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MultipleMeningiomas: CaseReportandReviewofLiterature

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1. Introduction

Wepresentacaseofa63-yearoldmanwithmultiplemeningiomas, withnoevidenceofneurofibromatosis2.

2. Case Presentation

A63-yearoldmanpresentedwithcomplaintsofasuddenleft-sidedweakness,propulsion,urinaryincontinence.Hehadahistoryof craniotomyforright-sidedparasagittalmeningioma18yearsago. After the surgery the patient developed left-sided spastic hemiparesis, which partially resolved in six months. After operation the patient developed also focal epileptic seizures twice a month. In May2011henoticedprogressiveworseningofhissymptomsand 20daysbeforeadmissionsuddenlydevelopedright-sidedhemiparesis, which gradually resolved.

MRI of the brain was obtained which revealed 12 meningiomas. All of them had supratentorial localization and one falcine meningioma appeared to have intratumoral hemorrhage (Figure 1, 2). MRIofthewholespinewastheobtainedwhichrevealedanother

nidusonthelevelofC4predominantelyontheleftsidewhichwas consistent with meningioma versus schwannoma (Figure 3).

On examination, the patient was awake, alert and oriented. However, he was a little depressive. He didn't have any meningeal symptomsandsigns. Cranialnerveswere WNL. He had left-sided hemiparesis. The muscle force on the right side was 4/5, on the left side 3/5. Left-sided Babinski sign. Patient had hyperostosison the right temporal region and postoperative scar on the central parietal region. The patient had no evidence of neurofibromatosis 2. In our clinic patient received dexamethasone 5 mg TID for 10 days, after that we performed the operation: left parietal craniectomy for removal of left parasagittal and falcine meningiomas. On first and seventh post-op days head CT scan was done (Figure 4, 5).

and seventh post-op days head CT scan was done (Figure 4, 5). 14 days after the operation patient was discharged from the clinicwithslightspastictetraparesis. The histological examination of the removed meningiomas showed, that all meningiomas were fibroplastic (WHO grade I).

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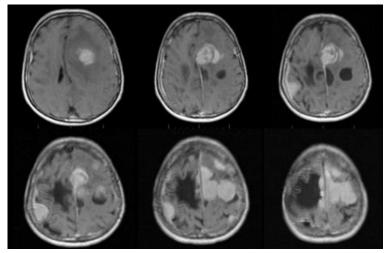


Figure1:

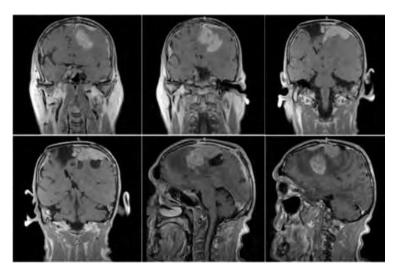


Figure2:

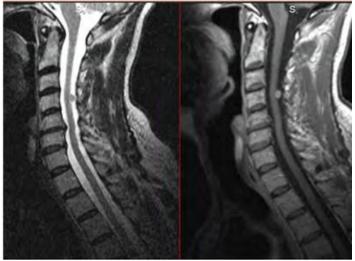


Figure3:

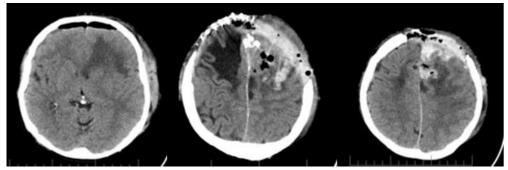


Figure4:POSTOPDay1

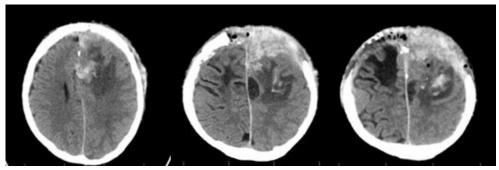


Figure5:POSTOPDAY7

3. Discussion

Meningiomasarethemostcommon,non-glial,primitiveintracranialtumors,theirprevalenceamongoperatedtumorsisaround13-19%.Theymayoccuratanyagebuthaveapeakincidencearound 45 years of age; 60% occur in females. Meningiomas may occasionally have an atypical appearance and atypical enhancement pattern secondary to necrosis, scarring, previous hemorrhage, or fat deposition [1].

