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Case Report

Intracranial Extracranial Huge Meningioma: Report of a Case

İntrakraniyal Ekstrakraniyal Dev Menenjiyom: Bir Olgu Sunumu

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ABSTRACT

Meningioma accounts for 15% to 20% of all intracranial neoplasms. Intracranial meningioma is a common brain tumor but huge meningiomas with extracranial extensions are extremely rare. Various symptoms, including neurological deficits and epileptic seizures are very frequent in these cases. Surgery still remains the principal form of the treatment and must be preceded by appropriate preoperative diagnostics. We report an unusual case of a huge meningioma of the brain (intracranial–extracranial extension) invading and destroying the skull bone and subcutaneous and cutaneous tissue. A 44–year old man in otherwise good general health was admitted to our University Neurosurgical Clinic. The MRI revealed an intracranial-extracranial lesion invading the skull bone, subcutaneous and cutaneous tissue. The tumour was removed by microsurgery. The bone was found to be predominantly lytic, and dura was infiltrated by the tumor. The bone and the dura were completely removed together with the entire tumor. Such cases are very rarely seen these days because of modern radiology techniques A giant intracranial–extracranial meningioma is therefore very unusual. Radiotherapy is generally recommended for incompletely resected meningioma and malignant meningioma.

KEYWORDS: Meningioma, Malignant meningioma, Huge intracranial-Extracranial meningioma

ÖZ

Menenjiyomlar tüm intrakraniyal neoplazmların %15-%20'sini oluşturur. İntrakraniyal olarak sık görülen bir beyin tümörüdür ama ekstrakraniyal uzantılı büyük menenjiyomlar çok nadirdir. Bu olgularda nörolojik defisit ve epileptik nöbetler dahil çeşitli belirtiler çok sıktır. Cerrahi halen temel tedavi şeklidir ve öncesinde uygun preoperatif tanı işlemleri kullanılmalıdır. Kafatası kemiğini ve subkütan ve kütanöz dokuyu istila ve imha eden büyük bir beyin menenjiyomu (intrakraniyal–ekstrakraniyal uzantı) şeklinde olağan dışı bir olgu sunuyoruz. Diğer bakımlardan sağlıklı 44 yaşında bir erkek hasta üniversitenin nöroşirürji kliniğine başvurdu. Manyetik rezonans görüntüleme (MRG), kafatası kemiğini ve subkütan ve kütanöz dokuyu istila eden bir intrakraniyal–ekstrakraniyal lezyon gösterdi. Tümör mikrocerrahi ile çıkarıldı. Kemiğin temel olarak litik olduğu ve duranın tümör tarafından istila edildiği görüldü. Kemik ve dura tüm tümörle birlikte tamamen çıkarıldı. Bu tür olgular günümüzde modern radyoloji teknikleri sayesinde çok nadir görülür, bu nedenle intrakraniyal–ekstrakraniyal menenjiyom çok olağan dışıdır. Genel olarak tam rezeksiyon yapılamayan menenjiyom ve malign menenjiyom için radyoterapi önerilir.

ANAHTAR SÖZCÜKLER: Menenjiyom, Malign menenjiyom, Dev intrakraniyal-ekstrakraniyal menenjiyom

INTRODUCTION

Meningioma accounts for 15% to 20% of all intracranial neoplasms (9). Ninety percent of meningiomas are slow growing and benign tumors, and the remaining ones are invasive or truly malignant (5). Meningiomas are usually located in the subdural space and they are the most common non glial intracranial primary tumors. Intracranial meningiomas are common brain tumors, but huge meningiomas with extracranial extensions are extremely rare. Due to the development of diagnostic tools, head swelling and deformities have decreased dramatically. Very infrequently they present as extracranial soft tissue masses. Sometimes patients with huge intracranial–extracranial meningioma may be in extremely poor condition when they are finally brought to the hospital. Various symptoms including neurological deficits and epileptic seizures are very frequent in these cases. Surgery still remains the principal form of treatment and must be preceded by appropriate preoperative diagnostics (6-8). We report an unusual case of a huge meningioma of the brain (intracranial–extracranial extension) invading and destroying skull bone, subcutaneous and cutaneous tissue. This case is the biggest in the world to the best of our knowledge.

