

Brain Tissue Adaptability to Slow-Growing Tumors: Case Report of Clivus Meningioma

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ABSTRACT

A 46-year-old female patient with a large slow-growing craniocervical junction tumor is presented. Her complaints began 6 months before with sensory and painful sensations, sphincter impairment, and motor events (spastic tetraparesis, more pronounced on the left extremities). Magnetic resonance of the head revealed a rounded tumor of 2.5 cm in diameter, by its characteristics corresponding to meningioma, at the level of C1 vertebra and craniocervical junction, with the base at spinal canal anterior wall, occupying most of the craniocervical junction, compressing spinal cord and medulla oblongata. Intracerebral computed tomography angiography showed spared lumen and a satisfactory image of vertebral arteries bypassing the expansive growth at the occipital foramen, confirming slow tumor growth. Antiedematous therapy led to transient improvement in extremity strength and partial recovery of neurologic deficit, which resolved completely upon neurosurgical operation and rehabilitation. This case report exemplifies brain adaptability to slowly growing expansive neoplasms, based on its volume reduction up to the moment when further adaptation is not possible anymore, i.e. breaking of the mechanism of adaptation. Because of brain adaptability, such slowly growing tumors may stay asymptomatic for a long time. Brain plasticity also includes adaptation and autoregulation of the circulation, thus ensuring stable blood flow.

Key words: *clivus, meningioma, brain tumor, adaptability, slow-growing tumors*

Introduction

Meningiomas are the most common primary non-neuroepithelial brain tumors that account for 20% of intracranial tumors, and are mostly characterized by slow growth and complete recovery after surgical extirpation *in toto*. Meningiomas are twice as common in women as in men^{1–4}. In rare cases, meningiomas can assume a locally invasive behavior or give rise to metastases; this is considered atypical or malignant form of meningioma⁵. The incidence of meningioma is 6 per 100,000 inhabitants. According to the World Health Organization, meningiomas are classified into three groups: benign (grade I), atypical (grade II), and malignant (grade III). About 90%–95% of meningiomas are histologically benign, 5%–7% atypical, and 1%–2.8% malignant⁶. Malignant meningiomas show a male predominance⁷. Genetic factors, hormones, ionizing radiation, electromagnetic waves, allergic diathesis and head trauma have

been implicated in the etiology of meningiomas. Clinical picture of meningioma is very variable and generally determined by tumor localization. Meningiomas are mostly slow-growing tumors that lead to gradual intracranial pressure (ICP) increase; therefore, the initial manifestation of meningioma usually is partial or generalized physical weakness persisting for months or years. Focal neurologic symptoms develop later, pointing to its anatomical localization.

Meningioma of the clivus is very rare⁸. The vicinity of the structures such as medulla oblongata, pons, superior segment of spinal cord, cranial nerves and vertebrobasilar basin leads to very variable symptomatology^{9,10}. Cases mimicking clinical picture of amyotrophic lateral sclerosis have been reported. Meningiomas can occur in any age group¹¹.

Case Report

A 46-year-old female patient was hospitalized at Department of Neurology, University Hospital Center Osijek for upper extremity weakness and loss of sensation, more pronounced on the left extremity, persisting for 2–3 weeks before admission. Six months before admission, the patient had trouble on starting urination, and a

month later felt dull cervical and occipital pain. About two months before admission, she felt trunk numbness, weakness of abdominal musculature, and difficult coughing. Several days before admission, she developed weakness of lower extremities, more pronounced on the left side.

On admission, signs of mild horizontal right gaze nystagmus and signs of spastic tetraparesis, more pronounced on the left extremities were present. Motor strength of the left hand as well as left elbow and left shoulder movements were reduced. Inspection revealed claw hand on the left. There were signs of sensitivity loss below C3 level, more pronounced on the left, and signs of mild urinary retention.

Magnetic resonance imaging (MRI) of the head showed a rounded tumor of 2.5 cm in diameter, at the level of C1 vertebra, i.e. at craniocervical junction, with its base at the spinal canal anterior wall, strongly compressing spinal cord and medulla oblongata. By its characteristics, the tumor corresponded to meningioma.

Computed tomography (CT) angiography revealed the previously verified tumor at the level of occipital foramen, i.e. at the level of C1 vertebra within the spinal canal, with preserved lumen and satisfactory image of vertebral arteries bypassing the expansive growth at the level of occipital foramen.

