



A SILENT CASE OF LARGE CYSTIC MENINGIOMA

Neurosurgery

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ABSTRACT

Meningiomas are the most common extra-axial neoplasms. Meningiomas, in general may be benign or malignant. Cystic meningiomas are one of the subtypes of meningioma, mimicking an intra-axial lesion radiologically and clinically. The common clinical manifestations may include but not limit to headache, vomiting, seizures & focal neurological deficits. The cystic component is an uncommon finding that makes it difficult to distinguish between a meningioma and a glial neoplasm radiologically. Here, we discuss a 54-year-old female patient who presented with sub-acute onset of transient memory disturbances and a sudden neurological deterioration who was found to have a cystic meningioma.

KEYWORDS

Cystic meningiomas, extra-axial, benign, malignant, glial neoplasm, neurological deterioration

INTRODUCTION

Meningiomas are the most common intracranial tumors with wide variety of clinical presentation and radiological findings. They have greater incidence in females compared to males. They are derived from arachnoidal (meningothelial) cells. Cystic meningiomas account for 1.5 to 10% of all the meningiomas [1]. They are extra axial and may have a dural attachment (tail). Cyst within the tumor may mimic neoplastic lesion. They are most located in the frontal and parietal lobes, cerebral falx and cerebellopontine angles. They most commonly occur near surfaces of brain and in parasagittal region. Meningiomas constitute approximately 15% of all intracranial tumors [2]. Certain risk factors for meningioma have been outlined like history of exposure to ionizing radiation [3] (greater incidence if the exposure was during childhood). It also has a greater incidence in people deletion and inactivation of NF2 on chromosome 22 [4]. These tumors are generally solid but rarely may have a cystic component (2-4% incidence). WHO classified meningiomas as benign (Grade 1), atypical (Grade 2) and malignant or anaplastic (Grade 3). Based on histology, they have been further divided into the most common subtypes of transitional, fibroblastic and meningothelial [5]. We present a case of cystic meningioma with unusual clinical and radiological presentation.

Case Presentation

A 54-year-old female, hailing from Bengaluru, India presented with the history of transient memory disturbance lasting for few seconds to minutes (45 seconds to 2 minutes) since 15 days; with no past history of trauma, radiation exposure, past history of similar episodes or positive family history. There was no headache, vomiting, loss of consciousness, giddiness. She was initially considered for age related degenerative disorder and admitted under neurology. Her neurological examination at the time of admission was unremarkable. Post Magnetic Resonance Imaging (MRI), her sensorium deteriorated, and she was not responding to verbal commands and painful stimuli. Her pupils were 2mm bilaterally equal and not reacting to light. MRI (Figure 1) revealed (64*54*41mm) cystic lesion with solid component, mixed signal intensity in the left inferior frontal region. There was left lateral ventricle compression and severe mass effect. Thereafter, her neurological condition deteriorated progressively with Glasgow Coma Scale (GCS) E1V1M3 and pupils 4mm not reacting to light. She also had an episode of generalized tonic clonic seizure associated with vomiting following which her contrast study was postponed and an immediate decompressive craniectomy was performed under general anesthesia. Tumor was approached via middle frontal gyrus after corticectomy. The cystic component contained xanthochromatic fluid which was collected (~20ml). The solid component was firm, highly vascular, grayish, encapsulated and attached to the underlying falx (dura) which was excised. Post operatively, she was continued on mechanical ventilation for a day, following which she was gradually extubated. Her neurological condition improved and was shifted to the ward after a day. Post operation CT scan with contrast is shown in Figure 2. The histopathological examination of the specimen revealed meningothelial cells with whorl formation and indistinct borders

forming syncytia. The cystic component did not reveal any atypical cell. Histopathological examination revealed the diagnosis of meningothelial cystic meningioma (Nauta et al Grade 1 and WHO Grade 1). Unfortunately, she had developed right sided hemiparesis with power of 1/5 in the upper limb and 2/5 in the lower limb but moved her left side spontaneously. She received regular physiotherapy and was mobilized. Her rest of the stay in the hospital was uneventful and was discharged with GCS of E4V3M6. On her 3 month follow up visit, she was alert, conscious and obeying commands with GCS of E4V5M6. She was moving her left side with power of 5/5 while right upper limb with power of 2/5 and lower limb with power of 4-/5. She exhibited decreased activity and labile emotions, post tumor excision, as compared to before she presented to us for the first time. With informed consent, she underwent Left Fronto-temporo-parietal autologous cranioplasty. She withstood the surgery well and was discharged after a day of observation and Post operation CT scan changes as shown in Figure 3.

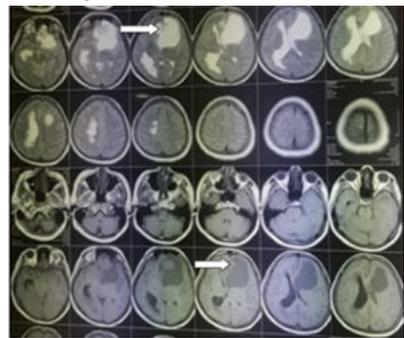


Figure 1: Preoperative MRI without contrast showing a heterogeneous T2weighted hyper tense, FLAIR hypo intense cystic lesion in the left basi-frontal region causing mass effect on ipsilateral lateral ventricle and midline shift.

