Tumors cyst of the brain with a mural nodule.

Poster No.: C-2786
Congress: ECR 2018
Type: Educational Exhibit
Keywords: Neoplasia, Cysts, eLearning, Education, MR-Diffusion/Perfusion, MR, CT, Oncology, Neuroradiology brain, CNS
DOI: 10.1594/ecr2018/C-2786

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Learning objectives

To learn common locations and imaging findings of tumors cyst of the brain with a mural nodule (CBMN).
Background

CBMN is one of the radiologic patterns of central nervous system tumors. In a classification of the patterns of contrast enhancement in the brain and meninges, this one is considered a subtype of intra-axial enhancement lesions included in the category of fluid-secreting low-grade primary neoplasm. The most common are: hemangioblastoma, pilocytic astrocytoma, ganglioglioma, pleomorphic xanthoastrocytoma and, as less common, tanycytic ependymoma, intraparenchymal schwannoma, desmoplastic infantile ganglioglioma and cystic metastasis. Other rare causes should be considered (infections or vascular anomalies).

Pilocytic astrocytoma

Formerly known as juvenile pilocytic astrocytoma, classified as World Health Organization (WHO) grade I, it is a well-circumscribed, often cystic, slow-growing tumor. Pilocytic astrocytomas are tumours of young people, with 75% occurring in the first two decades of life, typically late in the first decade (9-10 years). There is no recognised gender predisposition. There is a strong association with neurofibromatosis type 1 (NF1). NF1 associated tumours have a tendency to affect the optic nerves and chiasm. Typically it presents as a cystic cerebellar CMNT or as an enlarged optic nerve/chiasm/tract with variable enhancement. It is located in the cerebellum in 60% of cases. The pilocytic astrocytoma derives from astrocytic precursor cells and it is the most common primary brain tumor in children. Microscopically it shows classic biphasic pattern of two astrocyte populations: (a) compacted bipolar cells with Rosenthal fibers (Rosenthal fibers electron dense glial fibrillary acidic protein (GFAP) staining cytoplasmic inclusions); (b) loose-textured multipolar cells with microcysts, eosinophilic granular bodies. Immunohistochemistry reflects the astrocytic differentiation: GFAP: positive, S100: positive, OLIG2: positive, IDH R132H mutation: negative, p53 protein: negative or weak. Differential diagnosis include: medulloblastoma, atypical teratoid/rhabdoid tumour, ependymoma, haemangioblastoma, ganglioglioma, pleomorphic xanthoastrocytoma.

Hemangioblastoma

Hemangioblastoma, also named capillary hemangioblastoma, is a vascular neoplasm of uncertain origin currently classified as a WHO grade I tumor. It is found mostly in adults. Slight male predilection in adults: M:F ratio of 1.3-2.6:1. Peak incidence at around 30-60 years of age, but earlier in patients with vHL (Von Hippel Lindau). Sporadic cases make up approximately 75-80%, with the remainder being found in patients with vHL. Most frequently present with symptoms relating to: headaches: 70%, hydrocephalus and
symptoms of raised intracranial pressure: 50 %, cerebellar dysfunction: ~50-60%, altered mental state: 10%, polycythaemia due to erythropoietin production occurs in ~20% (range 5-40%) of cases. It presents as a CMNT in 60% of cases with the enhancing mural nodule abutting the pia; in 95% of cases, hemangioblastomas are in the posterior fossa, predominantly in the cerebellar hemispheres. The hemangioblastoma is a vascularized neoplasm with stromal cells of uncertain histogenesis. Historically they were assigned in the category of vascular meningiomas although they gained an independent WHO category from 1991. Haemangioblastoma is actually a capillary haemangioma and, despite the name with the affix of "blastoma", it is a low grade (WHO grade I) lesion (note that the calvarial haemangioma is a cavernous haemangioma). The tumour is usually well circumscribed with a highly vascular mural nodule almost always abutting pial layer and a peripheral cyst which has similar contents as blood plasma. Hence it is suggested that the cystic component most likely arises by exudation from the solid nodule vascular component. Regarding the pathogenesis of the formation of the cyst, a major hypothesis is that the absence of astrocytic end feet and tight junctions can be found in the microvessels of these neoplasms, which may lead to breakdown of the bloodbrain barrier, and have a role in cyst formation. The cystic fluid is xanthochromic, with a concentration of amino acids, alkaline phosphates, and mucoproteins similar to blood, suggesting that they originate by diffusion from the vascular component of the solid tumor.

