International Journal of Science and Research (IJSR)
ISSN: 2319-7064
ResearchGate Impact Factor (2018): 0.28 | SJIF (2018): 7.426

A Case of Psammomatous Histological Subtype of Who Grade I Meningioma Located in the Olfactory Groove

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Abstract: ***Psammomatous meningioma is a rare histologic subtype of meningioma with an incidence of 3.8% compared to the other more common histologic subtypes, meningothelial followed by fibrous and transitional meningiomas, which account for approximately 80% of all meningiomas.Also the psammomatous subtype is more common in the spine compared to the intracranial location. Here we present a case of a 40 year old female with psammomatous histological subtype of WHO grade I meningioma, located in the olfactory groove presenting to the outpatient department with headache and mild cognitive decline.***

Keywords: Psammomatous, WHO grade I meningioma, olfactory groove

1. Introduction

Meningiomas are the most common nonglial primary tumors of the central nervous system and the most common extraaxial neoplasms, accounting for approximately 15% of all intracranial tumors.[1]Meningiomas are typically slow-growing tumours that arise from the meningothelial cells of the arachnoid.They are most common after the 5th decade of life and are often asymptomatic Women are more than two times as likely as men to develop a meningioma. Meningiomas may be found along any of the external surfaces of the brain as well as within the ventricular system where they arise from the stromal arachnoid cells of the choroid plexus. The most common locations include the parasagittal aspect of the cerebral convexity, the lateral hemisphere convexity, the sphenoid wing, middle cranial fossa and the olfactory groove.[2]

Histologically, meningioma cells are relatively uniform, with a tendency to encircle one another, forming whorls and psammoma bodies (laminated calcific concretions). As such, they also have a tendency to calcify and are highly vascularized.

The **World Health Organization (WHO)** classification of brain tumors is the most widely utilized tool in grading tumor types. The WHO classification scheme recognizes 15 variations of meningiomas[3] according to their cell type as seen under a microscope. These variations are called **meningioma subtypes**; the technical term for these cell variations is histological subtypes (Table 1).

The majority of lesions are benign WHO Grade I lesions, representing approximately 90% of cases.The histological subtypes of grade I meningiomas include meningothelial, psammomatous, secretory, fibroblastic, angiomatous, lymphoplasmacyte-rich, transitional, metaplastic and microcystic. They differ from the more aggressive meningiomas, WHO grade II (atypical) and WHO grade III (anaplastic), 5–7 % and 1–3 % of cases respectively, in their number of mitoses, cellularity, nuclear-to-cytoplasmic ratio, histological patterns and their relatively low risk of recurrence or aggressive growth pattern.[2]

The incidence of meningothelial meningiomas is the highest, followed by fibrous and transitional meningiomas (Table 2). These three subtypes account for approximately 80% of all meningiomas, and thus could be regarded as typical meningiomas. For this reason, other uncommon histological subtypes may be considered as imaging variants, and diagnosis is often challenging for radiologists solely based on imaging features of typical meningiomas.[4]

Table 1: Histological grading of meningiomas based on the current WHO classification

WHO Grade I - Benign	WHO Grade II - Atypical	WHO Grade III - Malignant
Meningiothelial	Chordoid	Papillary
Fibrous (fibroblastic)	Clear Cell	Rhabdoid
Transitional (mixed)	Atypical	Anaplastic
Psammomatous		
Angiomatous		
Microcystic		
Secretory		
Lymphoplasmacyte-rich		
Metaplastic		

Table 2: Incidence of histological subtypes of meningioma

Subtype	Incidence (%)	Subtype	Incidence (%)
WHO grade I		WHO grade II	
Meningothelial	57.8	Chordoid	1.4
Fibrous	11.1	Clear cell	1.1
Transitional	10.4	Atypical	3.4
Psammomatous	**3.8**		
Angiomatous	1.6	**WHO grade III**	
Microcystic	0.3	Papillary	1.1
Secretory	1.5	Rhabdoid	2.3
Lymphoplasmacyte-rich	1.1	Anaplastic	1.4
Metaplastic	1.6		

The common histologic subtypes of **ossified meningiomas** are transitional, psammomatous, and metaplastic. **Psammomatous meningioma** is a histologic subtype

of meningioma usually presented as a heavily calcified intracranial or spinal mass lesion. The meningothelial and psammomatous types are the most common involving the spine. This densely calcified tumour is characterized by the presence of numerous psammoma bodies. This tumour is classified as a benign meningioma (WHO grade one).