Upon admission, the patient was prescribed anti-edematous therapy (20% mannitol and corticosteroids), which led to improvement of extremity strength and partial recovery of neurologic deficit.

On day 13 of her hospital stay, the patient was transferred to Clinical Department of Neurosurgery, Zagreb University Hospital Center in Zagreb, where she under-



Fig. 1. Expansive growth at craniocervical junction with a base at spinal canal anterior wall and clivus.

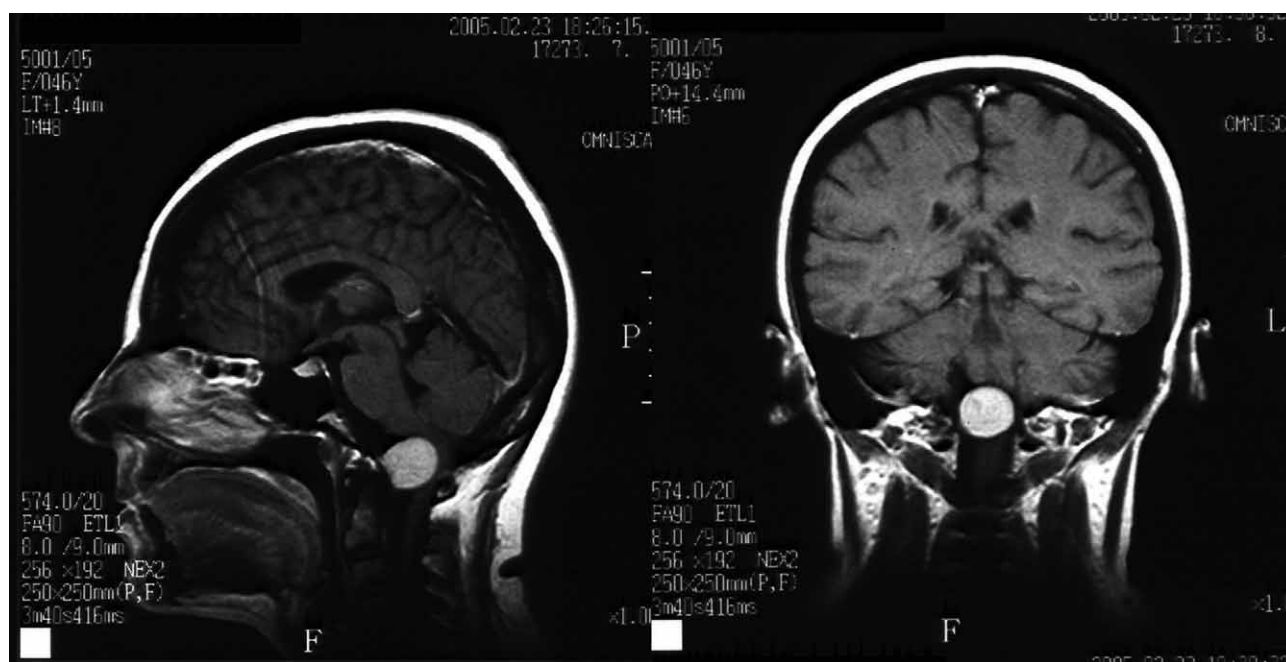


Fig. 2 and 3. Post-contrast magnetic resonance image of the expansive growth at craniocervical junction.

