

NEURO IMAGE

VOLUME 21, NUMERO 1
SEPTEMBRE 2005

NEURORADIOLOGY
MUHC-MNH

VOLUME 21, NUMERO 1
SEPTEMBER, 2005

SUMMER HEAT

IN THIS ISSUE ...

“New Angiography Suite”

Donatella Tampieri

“Neurenteric Cyst”

Federica Denegri

“Capillary hemangioma”

Maria Cortes

“Cyberspace”

Denis Melançon

The heat was high with the competitions at the XI FINA World Championships held in Montreal last July.

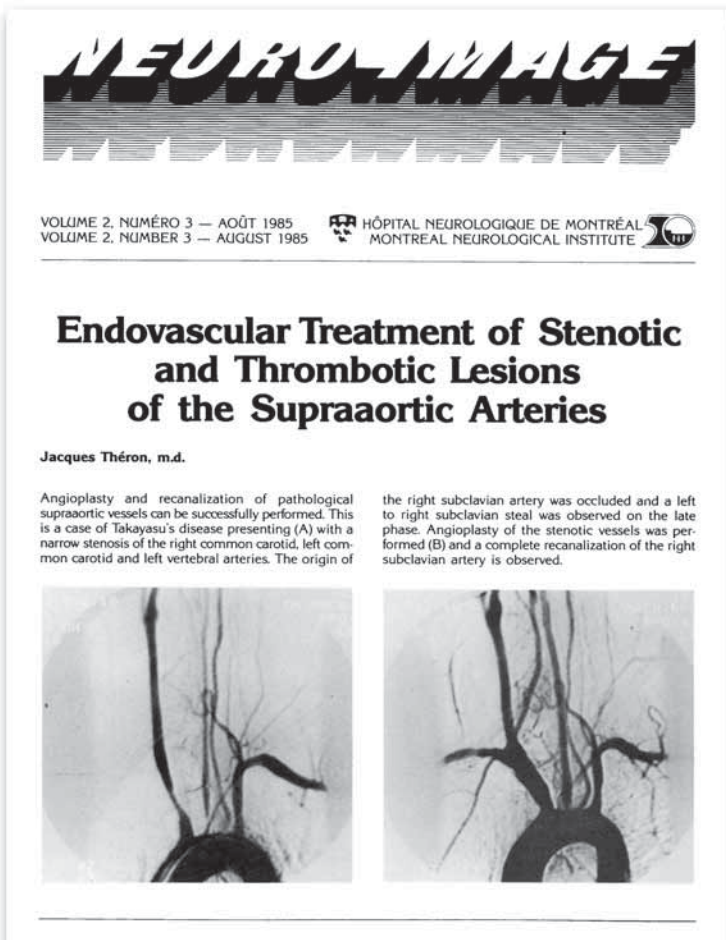
NEUROIMAGE HAS DIVED INTO CYBERSPACE

DR. DENIS MELANÇON

Everything is moving so fast, especially technology. We have gone through so many changes and advances, and since radiology likes to be with the state-of-the-art, cutting edge of technology, I would like to extend to you an invitation to connect to the world wide web and type in our website address:

<http://www.mni.mcgill.ca/neuroimage/index.html>

When you click onto this website, you can read the rest of this issue of NeuroImage Newsletter; and you can also see previous issues.



An earlier version of NeuroImage (1985)



A word of wisdom that easily applies to us radiologists

Best regards

Amicalement

Saluti affettuosi O Genki De

Herzliche Gruesse

Namaste

As-salaam alaykum

Filika

Respetos

Saudações

Bäst Hälsningar

Cordialmente

此致

敬意

NEURENTERIC CYST AT THE CERVICO-MEDULLARY JUNCTION: NEURORADIOLOGICAL FINDINGS, HISTO-PATHOLOGICAL CORRELATIONS & DIFFERENTIAL DIAGNOSIS

DRS FEDERICA DENEGR¹, MARIE-CHRISTINE GUIOT & DENIS MELANÇON

¹NEURORADIOLOGIA, OSPEDALE CTO, TORINO, ITALY

Neurenteric cyst (NC) are rare lesions occurring in the spinal canal, primarily in the cervico-thoracic spine; although they can occur anywhere from the cerebello-pontine angle to the coccyx, they are rarer in the posterior fossa or at the cervico-vertebral junction. We report a case of histologically proven neurenteric cyst an interesting MR finding of contrast fluid level inside the cystic lesion is probably explained by secretory function.

