

Atypical presentation of the World Health Organization Grade 1 meningioma: A case report



Rajnish Kumar Jaiswal¹, Sibaji Dasgupta²

¹NSPDT, ²Assistant Professor, Department of Neurosurgery, IPGME and R and SSKM Hospital, Kolkata, West Bengal, India

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ABSTRACT

Meningiomas are the most common benign neoplasm of the brain whereas ectopic presentation, although reported, is rare. Among these ectopic tumors, there are a group of purely intraosseous meningiomas, which usually are diagnosed differentially from common primary osseous tumor such as fibrous dysplasia and osteoid osteoma. The World Health Organization (WHO) Grade 1 meningioma with an involvement of scalp, calvarial bone, dura, and brain seen rarely. This type of atypical presentation of the WHO Grade 1 meningioma rarely documented. The main theories to justify the unusual topography appear to be embryological remains of neuroectodermal tissue or cellular dedifferentiation. Surgical treatment seems the best curative option. Meningioma with bony involvement could be very aggressive tumor, need to prepare accordingly for surgery.

Key words: Atypical presentation; Bone invasion; Bone reconstruction; Brain invasion; Subgaleal involvement; World Health Organization Grade 1 meningioma

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INTRODUCTION

Meningiomas comprises 36.6% of all primary central nervous system (CNS) tumors, as reported by histology, and 53.2% of non-malignant primary CNS tumors in the USA.^{1,2} However, atypical presentation of meningioma is rare, especially in case of the WHO Grade 1 meningioma.

Meningioma has an overall incidence of 8.3/100,000 people during the period 2010–2014, which has increased over the past decade from a rate of 4.52 during the period 1998–2002.^{1,2} Meningioma incidence is quite age dependent, increasing from 0.14/100,000 in children 0–19 years to 37.75/100,000 in the 75–84 years age group.¹

Data also reveal an incidence and prevalence are directly proportional to age.⁶ Meningiomas usually affect middle aged and older adults and are twice as frequent in women as in men.

Progesterone receptors have been found in meningeal tumor cells and possibly this hormone positively influences tumor development and progression.³ Furthermore, epidemiological associations, such as a history of head trauma, cigarette smoking, and cellular phone use, have not been consistently shown as associated with significantly increased risk of meningioma, although such studies are often confounded by recall biases and frequently lack pathologic confirmation.

There are several familial syndromes that predispose meningioma development, with the most common hereditary cause being neurofibromatosis type 2, an autosomal dominant condition.⁴

The World Health Organization (WHO) has classified the neoplasm into three grades with each grade having several histological variants.

1. Grade I: Mitotic count of <4/10 high-power field (HPF)

Address for Correspondence:

Rajnish Kumar Jaiswal, NSPDT, Department of Neurosurgery, IPGME and R and SSKM Hospital, Kolkata, West Bengal, India.

Mobile: +91-8975891129. **E-mail:** rajnish88rj@gmail.com

- Absence of brain invasion
 - Nine histological subtypes: Meningothelial, fibrous, transitional, psammomatous, microcystic, angiomatous, secretory, lymphoplasmacyte rich, and metaplastic.
2. Grade II (atypical): Mitotic count of 4–19/HPF
 - Or presence of brain invasion
 - Or 3 of 5 specific histological features: Spontaneous necrosis, sheeting, prominent nucleoli, high cellularity, and small cells
 - Three histological subtypes: Atypical, clear cells, and chordoid.
 3. Grade III (anaplastic): Mitotic count of 20 or more per 10 HPF
 - Or specific histologies: Rhabdoid or papillary meningioma.

CASE PRESENTATION

A 37-year-old lady Rupali Shaw presented to Bangur Institute of Neurosciences (BIN) Neurosurgery Outpatient Department at IPGME and R and SSKM Hospital, Kolkata, with approximately 2-month history of headache, gradually progressive swelling over the right side of head. There is a history of vomiting one episode. The neurological status during the clinical examination was normal. Locally, a skull tumor of 6 cm in diameter was felt. It was immobile, hard, and insensitive on palpation and the skin over the swelling was normal (Figure 1).

A computed tomography (CT) scan of the head showed expansile lesion of right parietal bone with extra-axial iso- to hyperdense space-occupying lesion (SOL) on the right parietal region (Figure 2a). Magnetic resonance imaging (MRI) showed a large expansile heterogeneous lesion centered in dipole of the right parietal calvarium in T2 WI. It showed isointense to hypointense signal in pre-contrast



Figure 1: Preoperative photo of head of the patient showing swelling at the right parietal region

T1 WI and intense but heterogeneous enhancement in post-contrast T1 WI (Figure 2b). Associated destruction of the inner and outer tables noted with subgaleal and intracranial extension, dural invasion, and cerebral compression. There is mass effect on ipsilateral middle cerebral artery and displacement of both anterior cerebral arteries due to midline shift. Short and intermediate time of echo magnetic resonance spectroscopy of the lesion reveal diminished N-acetyl aspartate with moderate to marked elevation of choline and choline/creatinine ratio, associated with marked elevation of lactate.

The patient was operated in BIN OT after pre-anesthetic checkup with all routine laboratory investigation with coagulation profile was normal. The right frontotemporoparietal craniectomy with SIMPSON Grade 1 excision of tumor done. Intraoperatively, there was diffused subgaleal involvement, there was severe bleeding from galea and outer table of bone while raising flap.

There was bony hyperostosis with multiple lytic regions in craniotomy flap. There was extra-axial SOL with an involvement of dura with extradural and intradural component noted with encroachment of brain parenchyma. Gross total resection with involve dura and bone done, SIMPSON Grade 1. Bony defect was covered with titanium mesh to reconstruct calvaria. Closure done after putting drain in layers (Figure 3).