Here we present a case of psammomatous histological subtype of WHO grade Imeningioma, located in the olfactory groove.

2. Case Summary

A 40 year old female presented to the outpatient department with headache which was not relieved on medicationsandslow onset gradual cognitive declinefor 3 months.A neurologic examination revealed anosmia, however there were no visual deficits, weakness, or frontal release signs. In view of her age and gender, a neurologic lesion causing cognitive impairment (e.g. frontotemporal meningioma) was considered. Patient was referred for MRI Brain in view of the same.

MRI Brain plain and contrast study was done which revealed large well defined extra-axial dural based supratentorial altered signal intensity lesion in the anterior cranial fossain midline centered on the planum sphenoidale at the site of the olfactory groove. The lesion appeared isointense to cortex on T1W images and hypointense on T2W images (Fig.1).It showed surrounding mild perilesional vasogenic edema in the deep white matter in bilateral frontal lobes (Fig.1). It is showed patchy areas of restriction diffusion on DWI. Multiple areas of blooming were seen within the lesion on gradient echo sequence (FFE) representing dense calcifications (Fig.2). On post Gadolinium T1W images, it showed strong homogenous post contrast enhancement with small central non-enhancing area (Fig. 3 & 4). Location of the lesion was that of olfactory groove and planum sphenoidale. Surgery was performed and intra-operative findings were consistent with extra axial mass in relation to olfactory groove. Histopathology study showed tumour predominantly comprised of numerous calcified psammoma bodies and was reported as WHO Grade 1 Psammomatous meningioma.