Meningiomas accounts for approximately 34% of primary brain tumors (tumors that start in the brain) in the United States and occurs in approximately six of every 100,000 people. Meningiomas are rare in children. The overall five-year survival rate (the percentageofpeoplewhosurviveatleastfiveyearsafterthemeningiomaisdetected, excluding those who die from other diseases) formeningiomais69% (70% forbenignand55% formalignant). Symptomsofmeningiomacan begeneral (caused by the pressure of the tumor on the brain or spinal cord), or specific (caused by thetumorstoppingthenormalfunctioningofaspecificpartofthe brain or by pressure on nerves or blood vessels). Generally, meningiomaisnotdiagnoseduntilsymptomsbegin[2]. Theincidence multiple meningiomas has increased since the introduction of computed tomographic (CT) scan and a high incidence of up to 8.0% has been reported (2). Meningioma is classified into subtypes based on the location of the tumor:

- Falx and parasagittal meningioma (accounts for 25% of meningiomas).
- Convexity meningioma (20%).
- Sphenoid wing meningioma (20%).
- Olfactory groove meningioma (10%).

- Posterior fossa meningioma (10%).
- Suprasellar meningioma (10%).
- Spinalmeningioma(lessthan10%).
- Intraorbitalmeningioma(lessthan10%).
- Intraventricular meningioma (2%) [2].
 - Meningiomas are classified according to the World Health Organization (WHO) schema, which is based upon morphologic criteria. The 2000 and 2007 versions of the WHO classification system divides meningiomas into three groups [8]
- WHO grade I Benign meningiomas (WHO grade I) are subdivided into a number of subtypes. WHO grade I meningiomasdo notmeetanyof thecriteriafor ahigher grade lesion based upon morphologic criteria. The treatment approach is the same for all of the subtypes of benign meningiomas.
- WHO grade II WHO grade II meningiomas have increased mitotic activity (≥4 mitoses per ten high powered fields) and three or more of the following features: increased cellularity, small cells with a high nuclear cytoplasmic ratio, prominent nucleoli uninterrupted patternless or sheet-like growth, or foci of spontaneous or geographic necrosis. Chordoid, clear cell, and atypical meningiomas are classified as WHO grade II.
- WHOgradeIII-WHOgradeIIImeningiomashave
 ≥20 mitoses per ten high powered fields and/or malignant characteristics resembling carcinoma, sarcoma, or melanoma. Features that support the diagnosis of malignament.

nant meningioma include the loss of usual meningioma growth patterns, infiltration of underlying brain, abundant mitoses with atypical forms, and multifocal microscopic fociofnecrosis.Papillary,rhabdoid,andanaplasticmeningiomas are classified as WHO grade III.

ItisusedalsotheSimpsonclassification [9].

Grade I- This is a macroscopically complete removal of the tumor, with excision of its dural attachment, and of anyabnormalbone. Wherethetumorarises from the wall of a dural venous sinus, such an operation necessarily entails resection of the sinus.

Grade II -This denotes a macroscopically complete removal of the tumorand of its visible extensions, with endother my coagulation (usually to the point of charring) of its dural attachment.

Grade III -This denotes a macroscopically complete removal of the intradural tumor, without resection or coagulation of its dural attachment, or alternatively, of its extradural extensions, e.g., an invaded sinus or hyperostotic bone.

Grade IV -This denotes a partial removal, leaving intradural tumor in situ.

Grade V-This is a simple decompression, without biopsy.

Multiple meningiomas mostly consist of benign tumors, in which a combination of differential histological types of meningioma is observed in approximately 30% of cases. However, the simultaneous occurrence of benign and anaplastic histological types is extremely rare [3]. In 1938 Cushing applied the term of multiple meningiomas to a condition in which a patient has more than one meningioma with different localizations. Confluent "en plaque" meningiomas are usually reported as diffuse meningiomatosis which is generally considered an extreme form of multiple meningioma [5]. There are 2 distinctly potheses for the occurrence of multiple meningiomas. The first suggests that tumors arise independently and this is supported by histological and cytogenetic examinations that have revealed microscopic and karyotypic differences in multiple tumors from the same patient. Another hypothesis suggests that a single transforming event occurs and the original clone of cells spreads throughout the meninges resulting in the formation of multiple, clonally related tumors [3]. Primary tumorsaredeemedtobemultiplewhentheyoccurwhollyseparate from each other in different parts of the central nervous system. They can develop at the same time or independently [4]. Sometimes, despite initial treatment, the meningioma may not go into remission (the temporary or permanent disappearance of a tumor) or it recurs (comes back after treatment) [2].

