Multiple Tumours Of The Central Nervous System (CNS): Two Cases

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Abstract: Multiple tumours of the CNS are relatively rare and almost half are accompanied by von Recklinghausen's disease. In this report two cases of multiple tumours of the CNS without von Recklinghausen's disease are presented. Case 1 had five CNS tumours (two intracranial meningiomas at different sites, two spinal meningiomas, one spinal ependymoma). Case 2 had two spinal tumours with different histological differentiation (thoracic spinal meningioma (Th3–Th4) and a ganglioneuroma arising from the first sacral root (S1)). These tumours were diagnosed by myelography and computed tomography (CT), and all were removed by microsurgery. The etiology and pathogenesis of multiple tumours of the CNS are discussed in the light of pertinent literature.

Key Words: Central nervous system, Multiple tumours.

The overall incidence of multiple tumours of the CNS is low. In the absence of neurofibromatosis, the coexistence of spinal and cranial meningiomas are distinctly uncommon, i.e. less than 1% of all meningiomas (4, 11, 15). The combination of two primary neuroepithelial tumours, e.g., meningioma + ependymoma, meningioma + ganglioneuroma is an uncommon coincidence.

The first case of multiple cerebral meningiomas was described by Anfimow and Blumenau in 1889 (2). The term "multiple meningiomas" was defined by Cushing and Eisenhardt in 1938 (6).

Multiple tumours reviewed in world literature were found to have a mean frequency of 2.4%, but only a few cases of cerebral and spinal meningiomas in the same patient have been described (3, 7, 10, 17).

Case reports

Case 1:

A 15-year-old girl was admitted in December 1992 complaining of headache and weakness of the lower extremities.

Cranial CT investigation disclosed two masses at the left pontocerebellar angle (PCA) and left parietal convexity (Figures 1, 2).

Fig. 1: A mass shows characteristic homogeneous enhancement and a well-circumscribed margin at the cerebellopontine angle.
At the first operation occipital and suboccipital craniotomy was performed and both tumours were removed.

Histological examination of these two tumours was reported as psammomatous meningioma. There was no complication postoperatively. For explanation of the tetraparesis lumbar myelography was performed which showed a complete block of the contrast medium at thoracic level (Th11) and an intradural defect at lumbar 2 (Figure 3).

Th 10–11 and L2 laminectomies were performed and two independent intradural, extramedullary tumours were removed by microsurgery.

The histology of these tumours was identical with the cerebral meningiomas removed one week before.

A week after the second operation quadriplegia developed and a total block at Th2 level was disclosed by myelography (Figure 4). C5–Th2 laminectomies were performed and an intramedullary mass extending from Th2 to C5 was removed completely. Histological examination disclosed an ependymoma.

The postoperative course was uneventful and the patient was referred to a rehabilitation centre.
Case 2:

A 63-years-old woman was admitted in May 1990 with low back and right leg pain. Myelography disclosed a typical filling defect at the right sacral 1 (S1) level (Figure 5) and lumbar disc hernia was diagnosed. The patient underwent an operation with a diagnosis of preoperative lumbar disc disease. A fusiform mass observed at this level, with no associated pathology in the disc space was removed.

Histological examination showed ganglioneuroma (Figure 6). The patient subsequently developed rapid weakness and numbness in both legs. A second myelography was performed. This disclosed a complete block consistent with an intradural spinal mass at Th3 (Figure 7). At operation a large intradural tumour was removed. Morphological examination showed meningioma (Figure 8). The postoperative course was normal. One month later, the patient was discharged symptom free and has had no other complaints since.
**DISCUSSION**

Multiple tumours of the CNS are relatively rare and account for only 1.2 to 9.5% of all spinal tumours (13). Since the introduction of CT scanning, frequencies of multiple meningiomas, ranging between 5.9% and 10.5%, have been reported (2, 11, 12). Since the era of CT, the rate of diagnosis of these tumours increased substantially to 8%, 9% and 12% respectively (1, 5, 11, 15).

Several theories have been proposed to explain the aetiology and pathogenesis of multiple CNS meningiomas.

- Spontaneous or surgical blood-borne spread.
- Spontaneous or surgical spread via the cerebrospinal fluid and.
- Multicentricity of dural foci (1, 11, 14).

Spontaneous dissemination via blood or cerebrospinal fluid was unlikely in our cases as the tumours were histologically benign and, seeding after surgery was impossible, because all were observed at same time and there was a short interval between operations.

In the literature, half of the multiple CNS tumours are accompanied by von Recklinghausen’s disease and most are meningioma (7, 8). But in our cases there was no sign of von Recklinghausen’s disease.

Multiple spinal cord meningiomas are less in frequency and constitute 2 to 3.5% of all spinal meningiomas.

The frequency of multiple intracranial meningioma is 3% to 10% of all meningiomas (11, 16). Only a few cases of cerebral and spinal meningiomas coexisting in the same patient have been described and their histological type may be essentially the same or different (7, 17).

The histological type of multiple spinal tumours is usually the same (7, 8, 9). Only 22 cases of different histological types have been reported and they were associated with von Recklinghausen’s disease.

During the investigation of cranial and spinal masses, the multiplicity of CNS tumors must be kept in mind. The development of neuroradiological techniques (particularly CT and MR) has led to an increase in the frequency of multiple tumours detected simultaneously.

**REFERENCES**

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