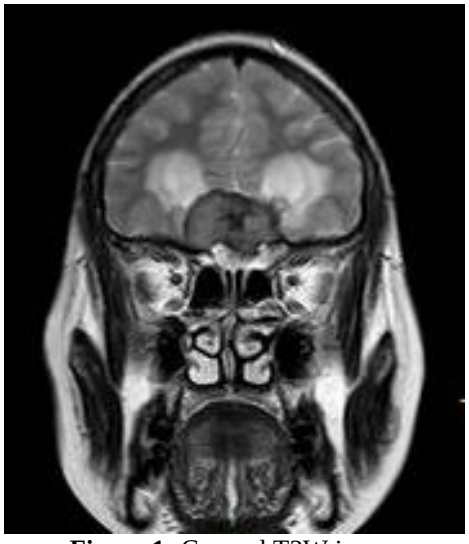

Figure 1: Coronal T2W image

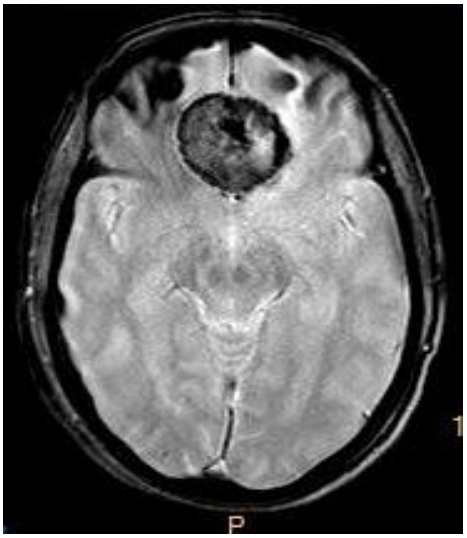

Figure 2: Axial Gradient image showing dense calcification of the tumor

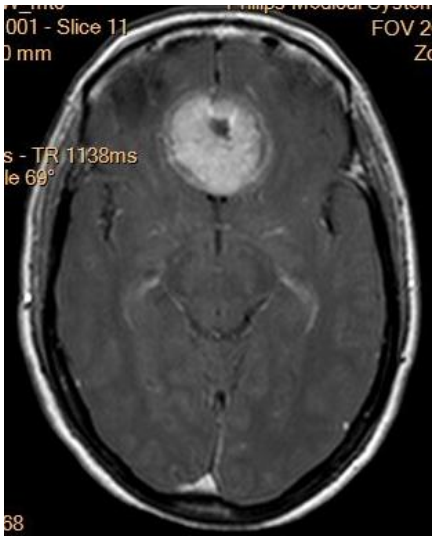

Figure 3: Post Gd T1W axial image

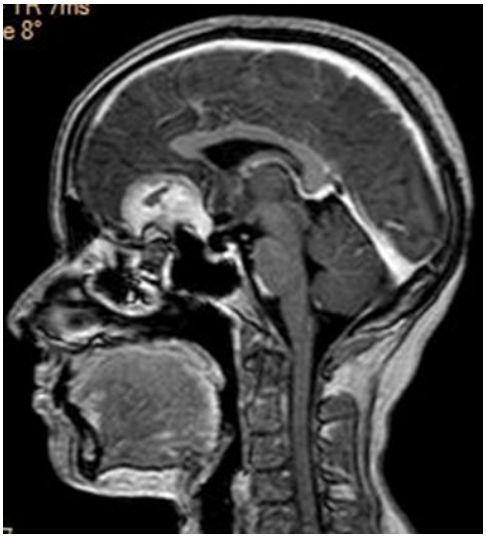

Figure 4: Post Gd T1W sagittal image

3. Discussion

Psammomatous meningioma is characterized by the predominance of psammoma bodies, when compared with

typical meningothelial meningiomas. This histological variant corresponds to WHO grade I. Psammomatous meningiomas share common locations with typical meningiomas in the cranium, but in the literature, many case reports described intraspinal psammomatous meningiomas, especially in the thoracic spine. CT reveals calcification diffusely or at the periphery of the tumor. T1-weighted images typically demonstrate isointensity with or without low signals. On T2-weighted images, the tumor commonly shows low intensity, but may show iso to high intensity as well. Such hypointensities on MR images do not always correspond to calcifications seen on CT, although calcifications are expected to cause signal reduction. ADC values of psammomatous meningiomas likely fall within the range of those of typical meningiomas. Post-contrast MR images show solid and strong enhancement in many cases (Fig. 4). Fewer cases can have weak contrast enhancement at the periphery of the tumor.Many rare histological variants, psammomatous subtype like in this case have their own imaging features, knowledge of which helps the radiologists to make a diagnosis of meningioma with more confidence.[4]

Olfactory groove and planum sphenoidale meningiomas are rare, constituting only 2% of all primary intracranial tumours. [6]

They are benign, slow growing tumours. These tumors arise from the floor of the anterior cranial fossa over the cribriform plate of the ethmoid bone. The site of origin frequently extends anteriorly or posteriorly and across the midline from this primary site. Although several speculations have been put forth, the exact reason as to why this site is more prone for occurrence of meningiomas is unclear. Due to their slow rate of growth, and their site and location in vicinity of the frontal lobes, these tumors achieve a significantly large size at the time of diagnosis and have relatively innocuous presenting clinical signs. A large tumor size, a firm and vascular nature, extension on both sides of the midline and a wide basal attachment are the general characteristics.[5]

Cognitive impairment and behavioural changes, the common presentation of this condition can be mistaken for dementia or depression.Anosmia is a common finding on physical examination, but it is not a typical presenting symptom similar to this case.

Although olfactory meningiomas are rare, they are treatable and the expected outcome is good for reversal of the change in thought processes.Hence, physicians should be aware that dismissing short-term memory loss as secondary to aging, depression, or dementia might lead to misdiagnosis and loss of opportunity for treatment. A high index of suspicion leads to earlier appropriate investigations, referral, and management.[6]

The case we present in addition to the emphasis on the psammomatous histological subtype of meningioma also illustrates the importance of suspecting an organic lesion when encountering a patient with features suggestive of early-onset dementia, especially in the primary care setting.

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