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Review Article

Transitional meningioma benign who grade 1 tumor-A case and review of the literature

Sunil V Jagtap[1,*], **Loknath Srimanto Sarkar**[1], **Shubham S Jagtap**[2]

[1]*Dept. of Pathology, KVV, Krishna Institute of Medical Sciences Deemed University, Karad, Maharashtra, India*
[2]*Dept. of Medicine, KVV, Krishna Institute of Medical Sciences Deemed University, Karad, Maharashtra, India*

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ABSTRACT

Background: Transitional meningioma (TM) is an uncommon benign meningioma. Transitional meningioma is a WHO grade I meningioma characterized by the coexistence of meningothelial cells and fibrous architectural patterns.

Case Report: A35 year female presented with history of intermittent fever, headache of 1 month duration. And since last 2 days with altered level of consciousness and vomiting.

Observations: MRI of the brain was performed using TI weighted spin echoI2 weighted turbo spin echo and turbo Fluid-attenuated inversion recovery: FLAIR diffusion sequences. It showed a large, lobulated, enhancing lesion in right fronto-temporal region, along right greater wing of sphenoid suggestive of meningioma. Surgical excision of tumor was done. On histopathology showed a benign tumor consists of meningothelial and fibrous proliferation. The meningothelial cells were arranged in fascicles, with the fibroblastic tumor in syncytial pattern. The nuclei were round, uniform without nucleoli. The chromatin was fine and open. The tumor cells were spindle having abundant pink cytoplasm, indistinguishable cell membranes and bland nuclear features. Mitotic figures and atypia were absent. Scattered Psammoma bodies were noted. Histopathological diagnosis was gives as benign WHO grade 1 tumor- transitional meningioma.

Conclusion: Due to enhanced and improved imaging, histopathological techniques the increased incidence of meningiomas is observed and it will be helpful for better outcomes of patients. We are presenting this case of transitional meningioma benign tumor WHO grade1for its clinical, radioimaging and histopatholgical findings.

1. Introduction

Meningioma accounts for 15% to 20% of all intracranial neoplasms. According to the latest Central Brain Tumor Registry of the United States (CBTRUS) meningioma was the most frequently reported histologic type, accounting for 37.1% of all CNS tumors.[1] Transitional meningioma(TM) is an uncommon benign meningioma of WHO grade 1.[2] It consists of meningothelial and fibrous patterns, are also known as mixed meningiomas. They also shows areas transitioning between the two patterns. Whorl formation and psammoma bodies are frequently noted in this variant.

In the routine clinical practice preoperative discrimination is challenging between the solitary fibrous tumor of CNS and transitional meningioma as they have similar clinical manifestations and conventional imaging characteristics. The severity of a meningioma is determined by its type, grade and location.

Due to enhanced and improved imaging, histopathological techniques the increased incidence

* *Corresponding author*.
E-mail address: drsvjagtap@gmail.com (S. V. Jagtap).

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of meningiomas is observed and it will be helpful for better outcomes of patients. The European Association of Neuro-Oncology (EANO) updated its recommendations for the diagnosis and treatment of meningiomas.[2]

2. Case Presentation

A35 year female presented with history of intermittent fever, headache of 1 month duration and vomiting 2days. Also history of intermittent altered level of consciousness.MRI of the brain was performed on MR scanner, using TI weighted spin echo, I2 weighted turbo spin echo & turbo FLAIR, Diffusion sequences. Post Contrast TIW images acquired. The observations were altered signal intensity, lobulated lesion measuring 3.3 × 3.8 x 4.3 cm seen in right temporal and frontal lobe region. There was mild to moderate perlesional edema noted. The involved sulci appears effaced. It was seen compressing right lateral ventricle and subfalcine herniation, midline shift to left side by 1.2 cm. The mild compression of basal cisterns and unchal herniation noted. It appears isointense on T2W and T1 W and FLAIR images and showed near homogeneous strong enhancement on post contrast images. The lesion was having broad base towards right greater wing of sphenoid. The lesion seen indenting MI and M2 segment of right middle cerebral artery, few of right middle cerebral artery branches seen coursing through lesion.