Pleomorphic xanthoastrocytoma

Pleomorphic xanthoastrocytoma (PXA) is a WHO grade II distinct type of benign astrocytoma. Typically it is a supratentorial cortical mass with an adjacent enhancing dural tail. CBMN appearance is present in upwards of 60% of cases. The temporal lobe is the most common location, followed by the parietal and occipital lobes. They are rare tumours accounting for only 1% of primary brain tumours. Typically these tumours are found in young patients (children or young adults), with a peak incidence in the second and third decade of life (10-30 years-of-age). As these tumours have a predilection for the temporal lobe, they most frequently present with seizures (~ 75% of cases). They usually present as cortical tumours with a cystic component and vivid contrast enhancement. Features of slow growth may be present, such as no surrounding oedema and scalloping of the overlying bone. A reactive dural involvement expressed by a dural tail sign can be found. Calcifications are rare. From a pathologic point of view, the PXA is noted for cellular pleomorphism and xanthomatous change. It may originate from cortical (subpial) astrocytes or from multipotential neuroectodermal precursor cells common to both neurons and astrocytes.

It has a pleomorphic appearance meaning that it can be highly variable in the size and shape of cells and/or their nuclei. CD34 antigen may help differentiate PXA from other tumors and may be associated with cortical
dysplasia. Immunohistochemistry demonstrates expected glial marker reactivity. Less obviously, there is also variable reactivity for neuronal markers. GFAP: positive, although often only weakly, S100: positive, neuronal markers including synaptophysin, MAP2 and neurofilament: variable. Pleomorphic xanthoastrocytomas, as well as pilocytic astrocytomas (and many non-CNS tumours), exhibit BRAF mutations 6,7. The only reported association is with neurofibromatosis type 1, although this is not a strong association.

Although prognosis is good following surgical excision, with a 5-year survival of 90% and 5-year-disease-free-survival of 70%.

**Ganglioglioma**

The ganglioglioma is a well-differentiated, WHO grade I or II, slow-growing neuroepithelial tumor composed of neoplastic ganglion cells and neoplastic glial cells. In a minority of cases (5%) these tumours show aggressive behaviour and histopathologic features and are then called anaplastic gangliogliomas (WHO grade III). At this stage, no criteria for WHO II gangliogliomas have been established.

Typical features are a partially cystic component with an enhancing, cortically based mass in a child/young adult with temporal lobe epilepsy. Malignant transformation into glioblastoma multiforme has been reported. There are two theories on the etiology of the ganglioglioma: (a) it may be a neoplastic transformation of glial hamartoma or (b) it may derive from subpial granule cell transformation. Genetics: BRAF V600E mutations are encountered in 20-60% of cases. IDH: negative (if positive then the tumour is most likely a diffuse glioma). Local resection is the treatment of choice and determines prognosis. In the brain, where a reasonable resection margin can be achieved, the prognosis is good, with recurrence-free survival reported to be 97% at 7.5-year follow-up.

**Desmoplastic infantile ganglioglioma**

Desmoplastic infantile ganglioglioma (DIG) is a WHO grade I tumor, slow growing glioneuronal tumours arising from either cortical or deep grey matter, that affects children in the first 2 years. Boys are affected more commonly than girls. Symptoms of DIG are intracranial hypertension, sunset eye, enlarging head circumference, bulging fontanels, variable localizing signs, including seizures, or paresis. From a pathologic point of view, DIG has prominent desmoplastic stroma plus neoplastic astrocytes, with a variable neuronal component. DIG arises from subpial astrocytes; the large cyst contains xanthochromic fluid and the tumor may be firmly attached to the dura and brain tissue. They demonstrate essentially no growth over time, although very gradual increase in size has been described. Only one case of malignant transformation has been reported.
Prognosis is excellent, however, due to the difficulty in managing seizure medically, patients usually undergo resection and even in cases of incomplete resection, seizures frequently cease.

**Ependimoma**

Represent a relatively broad group of glial tumours most often arising from the lining the ventricles of the brain or the central canal of the spinal cord. They account for ~5% of all neuroepithelial neoplasms, ~10% of all paediatric brain tumours and up to 33% of brain tumours occurring in those less than 3 years of age. Posterior fossa: 60%.

There is no recognised gender predilection. Although they can occur at any age, the posterior fossa tumours tend to present more commonly in the paediatric age group (mean age at diagnosis is 6 years of age), with a smaller second peak for supratentorial tumours around the 3rd decade.

