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Research Article

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Challenging Skull Base Lesions: A Neurosurgical Perspective

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Abstract

Lesions in the skull base represent a significant diagnostic and therapeutic challenge due to the complex anatomy of the area. Their close proximity to critical neural and vascular structures makes them potentially life-threatening, and their deep location complicates both diagnosis and treatment. Most skull base lesions are benign and don't spread to other parts of the body. We present 2 challenging cases we faced first case is 62 year old immunocompetent male, radiological investigation suggestive of skull base lesion but upon further investigation it was found to be high grade intracranial glioma invading skull base and second case is of young 19 year female who was diagnosed with primary extraosseous intracranial Ewing's sarcoma, also known as a peripheral primitive neuroectodermal tumor or "small round blue cell tumor," is an extremely rare entity with only 9 cases reported in the literature till date. Early detection, careful planning, and a multidisciplinary approach are essential for effective management, with the goal of maximizing the patient's functional outcomes and quality of life. Histopathological misdiagnosis can lead to devastating consequences of late treatment and disease progression therefore close follow up and reviewing of tissue blocks may be required as sometimes key histopathological features get masked by extensive fibrosis. therefore continuous reevaluation of the pathology, especially in the case of unexpected changes in the patient's condition is crucial.

Keywords: Skull Base, Intracranial, Extraosseous, Neuroectodermal, Functional

Abbreviations

CSF Cerebrospinal Fluid
ENT Ear Nose Throat
EW Ewing Sarcoma
GTCS Generalized Tonic-clonic Seizure GTR Gross Total Resection
MRI Magnetic Resonance Imaging
MRSA Methicillin-resistant Staphylococcus Aureus pPNET Peripheral Primitive Neuroectodermal Tumor

1. Case 1

62-year-old male with no known comorbidities presented with: headache, right-sided facial pain and fullness of the cheek, fever, hearing difficulty and an episode of GTCS (Generalized tonicclonic seizure) lasting for 30 mins. MRI Brain showed ill defined heterogeneous mass lesion involving right temporal and infratemporal fossa. This combination of symptoms suggests a possible intracranial pathology, with the facial pain and hearing issues indicating possible involvement of the trigeminal nerve or a lesion affecting the brainstem or tem-poral lobe.

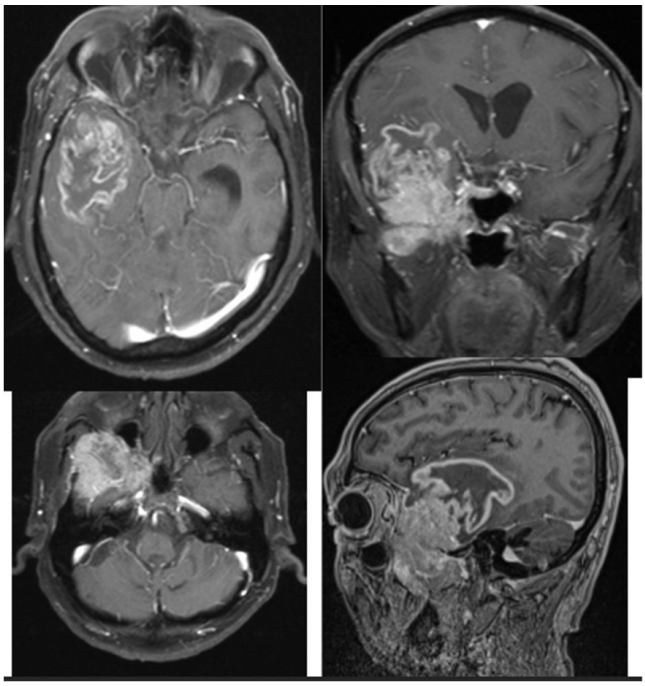

Figure 1: *MRI Brain: ill defined heterogenous mass lesion involving right temporal and infratemporal fossa.*