CASE REPORT

A 22 year old woman was seen in the Emergency Department complaining of sudden onset of discomfort in the head, nausea as well as a feeling of weakness or fainting. In the past days, she had experienced a feeling as though her head was too heavy for her neck. She also complained of pain in the back of the head and neck, getting worse during the night and exaggerated by sneezing and coughing. She was unable to turn her neck and she noticed difficulty swallowing; her symptoms were all worsened by moment. She denied any major illness or injury in the past, except a vague headache for the past year.

Neurological examination was normal. Blood tests obtained in the Emergency Room revealed no abnormality.

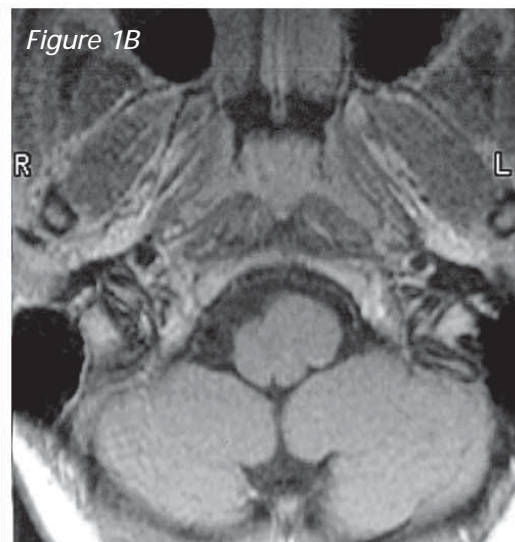
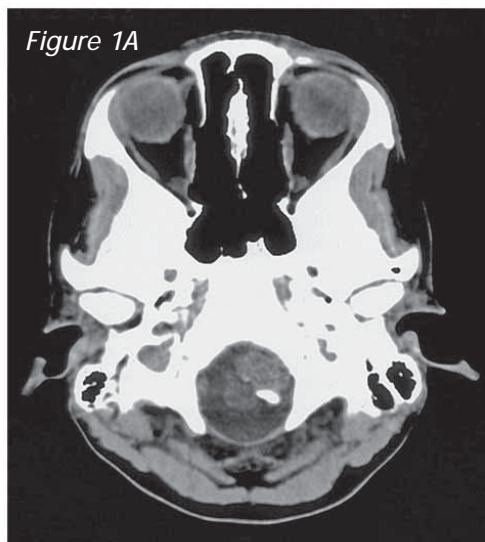
The CT scan performed with and without contrast showed an oval mass, isodense to brain, at the level of the foramen magnum, with some coarse calcifications (Fig. 1A). There was no abnormality in the bone, nor contrast enhancement of the lesion.

The MRI showed an intradural extra-medullary lesion, isointense in T1 (Fig. 1B), hyperintense in T2 and GR sequences (Fig. 2A, 2B), located in the anterior and left paramedian subarachnoid space, indenting slightly the medulla. A very thin tail arising from the cyst and going anteriorly was noted in all sequences. After gadolinium there was no real enhancement of the lesion but a iso-hyperintense fluid level pattern was seen inside the lesion in axial and sagittal T1 sequences.(Fig. 3A, 3B)

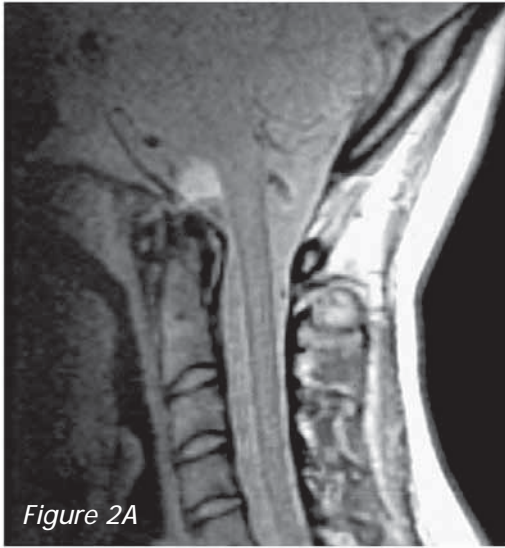
The patient underwent a left posterior fossa craniotomy to remove the lesion thought to be most likely a dermoid cyst. The operative findings described a cystic lesion, ventral to the medulla, indenting it and displacing the 11th and 10th cranial nerves posteriorly. There was a very diaphanous cyst wall and the contents varied: some parts were semisolid and others contained a mucoid liquid. The cyst wall was adherent to cranial nerves and brainstem. There was no clear attachment to the dura. Once the cyst was collapsed and most of its wall removed, the cranial nerves and brainstem structures appeared unremarkable. The cyst measured approximately 1.5-2.0 cm in diameter.