CASE REPORT

A 44 – year old man in otherwise good general health was admitted in our University Neurosurgical Clinic because of a huge skull deformation in the bifrontal and parietotemporal left region, which was growing progressively. He was operated in other institutions three times before and the last time he underwent Radiotherapy. Initially, during the first surgery done in 2007, the tumor was not removed

completely and the HP was meningioma WHO grade I. It was uncertain as to why the tumor was not removed completely. Next surgery occurred in 2010 and then again in 2011 (for the third time) at a different clinic, at which point the HP was not disclosed. However considering the appearance of the patient, the radical (removal) of the tumor was suspected and there was no doubt that we were dealing with a malign form (malignant alteration). Following the third surgery the patient underwent Radiation. The patient came to our Center after the tumor grew to enormous dimensions, after the skin on the head developed ulcers and after experiencing difficulties walking, with vision and communication to the point where his family and friends were frightened by his appearance. Upon hospitalization he was treated with wider spectrum Antibiotics. He received a pre op MRI and other pre op routine blood work. We were unable to perform an MRI venography at our Center due to technical difficulties. The neurological status during the clinical examination showed: huge mass located at bifrontal and left parieto–temporal region. The mass was insensitive on palpation and immobile. The covering skin was ulcerating (Figure 1). The ulcerated part of the tumor became infected, and began to hemorrhage. Cranial nerve examination revealed hiposmia. We were unable to examine the right eye because the tumor mass was covering it completely. The rest of the neurological results were normal. The MRI revealed an intracranial-extracranial lesion invading the skull bone, subcutaneous and cutaneous tissue (Figure 2, 3). Four days later, the patient underwent an operation. The patient was operated through a bicoronal skin flap. The extracranial part of the tumor was seen when the skin flap was reflected (Figure 4). The tumor was then microsurgicaly removed. The bone was found predominantly lytic, dura was infiltrated by the tumor. Bone and the dura were completely removed with the entire tumor. Dura was replaced with fascia lata. Six units of blood were given during the operation. Volume estimation yielded more than 3.4 kg, making this the largest meningioma ever reported (Figure 5). Extensive defects of the dura mater and bone were well reconstructed. Cosmetic results as well as brain protection were excellent.

Because the patient felt over exhausted by his previous treatments and was very content by the cosmetic aspect (his physical appearance) as well as the functional aspect (moving freely) he refused further radiation therapy. We state this considering the nature of the tumor and of course because at this phase it was impossible to remove it any further. Due to the gigantic tumor size we were left with an abundance of healthy skin which we used to reconstruct the scalp and we therefore did not experience any skin necrosis (Figure 8). The dura mater was replaced by fascia lata, whereas one part of the bone defect was replaced by the patient's own bone and the rest by Methyl Methacrylate (Figure 7). The surgical intervention lasted 10 hours and because the tumor caused a lot of bleeding, blood was given throughout the procedure. The control CT scan showed perfect position of the implant with no CSF collection underneath (Figure 9). Histological

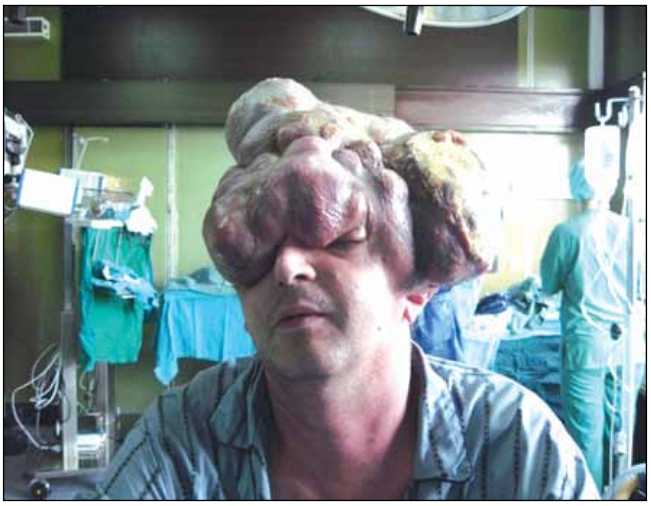

Figure 1: A/P view of patient before surgery.