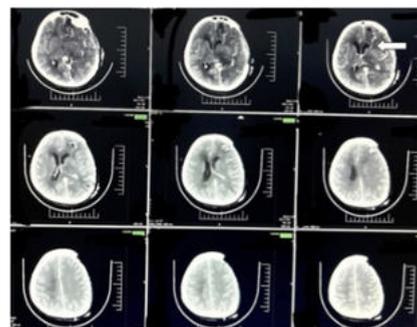


Figure 2: Post Decompressive craniectomy CT scan with contrast showing that the gross total resection of the tumor was achieved, and brain parenchyma edema was reduced. Left fronto-temporo-parietal craniectomy defect also appreciated.

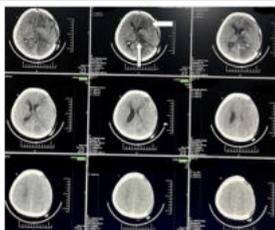


Figure 3: Post Cranioplasty CT scan with contrast showing a Left Fronto-temporo-parietal bone flap is replaced. Post operation gliotic changes are also seen.

DISCUSSION

Presence of cysts is a rare phenomenon in intracranial meningiomas in adults [6]. The term "cystic meningioma" is used for meningiomas with intra-tumoral or peritumoral cysts. The presence of an associated cyst makes it an unusual finding making it difficult to distinguish from glial neoplasms [2]. The cyst maybe (a) central or (b) eccentric in location; (c) its wall may contain nests of tumor cells; (d) it may be located adjacent to the tumor within brain parenchyma; or (e) it may consist of a CSF loculation trapped between tumor and brain [7-8] This is called the Nauta et al classification. There are several proposed mechanisms of cyst formation. One is cystic degeneration within the tumor; others are secretion of fluid by tumor cells, gliotic proliferation in adjacent brain with formation of fluid by glial cells, and loculation of CSF [8]. Cyst formation within the tumor also may be seen because of tissue necrosis in the malignant variants of meningioma. A cystic component or necrosis is more commonly seen in angioblastic and meningothelial meningiomas [7, 9], possibly because of their greater propensity for malignant transformation and secretion, respectively. The xanthochromic fluid from the cyst contains hemosiderin laden histiocytes, which can be due to intra-tumoral hemorrhage. Cystic meningiomas require special attention as they may lead to misdiagnosis upon initial evaluation. Combined MRI and Diffusion-weighted Imaging (DWI) maybe helpful in their diagnosis [10]. The cystic components are hypointense or mildly hypertense on DWI, whereas the Apparent Diffusion Coefficient (ADC) values are increased. Neuropathologic studies may be complicated by the difficulties frequently encountered in differentiating cystic meningiomas, particularly the meningothelial subtype from glioblastoma multiforme [11]. This subtype may simulate a glioblastoma at both the gross and microscopic levels. The cystic meningioma has been frequently misdiagnosed with CT, because the tumor may simulate glioblastoma, metastasis, abscess, or sarcoma [8] Presence of peritumoral edema can be a misleading finding too. Typical MRI findings include an extra-axial tumor with a broad dural base. The solid part shows intense enhancement, while the associated cysts may show wall enhancement, despite the absence of tumor cells [12]. However, the absence of Gadolinium enhancement of the cystic component does not rule out the presence of tumor cells [13]. The dural tail may also be seen in glioblastoma, parenchymal/dural metastasis and schwannomas. Small meningiomas in asymptomatic patients can be followed with serial imaging studies, preferably MRI, to assess for interval growth. Most accessible benign tumors undergo resection with a very low recurrence rate of 6%. It is necessary to remove the solid as well as the cystic parts to reduce the chances of recurrence. Surgically inaccessible tumors are treated with radiation therapy. Malignant tumors are treated with adjuvant radiotherapy or a combination of chemotherapy and radiotherapy, in addition to surgical resection [5].

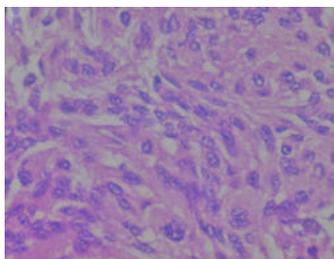


Figure 4: High-power view of the histopathology specimen showing lobules of spindle shaped monomorphic cells with ample cytoplasm and indistinct borders forming syncytia running in short fascicles and forming whorls indicating a meningioepithelial pattern. Nuclei are round to ovoid harboring fine chromatin and inconspicuous nucleoli.

CONCLUSION

A large cystic meningioma may have unusual clinical presentation which can lead to a misdiagnosis like an abscess, a glioblastoma multiforme (GBM), a metastatic tumor, hemangioblastomas etc. Therefore, in addition to Gadolinium enhancement MRI scan, microsurgical inspection and histological examination should be performed to confirm and differentiate. However, a combined MRI and Diffusion weighted imaging (DWI) are efficient in determining the type of cystic meningioma. Cystic meningiomas with peritumoral edema is considered an atypical and unusual presentation, hence, it is important to recognize variable features of meningioma, as to not confuse with another neoplastic/intracranial lesion.

Competing interest

No conflicts of interest or sources of funding.

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