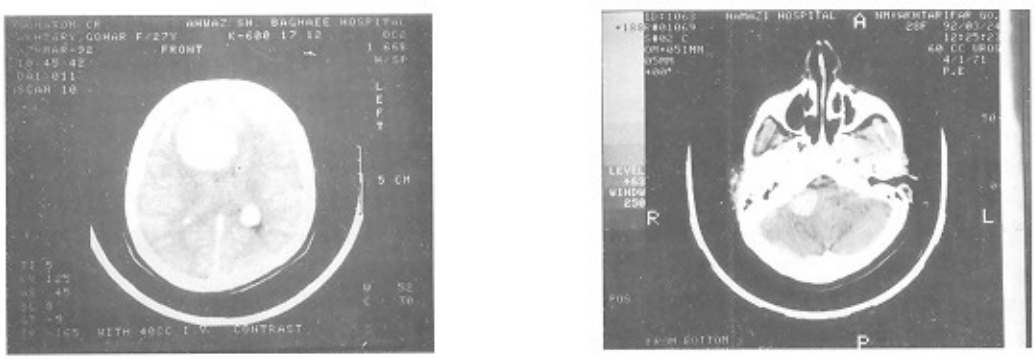

Figure 2: Axial contrast enhanced CT scan showing 3 enhancing lesions. **(Left)**: a right frontal parasagittal mass and a tumor in the trigone of the left lateral ventricle. **(Right)**: a mass in the right CP angle.

schwannoma. Three months after the second operation the patient developed gradual spastic paraplegia with 0/5 power and bilateral positive Babinski sign and a T4 sensory level. Thoracic myelography revealed an extramedullary intradural mass at the level of T4. The third operation was performed that showed a 2x4 cm calcified mass anterior to the spinal cord at the level of T4, which was resected. Pathologic examination revealed a meningioma. The small tumor in the trigone of the left lateral ventricle was not resected.

Discussion

Multiple primary CNS tumors of different histologic types are rare, accounting for only 0.4% of all CNS tumors[3]. Suggested etiologic factors for multiple tumors of different types include phacomatosis, irradiation and the presence of local or systemic tumorigenic substances.[1,3-6]

The first report of multiple primary brain tumors with histologic confirmation dates back to Shapland and Greenfield in 1935; their patient was a case of von Recklinghausen's disease with meningioma, neurofibroma and glioma.[7]

In the review of literature, multiple schwannomas,[8] multiple meningiomas,[9,10] multiple glioblastoma,[11] multiple intramedullary spinal cord tumors,[12] and combined occurrence of multiple different primary brain tumors in NF2 have been reported. Bilateral vestibular schwannomas are the hallmark of NF2.[2]

Meningioma has been present in more than 50% of cases of NF2, and also in most cases of multiple brain tumors not associated with von Recklinghausen's disease.[2,3,13,14]

Gokalp et al reported nine patients with multiple intracranial tumors, who did not have a history of irradiation or phacomatosis.[15]

Kogler et al., reported a case of concomitant bihemispheric cerebral ganglioglioma and hemangioma in an 18-month-old child.[16]

To our knowledge, only 2 cases of documented triple primary brain tumors of different histological types have been reported.[3,4]

In our second case with four CNS tumors, no NIH criterion of NF2 was present. Also other possible etiological factors for the presence of multiple tumors such as previous irradiation were not recognized.

We did not detect 3 of the tumors on the non-contrast MRI namely: the extramedullary tumor in cervical region, a left fifth nerve neurofibroma and a left jugular foramen tumor. Unfortunately, post-contrast MRI was not available in the pre-operative evaluation of this patient, therefore the inability to detect these tumors can be partly due to the absence of post-contrast MRI and also due to the small size of the tumors in left fifth nerve and jugular foramen. Therefore, in the presence of more than one primary CNS tumor especially in NF2, careful radiological and operative evaluation is crucial in order to obviate missing additional small tumors.

Kuroiwa et al[3] suggested the possibility of coincidence of multiple primary brain tumors not associated with von Recklinghausen's disease. They found that the most frequent combinations were those of meningioma and glioma, meningioma and pituitary adenoma, and that of meningioma and neurinoma. Because these tumors are quite common among the intracranial neoplasms, it may be reasonable to suppose that many of these tumors co-exist incidentally.

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