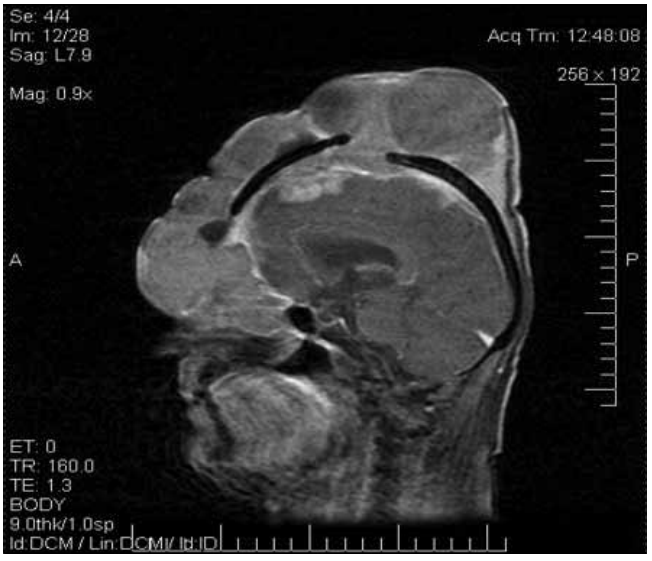

Figure 2: MRI with contrast - sagittal section.

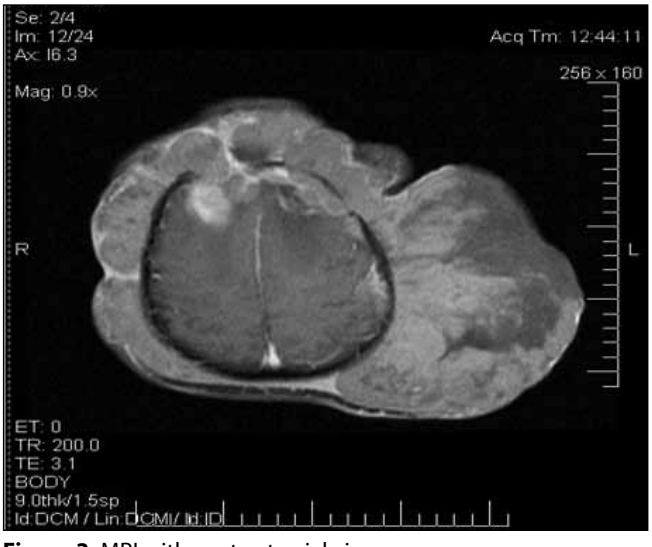

Figure 3: MRI with contrast-axial view.

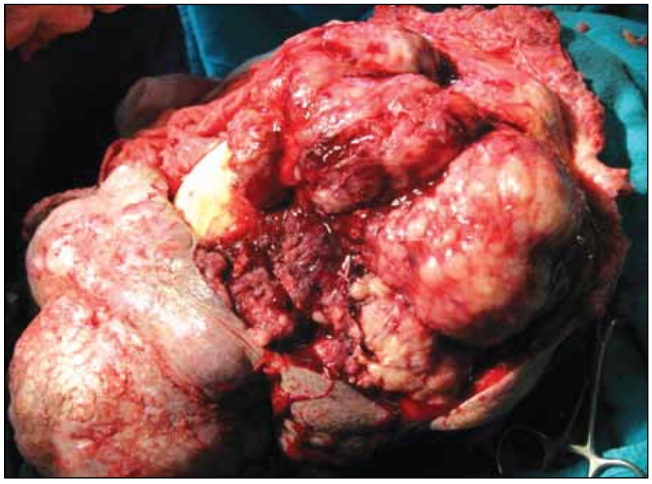

Figure 4: Intraoperative photo.

Figure 5: The amount of tumor.

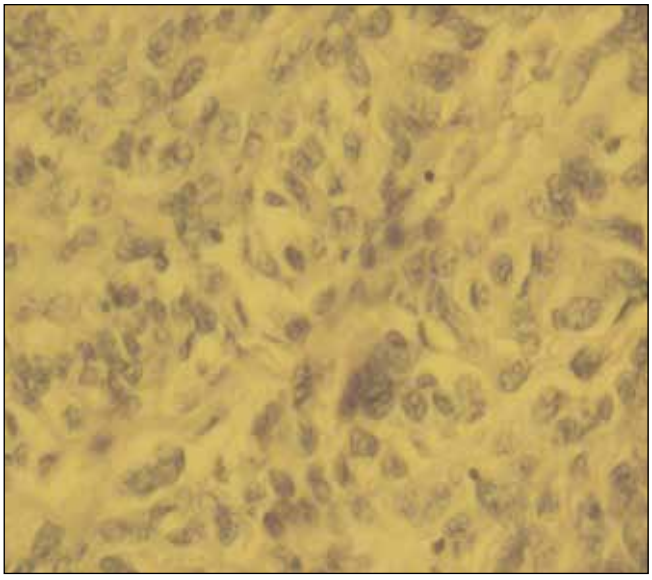

Figure 6: Loss of meningiomatous tissue pattern and numerous typical and atypical mitosis (H&E x400).