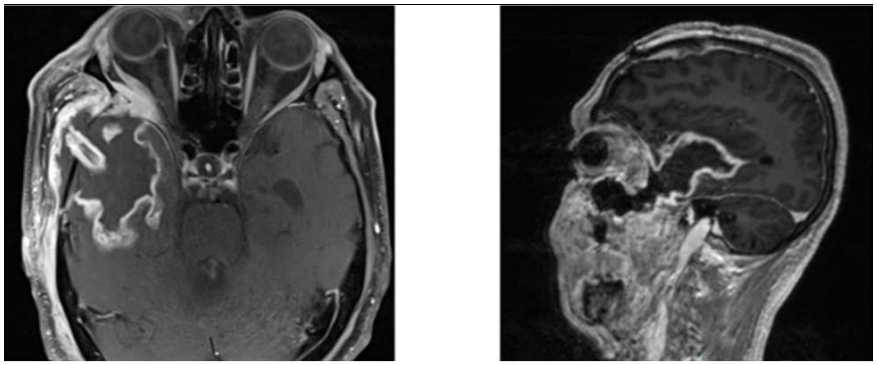

Figure 2: *MRI Brain a) axial section of brain b) sagittal section brain showing expected post operative changes.*

1.1. Surgical Intervention

The patient underwent a right fronto-temporal craniotomy, and intraoperative findings revealed dural invasion, suggesting the lesion had spread beyond the initial site. A gross total resection of the lesion was performed. Due to extensive spread of tumor involving maxillary sinus, facial structures and anterior skull base, ENT team was involved to resect a retromaxillary mass through the Weber-Ferguson approach.

1.2. Frozen Section and Cultures

The frozen section indicated a round cell tumor, which is a nonspecific finding and can be seen in various malignancies. The cultures showed the presence of MRSA (Methicillin-resistant Staphylococcus aureus), Klebsiella pneumoniae, and Candida aureus. Opinion of infection control specialist was taken on same and patient was started on appropriated antibi- otics based on sensitivity report.

1.3. Histopathology and Diagnosis

The primary histopathology report suggested supratentorial ependymoma grade 3, a type of malignant brain tumor originating from the ependymal cells lining the ventricles.

1.4. Post-Surgical Course

The patient showed symptomatic improvement after surgery, which is typical following successful resection of a primary mass and controlled infection with appropriate antibiotics. Recurrent Symptoms and Diagnosis Revision.

One week after discharge, the patient presented with fever, intermittent drowsiness, and a rash. The infection workup was negative, suggesting that the cause of the new symptoms may not be related to infection. A repeat MRI showed an increase in the size of the residual lesion, which raised concerns for a more aggressive tumor or recurrence. A review of the tissue blocks led to a revised diagnosis of glioblastoma grade IV, a much more aggressive

and fatal brain tumor, suggesting that the initial diagnosis of ependymoma was incorrect or that a secondary glioblastoma had emerged.

1.5. Management Considerations

Given the final diagnosis of glioblastoma grade IV, treat- ment typically includes chemotherapy (e.g., temozolomide) and radiation therapy. The prognosis is typically poor, with a median survival of about 12-15 months even with aggressive treatment [1,2]. The role of surgical resection in glioblastomas is limited to improving symptoms or reducing tumor mass, but complete resection is rarely possible due to the tumor's inva- sive nature [3,4].

Bony invasion in malignant gliomas have been classified into two categories. In the first type of invasion, tumors on the convexity invade the adjoining calvarium. The second type of invasion is seen in temporal lobe tumors that involve the middle fossa floor [12]. However there been reports of gliomas invading other skull base regions [1]. Macroscopically, extradural extension of gliomas has been described to occur through perivascular or dural slits; along the cranial nerves; or via direct dural destruction [2]. According to the 2007 WHO classification of central nervous system tumours, perivascular pseudorosettes are a "histological hallmark" of anaplastic ependymoma. However, glioblastomas may also feature perivascular pseudorosettes, and their distinction from anaplastic ependymoma may present a diagnostic challenge. This case highlights the challenges in diagnosing and treating ag- gressive brain tumors [3]. The progression from a diagnosis of ependymoma to glioblastoma underscores the importance of revisiting the pathology when there is a change in the clinical course. The patient's post-surgical complications, including infection and tumor recurrence, also demonstrate the complexity of managing such cases. The prognosis would now depend on the extent of the glioblastoma, the patient's overall health, and the ability to implement effective adjuvant treatments.