Associations a neurofibromatosis type 2 (NF2).

**Cystic meningioma**

Meningiomas are common tumors of the central nervous system that account for approximately 15% of all intracranial tumors and are the most common extra-axial neoplasm (i.e., neoplasm that does not arise from the brain or spinal cord). The WHO classifies meningiomas into many different subtypes based on histological parameters. Of these subtypes, transitional, fibroblastic, and meningothelial are the most common.

Histologically, transitional meningiomas are characterized by whorl formation with closely wrapped cells.

Most meningiomas are benign neoplasms with a WHO grade 1 classification. Approximately 8% of meningiomas are considered "atypical" (WHO grade 2) and tend to have a higher incidence of recurrence (2). The presence of necrosis, excessive mitotic activity, and evidence of brain invasion are histologic features that result in an "atypical" classification. Anaplastic meningiomas (WHO grade 3) account for <1% of all meningiomas and have a much shorter mean survival time, with a 5-year-survival rate of approximately 64%, compared with 95% for atypical meningiomas. "Benign" metastasizing meningiomas have also been reported, but are rare

**Metastases**

Brain metastasis is the most common intracranial tumor in adults. It presents clinically as headache, seizures or loss of cognitive or motor function. The incidence of brain
metastasis is rising with the increase in survival of cancer patients. Approximately, 20%-40% patients with cancer will develop brain metastases in the course of their disease.

A frank CBMN pattern in metastasis is rare. Recently, a case has been described in the literature. Cystic metastasis is much more common.

**Very rare cases**

Tanycytic ependymoma

Intracerebral schwannoma

Rosette-forming glioneuronal tumor (RGNT) of the fourth ventricle

Papillary glioneuronal tumor

Adjacent epidermoid cyst and primary central nervous system lymphoma

Vascular lesions

Neurocysticercosis
Findings and procedure details

Radiology Features

Pilocytic astrocytoma

Most common in the cerebellar hemispheres. Other typical location is optic nerve/chiasm/tract. In T1-weighted sequences, the solid portion of the CMNT is iso/hypointense to gray matter while the cyst content is iso- to slightly hyperintense to cerebrospinal fluid (CSF). In T2 and fluid attenuated inversion recovery (FLAIR) sequences, the solid portion is hyperintense to gray matter while the cyst content is hyperintense to CSF. In FLAIR sequences, the cyst content is not suppressed. The cyst wall occasionally enhances, but cyst wall enhancement does not necessarily mean the presence of neoplastic tissue and removal of the cyst wall does not affect the outcome in the case of pilocytic astrocytoma. Fig (1,2).

Hemangioblastoma

Most common in the posterior fossa. The nodular portion occasionally demonstrates flow voids visible in T1- and T2- sequences, while the cyst is slightly hyperintense in T1 images compared with CSF. In T2 and FLAIR images, both cyst and nodule are hyperintense. The enhancement of the wall of the cyst is rarely seen in hemangioblastoma. Fig (3,4).

Pleomorphic xanthoastrocytoma

Supratentorial superficial cortical mass. Usually in the temporal lobe. Cystic portion is isointense to CSF. Enhancement of adjacent meninges, with an appearance of dural tail.

The mass is round to oval but despite its circumscribed appearance, the tumor often infiltrates into the brain. On T1 images, the nodule appears hypointense or isointense to gray matter while the cystic portion is isointense to CSF. Sometimes an associated cortical dysplasia may also be seen. On T2 images, the pleomorphic xanthoastrocytoma looks hyperintense or may have mixed signal intensity, and surrounding edema is rare. On FLAIR images, the cyst content is normally suppressed. A peculiar feature is the presence of enhancement of adjacent meninges, with the appearance of a dural tail (in approximately 70% of cases) as visible in. The enhancing nodule often abuts the pial surface. Fig (5).

Ganglioglioma
Cortically based mass in a child/young adult with temporal lobe epilepsy. Calcifications inside the lesion are common. It is a well-circumscribed mass that often expands the cortex. Calcifications inside the lesion are a common finding. In T1 images it is hypointense to gray matter; rarely it can be hyperintense for the presence of Ca2+; and has associated cortical dysplasia. In T2 images it is typically hyperintense. Enhancement is usually moderate and heterogeneous. Their appearance on imaging is very variable: from a partially cystic mass with an enhancing mural nodule (~45% of cases) to a solid mass expanding the overlying gyrus. Contrast enhancement is variable. Fig (6,7,8).