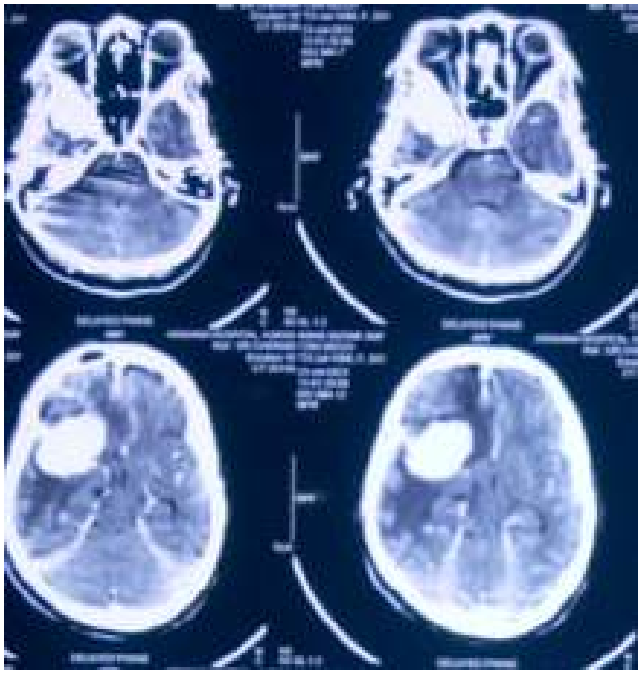

Fig. 1: MRI of the brain large, lobulated, enhancing lesion in right fronto-temporal region, along right greater wing of sphenoid.

MR spectroscopy reveals Choline peak with reduced NAA peak. There was mild confluent FLAIR hyper intensities seen in the bilateral fronto-parietal periventricular white matter, not showing enhancement on post contrast images and not showing restriction on DWI suggestive of ischemic changes. Rest of the cerebral cortical sulei, basal cisterns and ventricular system appear normal. The cerebellar foliae appear normal. There is no other abnormal focal area of altered signal intensity in the cerebral hemispheres, brain stem or cerebellum. Appearance and intensity of the brain parenchyma is normal. No evidence of obvious vascular anomaly is detected. Cerebellar hemispheres and brainstem show no

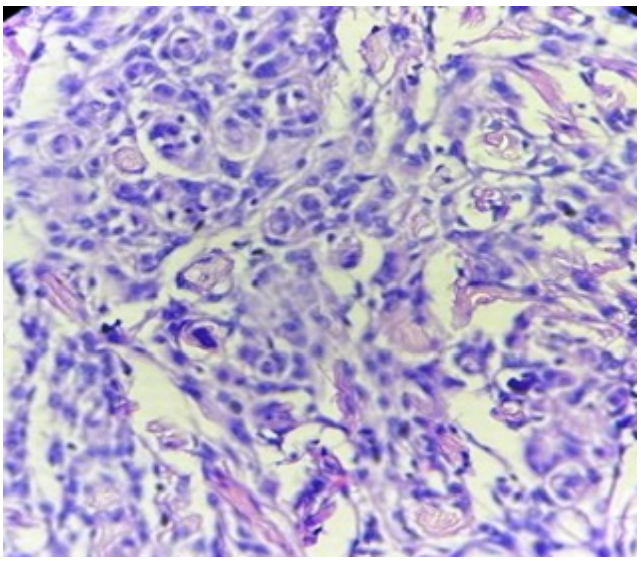

Fig. 2: Photomicrograph show a benign tumor consists of meningothelial and fibrous proliferation. (H&E stain, 100x)

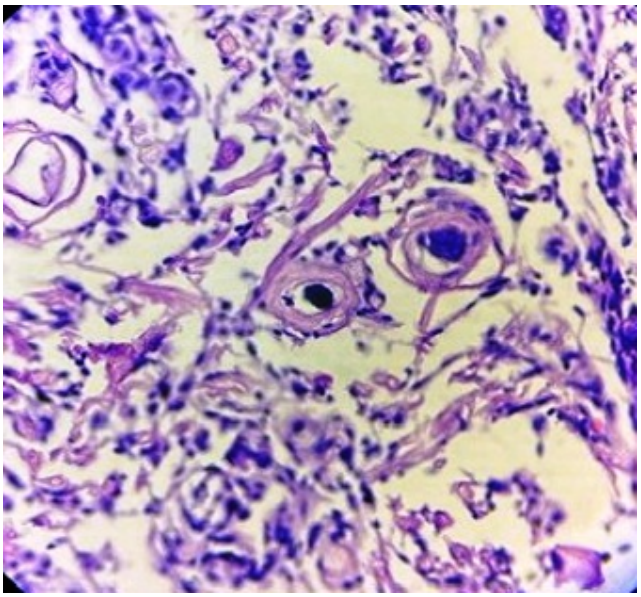

Fig. 3: Showed a benign tumor consists of meningothelial and fibrous proliferation with scattered Psammoma bodies (H&E stain,100x)

significant abnormality. No intra or extra-axial collection or hemorrhage was noted. The 7^{th} & 8th nerve complex was normal on both sides, Bilateral internal auditory canal was normal. Major intracranial structures showed expected flow voids. The paranasal sinuses and orbits appeared normal. The impression was large lobulated enhancing lesion in right fronto-temporal region, along right greater wing of sphenoid suggestive of meningioma (Figure 1) The histopathological correlation was advised.