The post-operative course was uncomplicated and she was subsequently discharged home.

Microscopic examination showed fragments of a single layer or pseudostratified epithelium, columnar in appearance and non ciliated. Most of the epithelial cells contained a secretory product strongly positive for PAS and mucin stains (Fig. 4). This type of epithelium resembling gastro-intestinal or respiratory epithelium is characteristic of a neurenteric cyst.



DISCUSSION



This kind of lesion was called an enterogenous cyst by Harriman in 1958, who described an anterior intradural cyst with respiratory epithelium.

Enterogenous cysts (EC) are rare mass lesions arising during notochordal development and resulting from entrapment of endodermal tissue between a segmentally split notochord.

More than 80% of EC are located in the spine, whereas only 10% to

15% are intracranial. They can occur at any age, from birth to fifth decade, but they are definitely more frequent after age 20 (1). Although all the levels may be involved in the spine, there is a slight predominance for the cervical spine, especially the lower levels.(2) There is a slight predominance in males, with male-to-female ratio 3:2.(3)

During the third week of embryogenic development, invagination and migration of cells at the primitive streak and node of the embryogenic disk result in the formation of the notochord and mesodermal germ layer. The notochord is located in the dorsal aspect of the embryogenic disk, anterior to the depression of the primitive streak. The depression deepens, yielding the neural groove and the neural crest infolding until the neural tube is closed. The neural tube closure starts in the region of the fourth somite (future cervical region) and continues in both cranial and caudal directions. The neural tube contains communications between the notochord and the yolk sac, and the amnion: these communications, referred to as the canals of Kovalevsky, normally involute about the 17th day of embryologic life (4). Subsequently, notochord detaches from the underlying endoderm. If a communication persists, an abnormal split of notochord may occur; also a persistent adhesion between endoderm and ectoderm or between notochord and endoderm, resulting in a variety of malformations which depend on the ability of the notochordal abnormal development to repair.(5)

In our case, a bone defect was not seen on CT. A very thin linear hyperintensity in T2 and Proton density extended forward to end blindly; it can be compatible with a remnant of a fistulous connection corresponding to Kovalevsky canal.(6)

RADIOLOGICAL FINDINGS

The MR has replaced CT and CT myelography although CT, done with thin sections, has an important role to pick up abnormality in the bone. Usually the density of NE is isodense to CSF with no calcifications and no contrast enhancement. In our patient, some calcifications were present in the inferior part of a grossly round hypodense lesion.

The signal features are very variable and different patterns of signal have been described in the literature. Neurenteric cysts may be isointense to hyperintense relative to CSF on T2-weighted MR images. On T1-weighted MR images, most neurenteric cysts are isointense or slightly hyperintense relative to CSF. Occasionally, homogeneous, very bright signal intensity on T1-weighted MR images and/or low intensity on T2-weighted MR images may be seen. These signal characteristics are thought to correlate with protein content or hemorrhage within the cysts (3). The absence of contrast enhancement is usually reported, according with cystic nature of these lesions and their poor vascularization.

In our patient, a smooth intradural extra-axial lesion, slightly hyperintense in T2 and fairly hyperintense in PD to CSF was seen in front of the cervical-medullary junction. After gadolinium injection a small hyperintense

level was demonstrated inside the cyst in axial and sagittal T1 images: this finding has never been described as no enhancement usually occurs.(5,7) We suppose that a secretion of contrast inside the cyst has to be considered to explain this behaviour since epithelial columnar pattern indicating a foregut origin and secretory products have been found in pathology.

