Imaging Spectrum of Cranial Meningiomas: you name it and it's there!

Poster No.: C-0474
Congress: ECR 2011
Type: Educational Exhibit
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Keywords: Neuroradiology brain
DOI: 10.1594/ecr2011/C-0474

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Learning objectives

1. To illustrate, discuss and review the diverse imaging appearances of cranial meningiomas.
2. To elucidate the typical, atypical and sometimes misleading imaging manifestations of this very common neoplasm.

Background

Meningiomas are the commonest extra-axial neoplasm and the most common primary nonglial intracranial tumor. Most meningiomas are benign, although atypical and malignant meningiomas also exist. Typical imaging characteristics include a well-circumscribed, homogeneously enhancing, extra-axial mass located over the cerebral convexity. There is seldom a doubt regarding their diagnosis owing to their characteristic appearance on imaging studies. However, at times the imaging features of meningiomas can be misleading. These atypical imaging manifestations can be in terms of signal characteristics, tumour location or behaviour. This educational exhibit illustrates a wide spectrum of atypical imaging appearances that can cause diagnostic dilemma. Since imaging plays a crucial role from the initial diagnosis to postoperative evaluation of meningiomas, the radiologist must be aware of the diverse, atypical and sometimes misleading imaging appearances of meningioma.

Imaging findings OR Procedure details

Meningiomas are the most common primary nonglial intracranial tumors which most frequently affect people in the middle and later decades of life, with a 2:1 female predilection [1-5].

The aetiology of the tumors is unknown in the majority of cases, although some are related to previous exposure to radiation and others occur as a manifestation of genetically inherited conditions such as neurofibromatosis type-2.

Histologically, meningiomas arise from meningothelial cells (arachnoid "cap" cells), and the tumor may occur in any area where arachnoidal cap cells exist [1-5].
As per the WHO classification, approximately 80% of meningiomas are Grade I (benign), with the remaining 20% being either Grade II (atypical) or Grade III (malignant) [6-8]. Meningiomas can be further classified into many different subtypes based on histological parameters. Of these subtypes, transitional, fibroblastic, and meningothelial are the most common ones.

**MACROSCOPIC CLASSIFICATION:**

Based upon their configuration, meningiomas can be classified into the commoner globular form known as "meningioma en masse" and the rarer flat variety called "meningioma en plaque" [1, 2] (Fig 1).

**TYPICAL IMAGING FINDINGS:**

A well-circumscribed, homogeneously enhancing, extraaxial mass on both computed tomography (CT) and magnetic resonance imaging (MRI).

Several imaging criteria help localize meningiomas as extracerebral and represent the key to diagnosis. These include:

"Cortical Buckling" of the underlying brain (Fig 2)

"Cerebrospinal Fluid Cleft"

"Pial Vascular Structures" interposed between the tumor and the brain surface

"Hyperostosis" of the adjacent bone is another highly specific finding for the extraaxial origin of meningioma (Fig 3)

On unenhanced CT scans the mass appears as an area of homogeneous hyperattenuation which enhances homogeneously following contrast material administration (Fig 4).

On magnetic resonance (MR) images, the typical features of meningiomas include a unilobar mass characteristically isointense on T1-weighted T2-weighted pulse sequences (Fig 5). On contrast enhanced MR images the mass homogeneously enhances (Fig 6) [1-5].
The dural tail sign which is thought to represent a reactive process in the meninges or neoplastic infiltration of the dura mater, is a characteristic but a non-specific sign of meningioma (Fig 6).

The most frequent calvarial change is hyperostosis of the adjacent skull, which may be seen on plain radiographs, as well as on CT and MR images (Fig 7).

**TYPICAL LOCATION**

There is usually a strong correlation between the location of the arachnoid granulations and the prevalent sites of origin for meningiomas. They arise in the following locations in descending order of frequency [2]:

- Convexity (20-34%) (Fig 8)
- Parasagittal (18-22%) (Fig 8)
- Sphenoid and middle cranial fossa (17-25%) (Fig 9)
- Frontobasal (10%) (Fig 10, 11)

**EN PLAQUE MENINGIOMAS** constitute 2-4% of all intracranial meningiomas. Morphologically, en plaque meningiomas are plaque-like or carpet-like lesions that grow in a radial fashion along the meninges (Fig 12). They tend to infiltrate the dura and many times invade the adjacent bones [9]. Enplaque meningiomas are much more commonly associated with adjacent bony hyperostosis which is disproportionately greater in size to the intracranial mass (Fig 13).

**ATYPICAL IMAGING FEATURES:**

The imaging features of meningiomas may be atypical in terms of signal characteristics, tumor location or behavior. However, atypical imaging appearances do not necessarily predict atypical histology [1].