Desmoplastic infantile ganglioglioma

Typical features are the presence of a large cyst with a cortical-based enhancing tumor nodule. Enhancement of the adjacent pia and the reactive dural thickening are quite specific findings. It tends to be frontal or parietal.

On T1 images, the hypointense cyst may contain septae while the nodule is heterogeneous. On T2 images the cyst is hyperintense and the nodule usually has a low signal. The solid tumor nodule enhances markedly along with the leptomeninges and dura adjacent to the solid tumor.

Ependymoma

The majority of intracranial ependymomas (60%) are located in the posterior fossa (infratentorial), usually arising from the floor of the fourth ventricle. This is especially true in children. The remainder (40%) are located supratentorially and up to half of these are intraparenchymal.

Posterior fossa ependymomas are apt to extend through the foramina of Luschka and Magendie, hence the term plastic ependymoma. This is a characteristic feature and can be seen on both CT and MRI.

Ependymomas are typically heterogeneous masses with areas of necrosis, calcification, cystic change and haemorrhage frequently seen. This results in a heterogeneous appearance on all modalities.

Intraparenchymal lesions (usually supratentorial) are usually large and variable in appearance, ranging from completely solid, enhancing masses to cysts with a mural nodule, or more heterogeneous masses. They are believed to arise from trapping of embryonic rests of ependymal tissue in the developing cerebral parenchyma.

CT: coarse calcification is common (50%), cystic areas (50%), solid component iso to hypodense, heterogeneous enhancement, variable haemorrhage.
MRI: T1 solid portions of ependymoma typically are isointense to hypointense relative to white matter. T2 hyperintense to white matter more reliable in differentiating tumour margins than non-contrast T1-weighted images (but less reliable than contrast enhanced T1).

T1 C+ (Gd) enhancement present but heterogeneous enhancement with gadolinium is useful in differentiating tumour from adjacent vasogenic oedema and normal brain parenchyma.

DWI/ADC restricted diffusion may be seen in solid components, especially in anaplastic tumour diffusion should be interpreted with caution in masses with significant haemorrhage or calcification.

MRS Choline peak elevation according to the cellularity of tumour,

NAA peak reduction elevated Cho/Cr ratio lipid and lactate rise when degeneration occurs. Fig (9,10).

**Cystic meningioma**

Meningiomas generally have the characteristic imaging appearance of a well-circumscribed, solid, homogeneously enhancing extra-axial mass on both CT and MRI. Adjacent osseous changes, most commonly hyperostosis, may also be seen. The term "cystic meningioma" has been used to describe meningiomas with intratumoral or peritumoral cysts. The presence of an associated cyst is an uncommon imaging feature that may make it difficult to distinguish the tumor from an intra-axial primary glial neoplasm. The presence of peritumoral edema, as in the case presented, can also be a misleading finding. The exact cause of cyst formation is not entirely known, although many mechanisms have been proposed. These include cystic degeneration of the tumor, secretion of fluid from tumor cells, or loculated cerebrospinal fluid from scar tissue within or adjacent to the tumor. Fig (11,12).

**Metastases**

The classical appearance of a metastasis is a solid enhancing mass with well-defined margins and extensive edema. Occasionally, central necrosis produces a ring enhancing mass. Multiple lesions with marked vasogenic edema and mass effect are typically seen in patients with brain metastases. Lesions are isointense to mildly hypointense on T1-weighted images, hyperintense on T2-weighted images or with FLAIR. Surrounding edema is relatively hypointense on FLAIR and T1-weighted images and hyperintense on T2-weighted images. Following administration of a contrast agent, solid, nodular, or irregular ring patterns of enhancement are seen. It has been shown that treatment with dexamethasone leads to a reduction in evidence on MRI of peritumoral edema and
occasionally, a lessening in the extent of contrast enhancement. Definitive diagnosis can be established on biopsy. Fig (13).
**Fig. 1:** Axial CT slices (A) without intravenous contrast and (B) with intravenous contrast. Intraaxial lesion in the cerebellar vermis. The lesion is cystic with a small solid area of 7 mm, which shows intense enhancement after intravenous contrast administration, compatible with Pilocytic astrocytoma.