Itdividesrecurrentmeningiomasinto:

- 1) Trueorlocalrecurrenceswhenthenewgrowthiseitherwithin thelimitsoftheinsertionareaofthepreviouslyresectedmeningiona or, if outside this zone, in direct continuity with (in practice, therecurrenttumorisusuallywithinthelimitsofthepreviouscraniotomy) and
- 2) False or regional recurrences, when the new growth is contiguous to but independent of the attachment surface of the primary meningioma and outside the previous craniotomy site. Those regional recurrences should be considered as new primary lesions originating from the multicentric tumor foci in the contiguous dura mater and the fibrous fringe. This pathological surrounding dura mater should be considered as part of attachment surface of "solitary" meningiomas [6]. The distinction is important because recurrence and regrowth represent two separate phenomena. The diagnosis of recurrence was based on computed tomography or magnetic resonance imaging findings [7] A number of factors have been studied for a possible relationship to the development of meningiomas and other brain tumors. They are Ionizing radiation, radiation therapy, genetic factors: neurofibromatosis type2, hormonal factors, breast cancer, head trauma, cell phone use). Multiple meningiomas can be associated with neurofibromatosis2 [5]. Neurofibromatosis type 2 (NF2) is an autosomal dominant disorder predisposing to multiple neoplastic lesion. This disorder is due to a mutation in the NF2 gene, a tumor suppressor gene on chromosome 22 which encodes a membrane cytoskeletal protein called merlin or schwannomin that appears to be involved in actin-cytoskeleton organization [8].

Treatingbrainandspinalcordtumorscanbechallenging.Surgery isthemostcommontypeoftreatment,butitcanbedifficultifthe tumor is near a delicate portion of the brain or spinal cord. The bloodbrainbarrier, which normally serves to protect the brain and cord from damaging chemicals, also keeps out many types ofchemotherapy. Meningiomagrowsoutsidetheblood-brainbarrier, so somedrugs doreachthese tumors; however, they are very resistant to currently available chemotherapy [2]. Surgery is the removal of the tumor and surrounding tissue during an operation. For meningioma, it is the most common type of treatment and is of ten the only treatment needed for benign tumors that are able tobecompletelyremovedbysurgery[2].Itcanuseradiationtherapy and chemotherapy. The goal of chemotherapy can be to destroy any tumor remaining after surgery, slow the tumor's growth, or reduce symptoms. However, chemotherapy is rarely used to treat meningioma, althoughresearchers are studying this form of treatment. It is also important to keep in mind that a treatment plan may change over time if it is no longer working. Most patients with a brain tumor will be prescribed steroids to help relieve swelling of the brain. Steroids occur naturally in the body in tiny amounts. In largeramounts, they are very powerful anti-inflammatories (drugs thathelpswelling). Youwillmostlikelyreceivesteroidswhenyou

arefirstdiagnosed,beforeandaftersurgery,beforeandafterradi- ation therapy, and if you have an advanced brain tumor. Steroids have many side effects, which include weight gain and water retention, increased appetite, difficulty sleeping, changes in mood, and stomach irritation.

Anticonvulsant medication. Aperson with a CNS tumor may experience seizures, and this type of medication helps to control the frequency of them.

Antidepressants. Depression can be common in people with a CNS tumor, but it is often undiagnosed. However, this does not mean that all people with a CNS tumor are depressed. For those who have symptoms of depression, the health care teammay decide to prescribe an anti-depressant to help with the symptoms [2].

4. Conclusions

Multiple meningiomas consist more than one meningioma with different localizations. Multiple meningiomas mostly consist of benigntumors. Therolefordevelopmentofmultiplemeningiomas playradiationtherapy, headtrauma, hormonal factors, ionization, genetic factors, especially neurofibromatosis type 2. But in our casemultiplemeningiomas were recurrent meningiomas, development who didn't play the factors who have signabove. The patient didn't have any signs of neurofibromatosis.

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