**(a) ATYPICAL APPEARANCES:**

- Signal Characteristics:
  - T1-Hypointense tumor (Fig 14)
  - T2-Hypointense tumor (Fig 15)
  - T2-Hyperintense tumor (Fig 16)
Mixed signal-intensity tumor (Fig 17)

- Heterogeneous enhancement (Fig 18)
- Sun-ray appearance (Fig 19)
- Target appearance (central enhancement) (Fig 20)
- Peripheral ring enhancement (Fig 21)
- Cystic Meningioma (Fig 22, 23)
- Nonneoplastic peritumoral cysts (Fig 24, 25)
- Disproportionate Peritumoral Edema (Fig 26)
- Diffusion restriction (Fig 27)
- Choline peak at MR spectroscopy (Fig 28)
- Pneumosinus dilatans (Fig 29)

(b) UNUSUAL LOCATIONS

- Posterior fossa (9-15%):
  Tentorium cerebelli (2-4%), (Fig 30)
  Cerebellopontine angle (2-4%), (Fig 31, 32)
  Cerebellar convexity (5%), (Fig 33)
  Clivus (<1%), (Fig 33)
  Petrous apex (Fig 34)
    - Intraventricular (2-5%) (Fig 35, 36)
    - Orbital (<1-2%) (Fig 37, 38)
    - Cavernous sinus (Fig 39)
    - Tuberculum sellar meningioma (Fig 40)
    - Falx cerebri (Fig 41)
    - Ectopic (<1%) (Fig 42-46)

(c) ATYPICAL BEHAVIOUR

- Anaplastic or Malignant meningioma (Fig 47-49)
- Extracranial extension (Fig 50)
- Dural venous sinus invasion (Fig 51, 52)
- Extracranial metastases
- Radiation induced meningioma
- Multiple intracranial meningioma (non-neurofibromatosis) (Fig 53-54)
- Neurofibromatosis associated multiple meningiomas (Fig 55-57)
**Fig. 1:** Macroscopic (Imaging) classification of Meningiomas
EXTRACEREBRAL LOCALISATION

1. CSF cleft

2. Pial vessels interposed between the tumor and brain surface

3. Cortical buckling

Fig. 2: Signs for lesion localization
Hyperostotic changes can be seen adjacent to a parasellar meningioma

Fig. 3: Hyperostotic changes
The mass appears as an area of **homogeneous hyperattenuation** which **enhances homogeneously** following contrast material administration.

**Fig. 4:** Typical CT findings of meningioma
Typical MR findings

A well circumscribed unilobar mass characteristically isointense to the cortex on T1- and T2-weighted pulse sequences.

Fig. 5: Typical MRI findings of meningioma
The mass shows homogeneous enhancement and a characteristic 'dural tail' sign.

**Fig. 6:** Enhancement characteristics
Hyperostotic calvarial changes are seen adjacent to a large parietal convexity meningioma.
**Fig. 8:** Cerebral convexity & Parasagittal Meningioma
**Fig. 9:** Sphenoid wing and Parasellar Meningioma
Fig. 10: Middle cranial fossa & Frontobasal Meningioma
Fig. 11: Planum Sphenoidale Meningioma
A flat plaque-like layer of enhancing tumor tissue which is growing radially along the meninges.

Fig. 12: Enplaque Meningioma
ENPLAQUE MENINGIOMA

The flat plaque-like meningioma is associated with disproportionate thickening of the adjacent sphenoid wings

Fig. 13: Enplaque Meningioma
Fig. 14: Atypical T1-dark Meningioma
**Fig. 15:** Atypical T2-dark Meningioma

**Atypical SIGNAL Characteristics**

T2-HYPOINTENSE meningioma
Fig. 16: Atypical T2-bright Meningioma
Fig. 17: Atypical mixed signal-intensity meningioma

Atypical SIGNAL Characteristics

MIXED SIGNAL INTENSITY on T2-weighted scan
Fig. 18: Atypical Heterogeneous Enhancement
Fig. 19: Atypical ‘Sunray’ Enhancement

Instead of homogeneous enhancement a ‘sunray’ pattern of angioarchitecture and enhancement is displayed.
Central enhancement

An atypical predominantly central enhancement is seen giving rise to a bulls-eye or target appearance

Fig. 20: Atypical Central(Target) Enhancement
Fig. 21: Atypical Ring Enhancement
CYSTIC MENINGIOMA

- Cystic meningiomas are rare
- Cystic meningiomas account for 10% to 19% of intracranial meningiomas in infants, while only comprising 2% to 4% in adults.
- Most cystic meningiomas are located along the cerebral convexity, especially the frontal-parietal region.

Fig. 22: Atypical Cystic Meningioma
**CYSTIC MENINGIOMA**

- Cystic meningiomas are frequently confused for other entities.