DIFFERENTIAL DIAGNOSIS

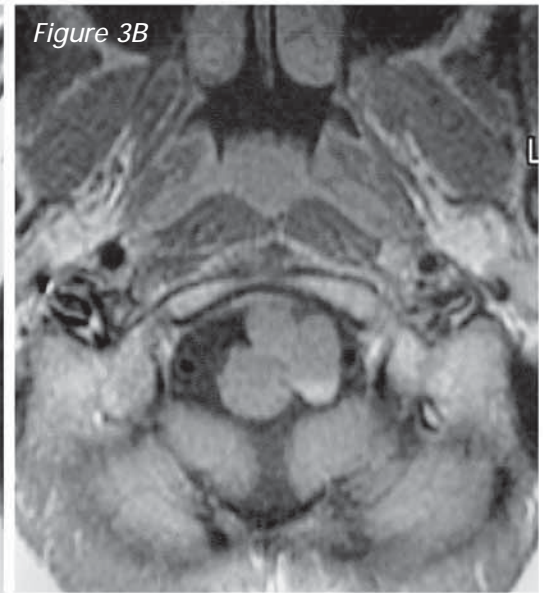
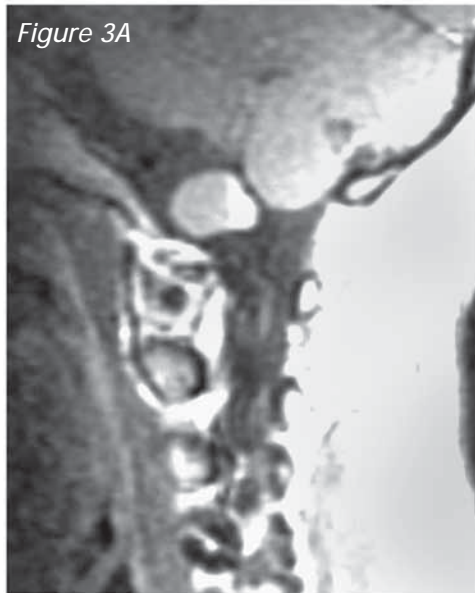
The differential diagnosis of cystic masses in this region include arachnoid cyst, epidermoid cyst, dermoid cyst; also the possibility of cystic neurinoma arising from lower cranial nerves has to be considered. This latter possibility should be distinguishable on the basis of contrast enhancement with ring-pattern due to marginal solid rim tissue.

Epidermoid cyst is a congenital disorder due an abnormal separation of the neural tube resulting in an epithelial inclusion cyst that has predominantly paramedian location,

with density identical to CSF on CT; on MR, the signal relative to CSF is slightly hyperintense in T1 images, hyperintense in proton density, and iso-hyperintense in T2 (10), although high signal epidermoid cysts on T1-weighted MR images have been reported (11). Flair MR may be helpful in differentiating epidermoid from arachnoid cyst, signals of epidermoid being similar to that of brain parenchyma, whereas that of arachnoid cyst are similar to spinal fluid(12). The low signal intensity of neurenteric cyst on diffusion-weighted images may enable differentiation from epidermoid cysts even if these cysts exhibit similar intensities on conventional MR images.(3) Dermoid cyst predominantly has a midline location and the density on CT and signal on MR are typically of fatty tissue.

The correct preoperative diagnosis of NC may be difficult since the MR signals are variable, depending on the secretory products from the epithelial lining. In addition, the numerous synonyms for these cysts have caused a degree of confusion regarding their histogenesis. NC have histological features similar to cysts of neuroectodermal origin and thus need to be differentiated from choroidal and ependymal cyst. A practical subdivision is provided by Elmadbouth(9) into two main group: endodermal or neuroectodermal. Within the former category are epithelial cysts, which have a connective tissue stroma, are lined by cuboidal or columnar epithelium, but have no definite histologic evidence of origin. When they occur within the ventricular system they are termed colloid cysts; otherwise, they are called enterogenous or neurenteric or simply epithelial cysts. Ependymal and choroidal cysts fall into the second category. Light microscopy often fails to differentiate between these cysts: for example, cilia are common to both ependymal and enterogenous cysts and there is a histologic similarity between these two entities and colloid cysts.

Electron microscopy helps to further differentiate. The walls of enterogenous cysts and colloid cysts are characterized by the presence of both columnar ciliated and nonciliated lining. The ciliated cells have no granular coating material. Nonciliated cells with microvilli are present in every case with characteristic surface granular glycocalyx coating. They possess features of secretory function, have prominent nuclei, and, in some cases, tight junctions are present. The appearance is similar to that of respiratory epithelium and is consistent with an endodermal origin. In contrast, cysts with ependymal and choroid plexus lining are composed of a single cell type and show no signs of keratinization, mucin production, or glycocalyx coating (9). Histochemical techniques suggest the origin of the cyst but do not provide a definitive diagnosis.



