Multiple Meningiomas in Different Neuraxial Compartments with Klippel-Feil Malformation: A Case Report

Ahmed I. Elghriany, Ahmed M. Elshanawany, Abdelhakeem A. Essa, Osama M. Ahmed

Department of neurosurgery, Faculty of Medicine, Assiut University, Assiut, Egypt

Corresponding Author: Osama M. Ahmed
oramadan1988@gmail.com

ABSTRACT

Though meningiomas are common neoplasms of the nervous system, the occurrence of multiple meningiomas in different neuraxial compartments is rather rare. We report a case of 30-year-old female who presented with spastic bilateral upper limbs weakness, and was found to have multiple homogenously enhancing tumors at the foramen magnum and lower cervical spine associated with Klippel-Feil malformation. She underwent surgery and total removal of these tumors was achieved, after which power improved in both upper limbs. Histopathology revealed psammomatous meningiomas of both lesions.

Keywords: Adult, foramen magnum, craniocervical, Klippel-Feil, meningioma, multiple, psammomatous

1. INTRODUCTION

Meningiomas are the most common primary non-glial brain tumors and comprise 13-19% of all primary intracranial neoplasms(1). Multiple cranial meningiomas are more common than multiple spinal meningiomas(2). Multiple meningiomas occurring in different neuraxial compartments are distinctly rare, with only 19 well-documented cases reported in world literature(3). The occurrence of such multiple meningiomas of spinal and cranial distribution in absence of neurofibromatosis is quite rare. Klippel–Feil malformation is a rare disease, characterized by the congenital fusion of any two of the cervical vertebrae. The syndrome occurs in a heterogeneous group of patients unified only by the presence of a congenital defect in the formation or segmentation of the cervical spine(4).

2. CASE REPORT

A 30-year-old female presented with backache, pain radiating to both upper limbs, and difficulty in handling objects. On examination, she had grade 4 power at all muscles in both upper limbs and increased tone and exaggerated reflexes. There were no multiple hypopigmented macules, neurofibromas, or other stigmata of neurofibromatosis. Magnetic resonance imaging (MRI) of the craniocervical junction and whole spine was performed, which revealed two well-defined, oval, intradural extramedullary, homogenously enhancing solid lesions at the level of foramen magnum and extending to the level of C2 and another lesion opposite C7, causing cord compression at these levels and bone.
anomalies (Samartzis classification type II) in the form of abnormal hypertrophy of the anterior arch of the atlas that is fused with the clivus, the occipital condyles are also fused with the lateral masses of the atlas, the bodies and lateral masses of C2-3 and C6-7 are also fused. The patient underwent surgery, regarding the foramen magnum lesion we did suboccipital craniectomy, removal of the posterior arch of the atlas with excision of the lesion. For the cervical lesion, we did hemilaminectomy of C6, C7 on right side and removal of intradural extramedullary lesion. Post operatively, the patient’s power improved and she was able to handle objects well after a week. A contrast enhanced MRI of the spine showed complete removal of tumors and decompression of cord. Histopathology revealed psammomatous meningioma of both lesions.

**Figure (1):** Contrast enhanced MRI spine showing the foramen magnum lesion and the cervical lesion

**Figure (2):** Contrast enhanced MRI axial cuts showing foramen magnum lesion

Figure (3): Contrast enhanced MRI axial cuts showing the cervical lesion

Figure (4): Multi slice CT scan showing the Klippel-Feil malformation
Figure (5): Post-operative contrast enhanced MRI spine sagittal view showing complete removal of both lesions

Figure (6): Post-operative contrast enhanced MRI spine coronal view showing complete removal of both lesions

3. DISCUSSION

We can define multiple meningiomas as at least two separated meningiomas occurring at the same time, or more than two meningiomas arising sequentially from two clearly distinct regions(5). Multiple spinal meningiomas are rarer than multiple cranial meningiomas. Multiple meningiomas occurring in different neuraxial compartments are rarer, with only six cases having multiple spinal and multiple cranial meningiomas(6).

The pathogenesis of multiple meningiomas can be explained in two ways, either these tumors arise independently as evidenced by the histological and cytogenetic differences between multiple tumors from the same patient, or a single transforming event occurs and the original clone of cells spreads throughout the meninges in the formation of multiple, clonally related tumors(1).

Of the intracranial meningiomas, one percent are multiple, usually in neurofibromatosis. The most common locations are Falx and parasagittal, convexity, sphenoid, and olfactory groove(7).

In the spinal canal, the preferred location of meningiomas is at the dorsal region then the cervical region, and finally, the lumbar region(2).

The clinical feature is characterized by a motor deficit, varying from a minimal paresis to paralysis, with pyramidal liberation signs, sphincter disturbances and signs of funicular or radicular impairment. The symptomatology is limited to a root or is associated with several sensitive and motor neurological signs(2).

Short neck, limited neck mobility and low posterior hair line due to the congenital fusion of two or more cervical vertebrae are composing the triad of Klippel-Feil malformation. Approximately 52% of patients are seen with this triad(8).

Klippel-Feil malformation has been classified and described by many systems, the simplest and the most accepted one is Samartzis Classification which is divided into 3 types:

- Type I: Single-level fusion
- Type II: Multiple, non-contiguous fused segments
- Type III: Multiple, contiguous fused segments(9)

Multiple neurological anomalies have been described with Klippel-Feil malformation. These include split cord malformation, syringomyelia, corpus callosum agenesis, meningocele. Association of Klippel-Feil malformation with intra cranial or spinal tumors are quite rare(10).

4. CONCLUSION

Although meningiomas are one of the most common tumors encountered in neurological practice, multiple meningiomas still remain a rare event.

Multiple meningiomas should be put in the mind of the surgeon to not be missed. Features that suggest the presence of multiple meningiomas include early age of
onset, female sex or presence of neurofibromatosis. When silent meningioma is discovered not causing the symptoms of the patient, a careful decision is made taking into consideration the presenting features, age, sex, tumor biology, associated diseases and patients expectations from surgery.

Uncommonly, Klippel-Feil malformation is associated with intracranial or spinal tumors. We report a unique case of Klippel-Feil malformation associated with foramen magnum and cervical meningiomas.

REFERENCES