- Reports in the literature have described incorrect preoperative diagnoses of glioma, malignant tumor, subdural hematoma, cerebral infarction and hydatid cyst, for histologically proven cystic meningiomas.

**Fig. 23:** Atypical Cystic Meningioma
Non-neoplastic peritumoral cysts

The pathogenesis of cysts accompanying extraaxial tumours is not well understood. However, it is postulated that CSF gets trapped within the cleft between the tumor & the adjacent brain leading to formation of these benign cysts.

**Fig. 24:** Nonneoplastic Peritumoral Cyst
Non-neoplastic peritumoral cysts

Most of these peritumoral cysts parallel CSF in signal intensity and do not show any enhancement unlike the intratumoral peripherally located (neoplastic) cysts. Occasionally, disproportionately large peritumoral cysts can be identified.

Fig. 25: Nonneoplastic Peritumoral Cyst
Disproportionate Peritumoral Edema

Although varying amounts of edema may be present with any of the meningioma cell types, most of the tumors have have only mild to moderate degrees of edema.

Severe edema tends to be associated with meningiomas of the syncytial or angioblastic cell type.

Fig. 26: Disproportionate Peritumoral Edema
Diffusion Restriction

DWI: Atypical and malignant meningiomas may show restricted diffusion

Fig. 27: Diffusion Restriction at DWI
Choline peak on MR-spectro

Usually MRS does not play a significant role in diagnosis.

Nonetheless certain features are present such as increase in Alanine and absent N-acetylaspartate and Glutamine.

A high Cho peak is unusual and can be misleading. However, recent reports have suggested that meningiomas can exhibit elevated choline with low or absent NAA and Cr and variable amounts of lactate.

Fig. 28: ‘Choline’ peak at MR-spectroscopy
Pneumosinus dilatans

An abnormally expanded, air-filled paranasal sinus can be associated with meningiomas or arachnoid cysts.

Detection of a Pneumosinus dilatans although rare can be a useful clue in identifying a cerebral meningioma, especially when it appears intrinsic to the brain.

Fig. 29: Pneumosinus Dilatans
Fig. 30: Tentorial Meningioma
Fig. 31: Cerebellopontine Angle Meningioma
Less than 5% of all intracranial meningiomas occur in the CP angle.

Meningioma is the 2nd most common mass lesion of the CP angle after acoustic schwannoma.

Meningiomas, tend to be larger, more hemispheric in shape and more homogeneously enhancing compared to acoustic schwannomas.

Fig. 32: Cerebellopontine Angle Meningioma
Fig. 33: Cerebellar Convexity & Clival Meningioma
Fig. 34: Petrous Apex Meningioma
Fig. 35: Intraventricular Meningioma
Intraventricular meningiomas account for 2-5% of intracranial meningiomas.

Intraventricular meningiomas arise from the tela choroidea.

80% arise in the lateral ventricles with a preference for the left trigone.

15% occur in the third ventricle, and about 5% within the fourth ventricle.

Meningioma is the most common trigonal intraventricular mass in an adult; typically demonstrates T2-hypointense signal and peritrigonal edema.

**Fig. 36:** Intraventricular Meningioma
Fig. 37: Orbital Meningioma
Orbital meningiomas account for < 2% of cranial meningiomas but constitute 10% of all intraorbital neoplasms.

Most arise from the optic nerve sheath between the globe and the optic canal.

May produce diffuse thickening of the optic nerve, a well-defined and rounded mass, or even an eccentric lesion with an irregular border.

Characteristically show a ‘tram track’ sign on contrast enhanced CT or MRI.

Fig. 38: Orbital Meningioma
Cavernous sinus meningiomas are rare.

Although, meningiomas are the most common primary tumor involving the cavernous sinus.

Most cavernous sinus meningiomas arise from the lateral dural wall, but sometimes they may be exclusively inside the sinus.

A dural tail frequently is seen extending away from the edge of the tumor.

Meningiomas characteristically constrict the lumen of the internal carotid artery (ICA).

**Fig. 39:** Cavernous Sinus Meningioma
Approximately 5% of meningiomas are suprasellar either arising from the diaphragma sellae or tuberculum sellae.

These meningiomas may invade the sella turcica or sometimes can be predominantly intrasellar.

Thickening and ossification of the jugum (arrows), presence of a ‘dural tail’ and downward displacement of the intensely enhancing pituitary gland favour a tuberculum sellae meningioma.

Fig. 40: Tuberculum Sella Meningioma
Fig. 41: Meningioma of the Falx-Cerebri
ECTOPIC MENINGIOMAS

- <1% of meningiomas develop extradurally.

- These ectopic meningiomas may arise within the intradiploic space, from the outer table of the skull, inside the paranasal sinuses, in the parotid gland, and from the parapharyngeal space.