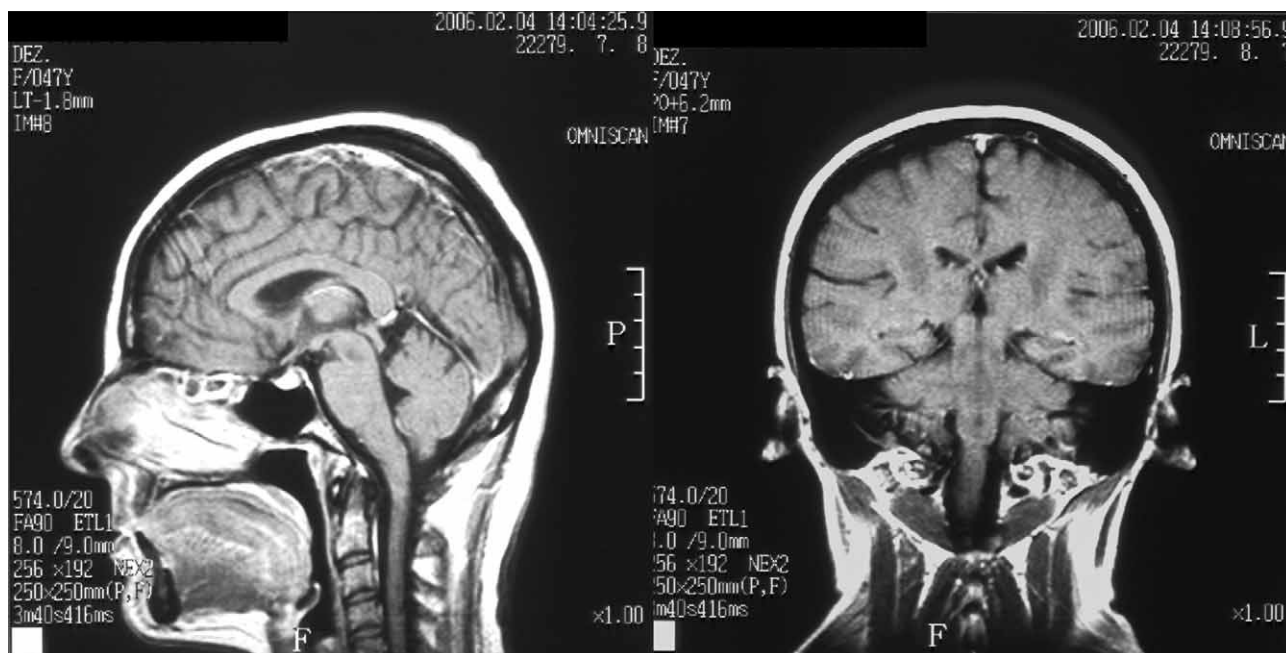


Fig. 3 and 4. Postoperative magnetic resonance image of the brain and craniocervical junction shows normal finding.

went radical surgery with tumor extirpation *in toto*, followed by 3-week rehabilitation, which resulted in complete recovery, leaving the patient free from any neurologic deficit. Postoperative MRI showed normal finding of intracranial and spinal structures.

Discussion

The clinical picture and the course of disease in our patient pointed to subcutaneous development of spastic tetraparesis and loss of all sensations inferior to C2 level, with previous development of partial urinary retention. Imaging examinations (brain MRI and CT angiography) indicated a slow-growing (long-standing) expansive growth in the area of craniocervical junction (meningioma of the clivus origin), which was supported by discrepancy between tumor size and resulting neurologic deficit. Measurement of the tumor size and surrounding structures revealed that the tumor occupied more than two-thirds of the spinal canal and great occipital foramen (craniocervical junction) dimensions. Such a large tumor size without clinically and historically visible neurologic deficits could only be achieved by very slow growth and excellent adaptation of the surrounding neural structures, which was the reason for the long absence of the spatial compressive effect and consequential neurologic deficit. CT angiography confirmed adaptation of vascular structures (vertebral artery and its branches), which bypassed the tumor mass, supporting its very slow growth, along with good adaptation of the adjacent structures. Clinical picture was the consequence of compressive effect upon descending corticospinal pathways involved in the regulation of urinary bladder and ascending sensory pathways. Nystagmus most probably developed consequen-

tially to compression of the inferior segment of medulla oblongata. Patients with meningioma of this region may show symptoms that are due to compression of the inferior segment of cranial nerves (glossopharyngeal neuralgia, geniculate neuralgia, etc.) or symptoms associated with impaired vertebrobasilar basin circulation.

The case presented exemplifies good compensation with long-standing balance of the tumor, cerebrospinal fluid (CSF), blood and cerebral tissue volumes. The increase in tumor mass was compensated for (Monro-Kellie theory) by a decrease in the volume of the other three structures (cerebral tissue, CSF and blood)^{12–15}. The Monro-Kellie hypothesis states that the cranial compartment is incompressible, and the volume inside the cranium is a fixed volume. Therefore, any increase in the volume of one of the intracranial constituents must be accompanied by a decrease in the volume of another. The main buffer mechanism implies a shift in CSF volume and to a lesser extent in blood volume. These changes occur following an increase in the volume of other intracranial structures. For example, acute occurrence of epidural hematoma results in caudal displacement of CSF and venous blood volume in order to maintain normal ICP to the volume of 100–120 mL. Then, however, an ICP increase follows, resulting in damage to intracranial structures⁵.