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**Fig. 2:** Axial MR slices: (A) T1 sequence with intravenous contrast and (B) T2 sequence without intravenous contrast: Intraaxial lesion in the cerebellar vermis. The lesion is cystic with a small solid area of 7 mm, which show intense enhancement after intravenous contrast administration. And (C) Difusion-ws(B=1500): Intraaxial lesion in the cerebellar vermis. The lesion is cystic with a small solid area of 7 mm, without restriction in the diffusion, compatible with Pilocytic astrocytoma

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Fig. 3: Axial CT slices (A) without intravenous contrast (B) with intravenous contrast. Cystic tumoration with a solid pole in its lower region, located in the right cerebellar hemisphere, compresses and partially displaces the IV ventricle. The solid pole presents important enhancement after the iv contrast administration, compatible with Hemangioblastoma.

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**Fig. 4:** Coronal and Axial MR slices (A) T1 sequence with intravenous contrast (B) T2 sequence. Cystic tumoration with a solid pole in its lower region, located in the right cerebellar hemisphere, compresses and partially displaces the IV ventricle. The solid pole presents important enhancement after the iv contrast administration, compatible with Hemangioblastoma.

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Fig. 5: Axial and coronal MR slices. T2 sequence and T1 sequence with intravenous contrast: Pleomorphic xanthoastrocytoma. Superficial cortical expansive lesion in the left inferior temporal gyrus, with minimal bone remodeling of the temporal scale. It presents a heterogeneous signal, with a cystic area mainly in its medial side, and a more solid zone in its most peripheral portion. After intravenous contrast administration, there is a discrete enhancement of its solid portion and a discrete adjacent meningeal enhancement.

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Fig. 6: Axial CT (A) without intravenous contrast (B) with intravenous contrast. Partially cystic tumoration of 4 cm in diameter, with a solid pole in its lower region, located in the left parietal region, compresses and displaces the ventricular body and atrium of the left lateral ventricle. With heterogeneous enhancement after administration of iv contrast, compatible with Ganglioglioma.

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Fig. 7: Axial MR slices (A) T1 sequence without intravenous contrast (B) T2 sequence. Partially cystic tumoration of 4 cm in diameter, with solid pole in its lower region, located in the left parietal region, compresses and displaces the ventricular body and atrium of the left lateral ventricle, compatible with Ganglioglioma.

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**Fig. 8:** Sagital and axial MR slices: (A) T1 sequence with intravenous contrast (B) diffusion-weighted sequence (1000b). Partially cystic tumoration with solid pole in its lower region, with important and heterogeneous enhancement after administration of iv contrast, located in the left parietal region, 4 cm in diameter, compatible with Ganglioglioma.

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**Fig. 9:** Coronal and sagital CT slices: Mass in the left posterior fossa, 31 x 26 x 23 mm (CC x AP x T), which presents an hyperdense solid portion, gross calcifications and a cystic component with a thin wall. The mass seems to have its origin in the IV ventricle. The findings suggest ependymoma.

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Fig. 10: MR slices: (A) Axial, T2 and (B) Sagital, T1 sequence. Mass in left posterior fossa, 31 x 26 x 23 mm (CC x AP x T). The lesion is cystic with a small solid area, which show intense enhancement after intravenous contrast administration. The mass seems to have its origin in the IV ventricle. The findings suggest ependymoma.

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**Fig. 11:** Axial CT slices (A) without intravenous contrast (B) with intravenous contrast. A space-occupying lesion is visualized in the suprasellar cistern, with discrete compression but without clear invasion of the pituitary gland. It presents mixed component, solid - cystic, with heterogeneous enhancement of the solid parts after the administration of intravenous contrast.

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Fig. 12: MR slices (A) Coronal, T2 and (B) Sagital, T1 sequence with intravenous contrast. A space-occupying lesion is visualized in the suprasellar cistern, with discrete compression but without clear invasion of the pituitary gland. It presents a mixed component, solid - cystic, with heterogeneous enhancement of the solid parts after administration of intravenous contrast. A size of 2.2 x 2 x 3 cm (T x CC x AP). Compresses and later displaces the optic chiasm.

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**Fig. 13:** Axial CT slice (left) and MR slice (right) T1 sequence with intravenous contrast. A cystic lesion is visualized in the right parietal-occipital region, with a solid pole, which is intensely enhanced after the administration of intravenous contrast. Compatible con metastasis.

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Conclusion

CBMN is an important radiological pattern of brain lesions. So that, radiologic characteristics of the lesions can help to the differential diagnosis, that can be related, more frequently, to primary or secondary brain tumors.
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