On histopathology showed a benign tumor consists of meningothelial and fibrous proliferation. The meningothelial cells were arranged in fascicles, and the fibroblastic tumor in syncytial pattern (Figure 2). The nuclei were round, uniform without nucleoli. The chromatin was fine and open. The tumor cells were spindle having abundant pink cytoplasm, indistinguishable cell membranes and bland nuclear features. Mitotic figures and atypia were absent. Scattered Psammoma bodies were noted (Figure 3). The delicate vasculature was noted. There was no evidence of necrosis, brain invasion, or increased cellularity. Histopathological diagnosis was given as benign grade 1 tumor- transitional meningioma. We are presenting this case for its clinical, radioimaging and histopatholgical findings.

3. Discussion and Review of Meningiomas

3.1. Epidemiology

Harvey Cushing an American neurosurgeon in 1922 first to use the term "meningioma".[3]Meningiomas are neoplasms arises from arachnoidal cap cells from the meningeal coverings of the brain or spinal cord. More than 80% of meningiomas originate from the cerebral meninges, whereas less than 5% are noted extracranial in the spinal meninges. Meningiomas are most common benign intracranial tumors and accounting for 36% all CNS tumors.[4] It comprises about one fourth of all primary tumors of the central nervous system. Most of these tumors fall into WHO grade I, with a low risk of recurrence and less aggressive behavior. The mean age of among all cases of meningioma is 51.45 years, and that of transitional meningioma is 50.54 year and incidence is 4.56% of all meningiomas.[5] The female:male ratio is 2:1. In female occurrence to develop meningiomas is twice. The etiologic role for hormones is likely to develope these tumor as noticed the presence of estrogen, progesterone, and androgen receptors on some meningiomas.[6]

3.2. Clinical presentations meningiomas

The presentation of meningiomas are often non-specific. They may produce neurological symptoms as focal neurological deficit due to tumor compression to adjacent structures. Meningiomas are generally benign, slow growing tumor along external surface of brain, spinal cord or rarely, within the ventricular system. The common clinical presentation are headaches, deziness, seizures.Lateral sphenoid wing meningiomas often present with painless unilateral exophthalmos, followed by unilateral loss of vision . Parasagittal meningiomas can grow to considerable size before presenting with symptoms . The clinoidal location of meningiomas often present with a visual impairment, cranial nerve palsies, and exophthalmos.[7]

3.3. Natural history meningiomas

Meningiomas are the most common benign intracranial tumor. About 90% of meningiomas are located intracranially, and remaining 10% are found in the spinal meninges.[8] The various locations are intradural, intraventricular,cerebral convexity, parasagittal/falx,intraorbital, cerebellopontine angle,optic nerve meningioma. Also rare forms are spinal meningioma, ectopic meningioma or primary intraosseous meningioma.

The majority of intracranial meningiomas are located anteriorly, usually in close vicinity to the superior longitudinal sinus, and present as a dura-based mass covering the cerebral hemisphere or attached to the falx cerebri. Grade I meningiomas are more likely to be found at the skull base. Meningiomas are generally slow growing lesions with a linear growth rate of 2–4 mm/year for asymptomatic meningiomas.Rarely multiple meningiomas may be seen in 1% and 4% patients.[9]

3.4. Etiology meningiomas

A primary modifiable risk factor for the development of meningioma is exposure to ionizing radiation, resulting in a 6- 10 fold increase in risk.[10] Inherited susceptibility to meningioma is related to genetic and family history associated. Meningioma occurrence associated with hereditary syndromes, such as Neurofibromatosis type 2 (NF-2), Gorlin syndrome ,Cowden syndrome. The familial syndromes associated with meningiomas include Rubinstein-Taybi syndrome, Li-Fraumeni syndrome, Gardner syndrome, multiple endocrine neoplasia type 1. Deletion and inactivation of NF2 on chromosome 22 is a predominant feature meningiomas, Also genomic regions which are recurrently lost in meningiomas include 14q, 1p, 6q, and 18q.[11] In the United States, reported rates for Black Non-Hispanics are slightly higher of meningioma than other ethnic groups. With the advances in brain tumor research by new genetic and molecular knowledge the causes of meningioma are significantly updated.