Brain volume includes the volume of glial cells (700–900 mL), neurons (500–700 mL) and extracellular fluid (75 mL). Glial cells and neurons make up to 70% of the intracranial content, while CSF, blood and extracellular fluid of the brain account for 10% each. While growing, acute expansive processes (e.g., acute epidural hematoma) compress and displace cerebral tissue, which can be visualized by various radiological techniques (angio-

graphy, CT, MRI), and very soon lead to cerebral parenchymal lesion and early onset of neurologic symptoms. Development of brain edema (brain volume increase due to increased amount of water in cerebral tissue) leads to rapid ICP increase, damage to cerebral tissue, and manifestation of neurologic symptoms. In case of brain tumors, vasogenic cerebral edema develops, accompanied by increased blood-brain barrier permeability (cranio-cerebral injuries, tumors, abscesses, stroke, etc.) and manifested by protein-rich fluid accumulation in the extracellular space. Further ICP increase results in breaking the mechanism of circulation autoregulation with progressive vasodilatation and vasomotor paralysis, and thus enormous ICP increase. Therefore, prompt administration of potent antiedematous therapy is mandatory.

The slow-growing processes such as tumors like meningioma also compress and displace cerebral tissue, but owing to brain tissue adaptability and plasticity the volume of cerebral tissue decreases, thus these processes may long stay asymptomatic. Brain plasticity includes adaptation and autoregulation of the circulation, thus ensuring stable blood flow.

Conclusion

The case presented shows the possibilities of brain tissue adaptation and plasticity to the slow-growing expansive process by a decrease in the volume of particular cerebral constituents up to a point when further adaptation is not possible anymore and the mechanism of adap-

tation breaks down. These slow-growing tumor processes may long stay asymptomatic owing to this brain adaptability including circulation adaptation and autoregulation to ensure stable cerebral blood flow. Complete recovery of the associated neurologic deficit is only possible with rapid diagnostic workup and therapy administered immediately upon the onset of initial neurologic symptoms.

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ADAPTACIJA MOŽDANOG TKIVA NA SPORORASTUĆE TUMORE: PRIKAZ SLUČAJA MENINGEOMA KLIVUSA

SAŽETAK

U radu smo opisali bolesnicu u dobi od 46 godina sa velikim spororastućim tumorom kranio-cervikalnog prijelaza. Tegobe počinju unazad 6 mjeseci postupno sa senzornim i bolnim senzacijama, smetnjama sfinktera, a kasnije i motoričkim ispadima (spastična tetrapareze, više izraženo na lijevim ekstremitetima). Po prijemu učinjen je MR glave koji pokaže okruglu tumorsku tvorbu u nivou C1 kralješka odnosno kranio-cervikalnog prijelaza sa bazom na prednjoj stijenci spinalnog kanala, promjera oko 2.5 cm, koja zauzima veći dio kranio-cervikalnog prijelaza sa jakim kompresivnim efektom na medulu spinalis i medulu oblongatu, a koja po svojim karakteristikama odgovara meningeomu. Intracerebralna CT angiografija pokazala je očuvan lumen i zadovoljavajući prikaz vertebralnih arterija koje obilaze oko već spomenute ekspanzivne tvorbe u visini okcipitalnog foramena što potvrđuje spori rast tumora. Na primijenjenu anti-edemsku terapiju dolazi do prolaznog poboljšanja snage ekstremiteta i djelomičnog povlačenja neurološkog deficita, a nakon neurokirurškog zahvata po provedenoj rehabilitaciji bolesnica je bez neurološkog deficita. Ovaj primjer pokazuje nam mogućnosti adaptacije (plastičnosti) moždanog tkiva na rast spororastućih ekspanzivnih procesa, temeljem smanjenja volumena istog do jednog određenog trenutka kada prestaje daljnja mogućnost adaptacije i dolazi do prekida (break) mehanizama adaptacije. Zahvaljujući ovom procesu takvi spororastući tumorski procesi dugo ostanu asimptomatski. Pri tome isto tako dolazi i do adaptacije krvotoka i autoregulacije istog čime je osiguran stabilan protok krvi.