- Theories to explain these sites of origin include derivation from the arachnoid around the cranial nerve sheaths or from arachnoid cells disseminated during the formation of the skull (i.e. ectopic inclusions).

Fig. 42: Ectopic Meningiomas
Fig. 43: Ectopic Intradiploic Meningioma
Intradiploic meningiomas are highly uncommon.

They are thought to arise from ectopic meningocytes or arachnoid cap cells trapped in the cranial sutures during moulding of the head at birth.

Frontoparietal and orbital regions are the most common locations.

Primary calvarial meningiomas are frequently confused preoperatively with a primary bone tumor of the skull.

**Fig. 44:** Ectopic Calvarial Meningioma
Fig. 45: Paranasal Sinus Meningioma
Meningioma occurring in the paranasal sinus are notably rare.

They are believed to arise from arachnoid ‘cap cells’ or meningocytes, which have migrated in the nerve sheath and become detached during development.

They are frequently seen in the frontal and ethmoid air cells.

Fig. 46: Paranasal Sinus Meningioma
Fig. 47: Malignant Meningioma
Up to 10% of meningiomas are atypical or malignant, characterized by nuclear disorganization, necrosis, prominent nucleoli, and increased mitoses on histology.

They are also more prone to recur (in 29%–41%) than typical meningiomas where the recurrence rate is low (7%–20%).

The age group for atypical and malignant meningiomas is about 10 years before the typical counterparts.

Fig. 48: Malignant Meningioma
The definitive diagnosis is histopathological; however, there are some radiological findings that can suggest the possibility of an atypical/malignant meningioma.

These include a heterogeneous appearance, indistinct tumor margins, a inhomogeneous enhancement and marked perilesional edema. T2-weighted images may exhibit flow void phenomena, corresponding to dilated high-flow drainage veins. Bone destruction, ‘mushrooming’ on the outer edge of the lesion, and extracranial extension may be seen.

**Fig. 49:** Malignant Meningioma
Osseous invasion, Extracranial extension and infiltration of the scalp is highly uncommon finding associated with intracranial meningioma.

Fig. 50: Extracranial extension
Fig. 51: Dural venous sinus invasion

A large parasagittal meningioma is seen invading and occluding the superior sagittal sinus.
Dural Venous Sinus Invasion

Parasagittal meningiomas often grow adjacent to cerebral venous sinuses, especially the superior sagittal, and may occlude the lumen of a sinus by either invasion or compression against the outer wall of the sinus. The invasion of the dural sinuses by meningiomas is a common event that may limit the extent of surgical excision.

**Fig. 52:** Dural venous sinus invasion
Multiple Meningiomas

- Multiple meningiomas is a condition in which the patient has more than one meningioma in several intracranial locations without signs of neurofibromatosis.

- The incidence of multiple intracranial meningiomas varies from 1 to 10% in different series.

Fig. 53: Multiple Meningiomas(Non-Neurofibromatosis)
Multiple Meningiomas

- Multiple meningiomas are not a specific disease entity and have no distinctive clinical, pathological or surgical features.

- Despite the multiplicity of sites, multiple meningiomas do not differ in prognosis from benign solitary meningiomas.

Fig. 54: Multiple Meningiomas (Non-Neurofibromatosis)
Multiple Meningiomas in NF-2

Contrast enhanced MR in a patient with NF-2 reveals multiple enhancing supra- and infratentorial dural based extra-axial masses consistent with meningiomas.

Fig. 55: Neurofibromatosis associated Meningiomas
Multiple Meningiomas in NF-2

- Neurofibromatosis 2 (NF-2) is an inherited autosomal dominant neurocutaneous syndrome.

- It is characterised by development of multiple central nervous system tumours such as multiple schwannomas, meningiomas and ependymomas.

- Meningiomas are the second most frequent tumor affecting about half of the patients and are often multiple.

Fig. 56: Neurofibromatosis associated Meningiomas
Multiple Meningiomas in NF-2

National Institutes of Health, Manchester
diagnostic criteria of NF-2 include:

[1] Bilateral acoustic schwannoma seen at CT
or MRI; or

[2] A first-degree relative with NF2 and either a
unilateral acoustic schwannoma or 2 of the
following: neurofibroma/ meningioma/
glioma/ schwannoma/ juvenile posterior
subcapsular lenticular opacity.

Fig. 57: Neurofibromatosis associated Meningiomas
Conclusion

Because meningiomas are so common, the radiologist must be aware of their less frequent and uncharacteristic imaging findings in order to suggest the correct diagnosis in cases that are atypical. This is often very satisfying since the imaging characteristics of meningioma are usually, but not always, diagnostic.

Personal Information

References