2. Case 2

The patient, a 19-year-old female, presented with symptoms of tinnitus, mild headache, and imbalance, which prompted an MRI revealing a left temporal lesion. A left temporal craniotomy was performed for excision, and Ewing sar-coma was diagnosed based on histopathological examination.

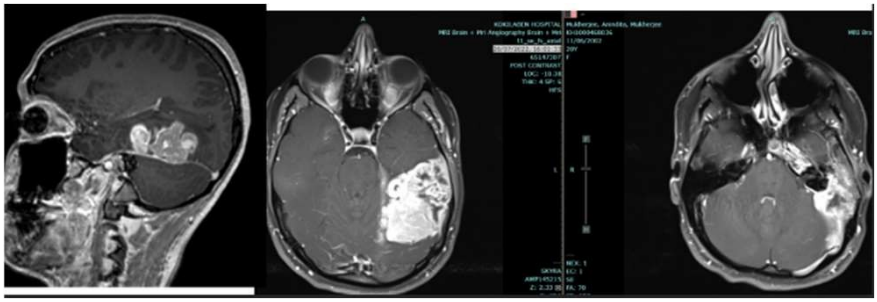

Figure 3: *MRI Brain a) sagital section b) axial section showing left temporal lesion.*

The patient was treated with ifosfamide and doxorubicin chemotherapy, followed by radiation therapy—standard treatments for Ewing sarcoma. This regimen aimed at controlling the tumor and preventing recurrence.

2.1. Recurrence of Lesion After 1 Year

After one year, the patient presented with left-sided facial palsy, which led to an MRI that suggested tumor recurrence. She underwent left retromastoid craniotomy for reexcision of the lesion. Histopathological examination confirmed Ewing sarcoma recurrence, and FISH testing showed EWSR1 gene rearrangement, a hallmark of Ewing sarcoma [11]. After surgery, she continued with chemotherapy and radiotherapy.

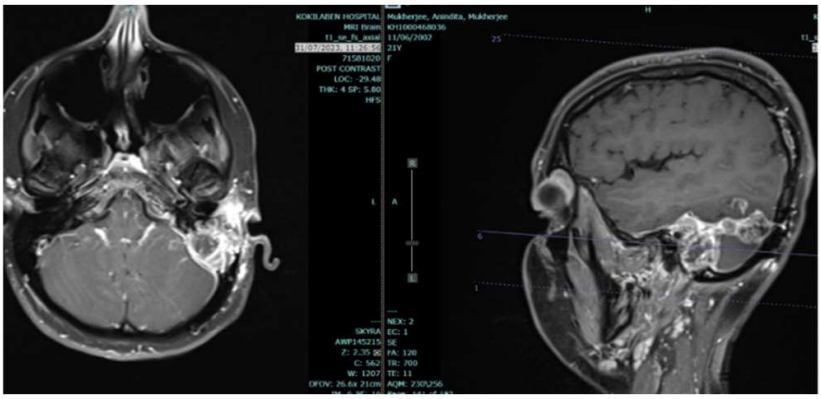

Figure 4: *After 1 year MRI a) axial section b) sagital section showing recurrent left temporal lesion.*

2.2. Complications and Worsening Symptoms

Six months later, the patient developed severe headache, worsening imbalance, and became bed-bound. Her wound became gaping, which could suggest wound healing complications or infection. MRI revealed the presence of a giant vertebral artery pseudoaneurysm at the V2-V3 junction, a serious and potentially life-threatening complication. Large pseudo-aneurysms can masquerade as mass lesions, making it challenging to identify the correct diagnosis. A pseudoaneurysm of the vertebral artery can lead to hemorrhage or compression of adjacent structures, and may necessitate urgent intervention [8,9]. The patient underwent coiling of aneurysm followed by re-suturing of wound gap. The patient also developed a CSF leak, which could be related to previous surgeries, tumor recurrence, or the effects of radiation. This, combined with her worsening neurological symptoms, led to a progressive decline in her condition, eventually requiring palliative care.