During intraoperative period due to severe blood loss, there was drop in blood pressure so 3 unit blood transfusion given intraoperatively and 1 unit blood transfused in post-operative period. The patient shifted to neurointensive care unit for post-operative care.

HPE report showed neoplastic lesion composed of plump meningoepithelial cells arranged in sheets, whorls, and lobules. There is also the presence of few psammoma bodies and hyalinization, suggestive of meningothelial meningioma (WHO Grade-1).

DISCUSSION

Our patient was operated on for the WHO Grade 1 meningioma, which was especially interesting because of invasion into the skull bone and its destruction along with invasion to subgaleal soft tissue and brain parenchyma, as well as finding of intraoperative severe bleeding and post-operative requirement of reconstruction.

The majority of meningioma is benign tumors that behave as expansive lesions.⁵⁻⁷ Symptoms usually arise due to compression of the brain and erosion of neighboring tissue.

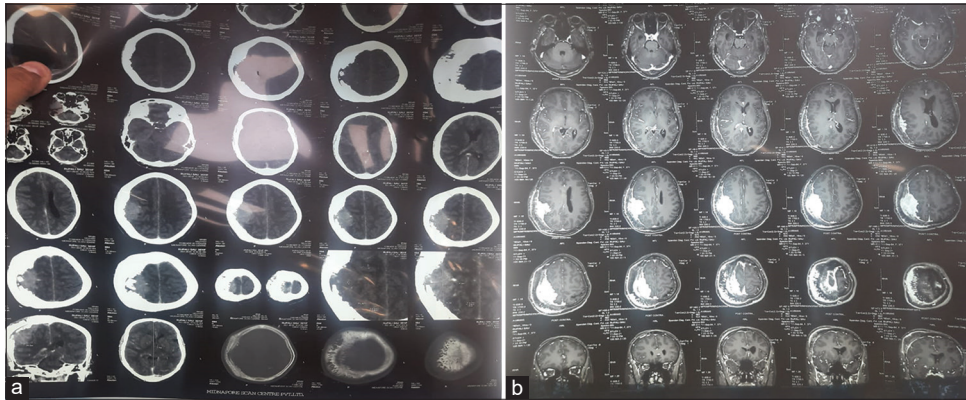


Figure 2: (a) Non-contrast computed tomography head, (b) magnetic resonance imaging brain (T1 contrast) of patient

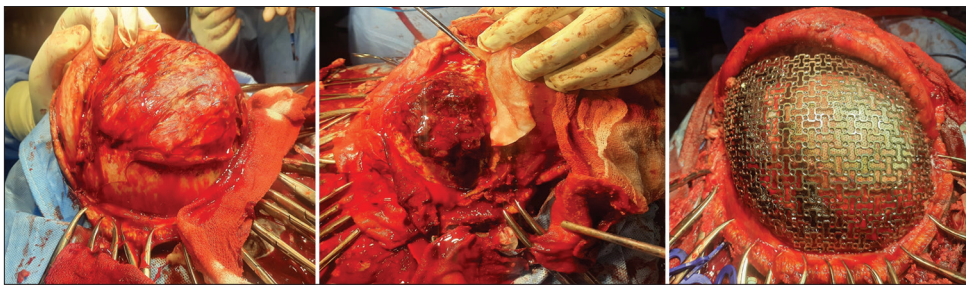


Figure 3: Intraoperative photo of the patient

Some of meningioma are invasive and about 5% of meningiomas are malignant, more likely causing direct invasion of surrounding structure.^{5,6,8-10} Beside invasive and malignant meningiomas, benign meningioma may also invade bone. CT scan and MRI brain if showing bony erosion and subgaleal involvement, we should predict severe blood loss and prepare accordingly and prognosis to be explained accordingly. In all cases, the reconstruction of removed bone is necessary. Because of tissue deficit and extensive operation, the reconstruction of the missing tissue, especially the skull bone and soft tissue, is problematic.¹¹⁻¹³ There are many alternatives to repair the missing tissue, nowadays, three main techniques are used: Autografts, allografts, and artificial replacement material.^{7,14}

To accomplish a complete resection, a combined intra- and extracranial resection is required, involving the removal of the hypertrophic bone. It was suggested that strict adherence to oncological principles should be applied also in the case of benign neoplasms to prevent contamination of wounds with tumor cells and potential recurrence.¹⁵ Often, a radical resection may be attained with low morbidity in operated patients, providing a significantly better long-term clinical outcome.¹⁰ In such extensive resections, the esthetic reconstruction of large bone defects may pose a significant issue during the operation. Viable tissue in the form of autografts and allografts is one attractive option, another one is artificial replacement material.¹⁴⁻¹⁶

In this particular case, we decided for titanium mesh cranioplasty, as it is easily available at our institute and quick procedure.

CONCLUSION

This case illustrates atypical presentation of benign meningioma and also makes us aware how much difficulty we can face in these kind of cases intraoperatively and predict blood loss. This case also illustrates intraoperative reconstruction of the skull bone is sometimes needed after the benign meningioma excision. Titanium mesh may be suitable, allowing fast intraoperative reconstruction with excellent brain protection and cosmetic effect during the one-stage procedure.

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Authors' Contributions:

RKJ- Manuscript preparation, revision of manuscript, concept and design, and resident in charge; **SD**- Review of manuscript and treating neurosurgeon

Work attributed to:

IPGME and R and SSKM Hospital, Annex1 BIN, Kolkata - 700 020, West Bengal, India

Orcid ID:

Dr. Rajnish Kumar Jaiswal -  <https://orcid.org/0000-0003-3192-7071>

Dr. Sibaji Dasgupta -  <https://orcid.org/0000-0002-2275-3519>

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