Figure 7: Bone reconstruction with Methyl Methacrylate.

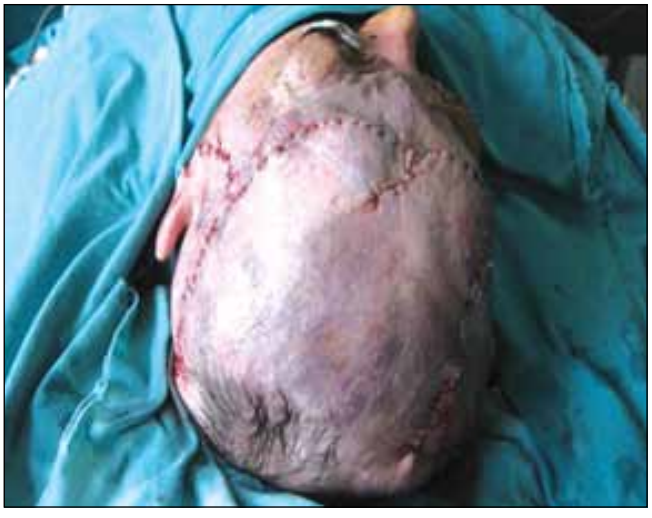

Figure 8: Skin closure without tension.

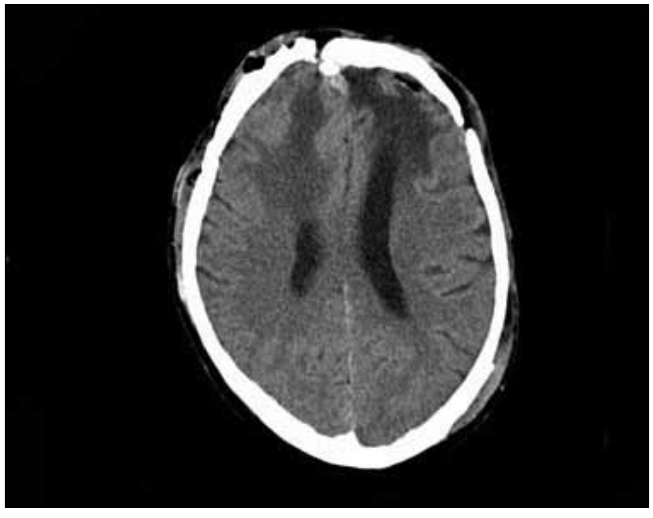

Figure 9: Postoperative cranial CT; axial section.

examination showed anaplastic malignant meningioma WHO III (Figure 6). Postoperatively, there was no change in his neurological status. Eleven days later the patient was discharged in excellent condition. Follow up after 8 months within normal expectations.

DISCUSSION

Meningiomas are common brain tumors and create up to 13% - 18% of central nervous system tumors, but large intracranial-extracranial meningiomas are very rare. Cushing and Eisenhardt addressed some cases of giant meningiomas described in the 18th and 19th centuries as well as in prehistoric times (2). These days because of modern radiology techniques, such cases are very rarely seen. Therefore, extremely giant intracranial–extracranial meningiomas are very unusual. In 1937 Davidoff removed an 835 gr. tumour including hyperosteotic bone (3) and two recent cases were described by Cech et al. and Djindjian and Raulo (1,4). In the case of Cech et al., the tumor was a parieto occipital meningioma, weighing 260 gr (1). In the case reported by Djindjian and Raulo an 800 gr. parietal meningioma was removed in a two stage operation after embolization of the feeding arteries (4). Our patient was operated on for a huge meningioma, which was especially interesting because of invasion and destruction of skull bone and massive invasion into the subcutaneous tissue. Surgical treatment of these tumors presents a surgical challenge. In the past, surface ulcerations leading to infection, massive intraoperative blood loss were very common. Our patient required 6 units of red blood cells during operation and a total of 9 units during his entire hospitalization. From previous surgery 1/3 of frontal sagittal sinus was ligated. Another surgical problem is reconstruction of the scalp if a large area of skin is involved but a myocutaneus skin flap reposition can be used (1). Due to the limited number of such cases published, definitive management guidelines do not exist. MRI is very helpful in preoperative diagnosis or if palpable extracranial/ subcutaneous soft tissue mass is present. Percutaneous needle biopsy is very helpful in establishing preoperative diagnosis. We strongly recommend surgical removal of tumor because it is very important to prevent further recurrences and malign alterations, especially when the patient is young. Radiotherapy is generally recommended for incompletely resected meningioma and malign meningioma.

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