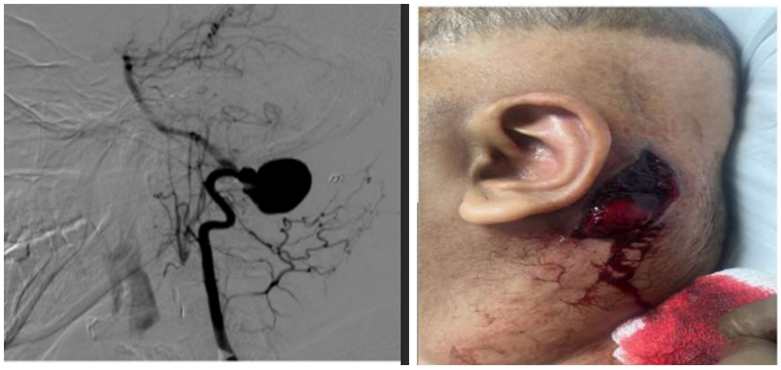

Figure 5: *a) giant verteberal artery aneurysm b) gaping of wound*

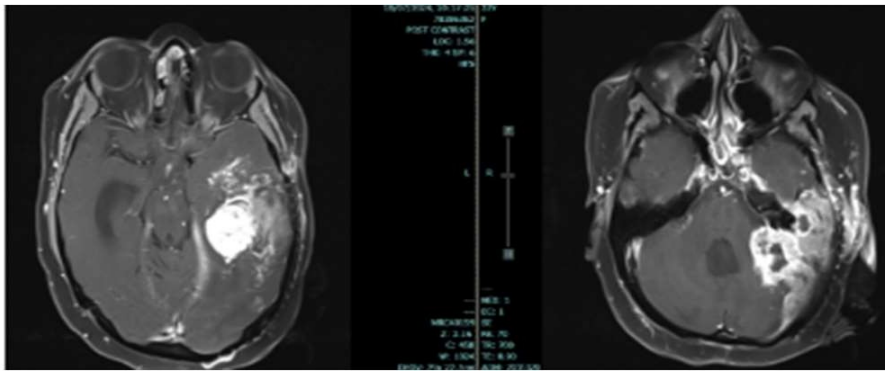

Figure 6: After 6 month MRI showing recurrent lesion.

2.3. Prognosis

Ewing sarcoma is a rare and aggressive cancer typically af- fecting bones or soft tissues but can present in atypical loca- tions, such as the brain in this case [6,7,10]. Primary intra- cranial ES/ pPNETs are aggressive malignant tumors with a poor prognosis, and ITR is one of the most important reasons for its recurrence [2]. Therefore, GTR should be the surgical aim. However, even when treated with total resection, recurrence may be unavoidable Given the recurrent nature of Ewing sarcoma and the complex complications such as the vertebral artery pseudoaneurysm and CSF leak, the prognosis remains poor. The patient's condition has necessitated a shift toward palliative care, where the primary aim is to ensure comfort, dignity, and quality of life in her final months. This case underscores the challenges in managing a rare and ag- gressive tumor like Ewing sarcoma, particularly when it involves the brain and is complicated by recurrent disease, treatment-related complications, and life-threatening vascular is- sues.

3. Conclusion

Skull base tumors are rare tumors that grow either within the cranial base or between the brain and base of the skull. Surgery is the primary treatment option of many skull base tumors. Choice of treatment depends on tumor size and location. Neurosurgical intervention for skull base lesions requires careful planning and expertise, as it involves delicate, often life-sustaining structures. Post surgery close follow up is required. Depending upon histopathological diagnosis further line of treatment can be planned. Option of re-evaluation of histopathological diagnosis should be considered in case of no response or worsening of the patient's condition. In our cases intracranial high grade glioma masquerading as skull base lesion superimposed by infection and primary intracranial extraosseous Ewing Sarcoma signifies diagnostic, treatment challenges and importance of multidisciplinary approach.

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