3.5. Diagnostics approach meningiomas

Meningiomas diagnosis is based on history, physical examination, and radiological investigations and histopathological examination of biopsy tissue. Today

with better radio imaging facilities the rate of diagnosis of meningiomas has considerably increased**.** Also number of incidentally detected asymptomatic meningiomas are increased with recent widespread use of intracranial imaging. The investigations include contrast-enhanced CT and MRI, and angiography.[12] Meningiomas characteristically demonstrate hyperdensity on non-contrast CT, iso- to hypointensity on T1-weighted, and iso- to hyperintensity on T2-weighted images. The imaging shows uniformly contrast enhancing extra-axial mass with dural tail. Peritumoral brain edema is noted. On MRI typically appear as homogeneous, well-circumscribed, extra-axial masses with a broad dural base and is the investigation of choice for the diagnosis and characterization of meningiomas. MR spectroscopy has no significant role in diagnosis but can help in distinguishing meningiomas from mimics. The other unusual imaging features are hyperostosis in the underlying bones, and meningeal cysts, ring enhancement, calcification and various metaplastic changes are also observed.[13]

3.6. Histopathological findings meningiomas

On gross meningioma is a single, firm, a homogeneous, hemispheric or circumscribed mass. It is widely attached to the dura mater. And the tumor separates readily from brain tissue. Meningiomas usually form sessile, extra-axial mass lesions with broad-based dural attachment.

The diagnosis is based on microscopy of routinely stained haematoxylin-eosin sections with criteria given by WHO classification of tumors of the central nervous system. This classification scheme provides guidelines for tumor grading and subtypes. The present World Health Organization (WHO) grading of these tumors is based almost entirely on histopathological features.[14]

Meningiomas are classified into various grades according to their risk of recurrence and distant metastasis. The meningiomas are classified into 3 grades and 15 different variants by the World Health Organization (WHO) 2016 classification as grade 1, common meningioma with benign behavior; grade 2, intermediate or atypical meningioma; and grade 3, anaplastic meningioma or malignant meningioma. Meningothelial, fibroblastic and transitional meningiomas are the most common subtypes of meningioma in the WHO classification. Less common subtypes are metaplastic, lipomatous, osseous, cartilaginous, myxoid, xanthomatous, psammomatous, secretory, angiomatous, lymphoplasmacyte-rich and microcystic meningiomas.[14,15]

The study by Wang DJ et al, among 7,084 cases, there were 323 cases of transitional meningioma, accounting for 4.56% of all meningiomas.[16]

On histopathology transitional meningiomas showed a benign tumor consists of meningothelial and fibrous proliferation. Psammoma bodies are frequently found in transitional meningioma. In our case few Psammoma bodies were noted.

The differential are schwannoma, solitary fibrous tumor, hemangiopericytoma, dural metastases (e.g. breast cancer, oesophagael cancer).[17]

3.7. Immunohistochemistry, molecular, cytogenetic studies in meningioma

Meningioma cells exhibit a striking similarity to arachnoid cap cells, which are the likely tumor cell of origin.

Immunohistochemical staining for Somatostatin receptor 2a (SSTR2a) is a specific meningioma marker in CNS tumors. Other shows EMA, progesterone receptor positive and negative for Glial fibrillary acidic protein (GFAP) . Nassiri F et al observed that the aggressiveness of the tumor and recurrence rate can be predicted by methylation status of DNA within meningiomas as noted in recently studies.[18] Platelet-derived growth factor and its receptors (PDGFR) are frequently co-expressed in meningiomas.

The molecular cytogenetic can be detected through genetic karyotyping studies. Commonly noted abnormalities are the mutations in chromosome 22 for neurofibromatosis 2. In some cases of meningiomas abnormalities noted in chromosomes 1p, 6q, 14q, and 18q. In the literature various genetic alterations reported include SUFU, AKT1, PIK3CA, SMO, TRAF7, KLF4, PRKAR1A, and POLR2A.[19]

3.8. Treatment meningiomas

The various treatment modalities are, in cases of asymptomatic and small meningiomas regular observation are recommended. The clinical observation and MRI screening are performed every six months following initial diagnosis.[20]Gross total resection is usually curative. Gamma knife radiosurgery might be an effective therapy for patients with tumor recurrence.[21] In cases who are not fit for surgery like elderly or disabled individuals, have the option to choose either stereotactic radiotherapy or radiosurgery or chemotherapy as a primary treatment.[22]

For transitional meningioma gross total resection during the first surgery should be done. Ma XJ et al noted that Gamma knife radiosurgery might be an effective therapy for patients with tumor recurrence.[23] Perry A et al,reported recurrence rates of grade I, II, and III meningiomas are 7-25%, 29-52%, 50-94%, respectively.[24]

4. Conclusion

Due to enhanced and improved imaging, histopathological techniques the increased incidence of meningiomas is observed and it will be helpful for better outcomes of patients. We are presenting this case of transitional meningioma benign tumor WHO grade 1for its clinical, radioimaging and histopatholgical findings.

5. Source of Funding

None.

6. Conflict of Interest

None.

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Author biography

Sunil V Jagtap, Professor https://orcid.org/0000-0002-4844-7624

Loknath Srimanto Sarkar, Resident https://orcid.org/0009-0003-2473-7760

Shubham S Jagtap, Resident https://orcid.org/0000-0002-8304-8581

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