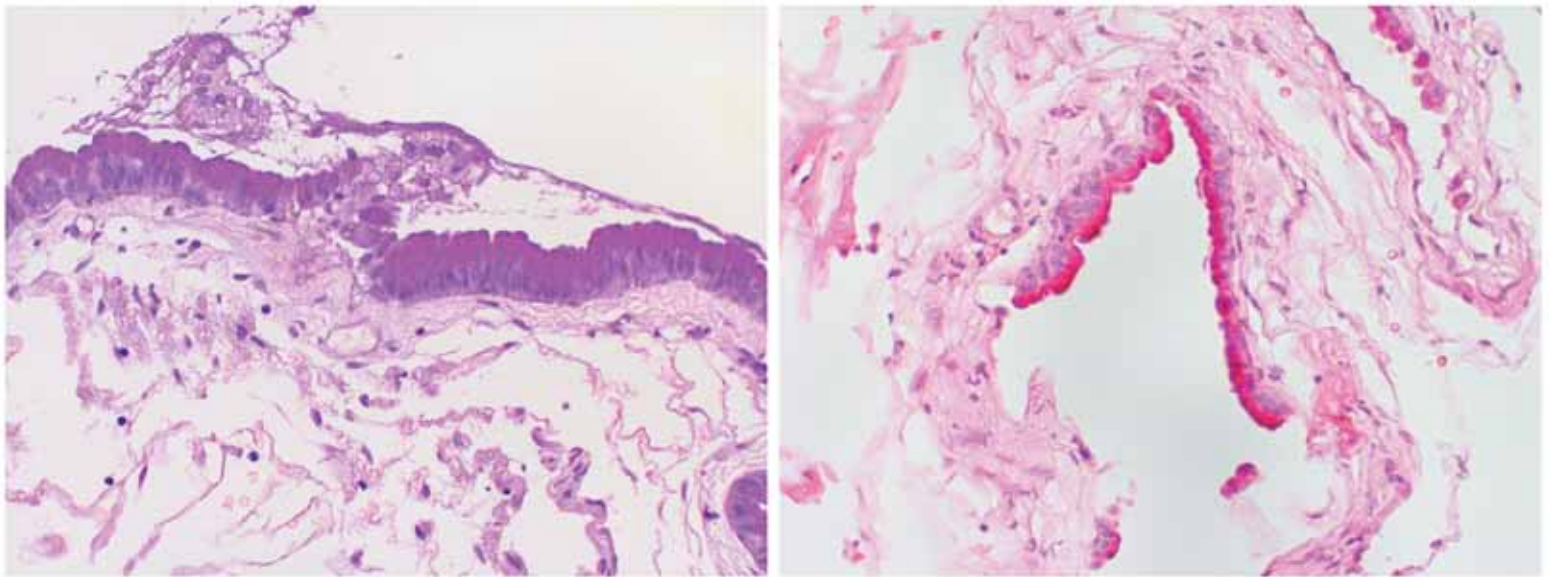


Figure 4 (Left) Periodic Acid Shift (PAS) Stain (right) Mucin Stain

PATHOLOGY

The pathology findings indicate the real origin of these lesions, showing an epithelium of digestive or respiratory type lining the cavity of the cyst. A fibrous capsule, variable in thickness, is always present and the cysts are lined by epithelium varying from cuboidal to columnar and occasionally pseudo-stratified type. The epithelium is strongly PAS positive and rests on a connective tissue base. A varying number of goblet cells are present within the epithelium and are responsible for the mucinous content. (8). Electron microscopy can reveal areas of ciliated epithelium whereas non ciliated cells with microvilli are present in every case with characteristic surface granular glycocalyx coating. They possess features of secretory function, having prominent nuclei, and, as well, tight junctions are present (9).

In our patient, a typical fragment of foregut epithelium was found with secretory products: the secretory function gives support to the possibility of secretion of contrast inside the lesion, a fact never seen and described before in literature. In addition several calcifications have been found.

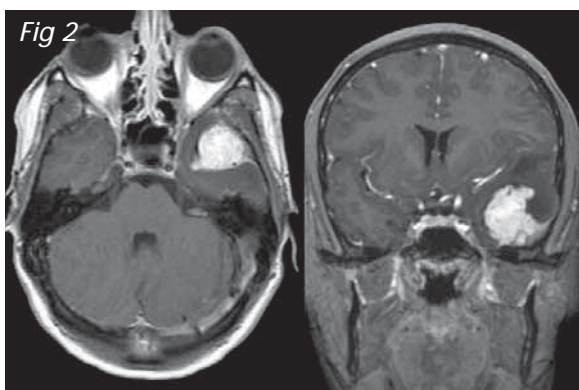
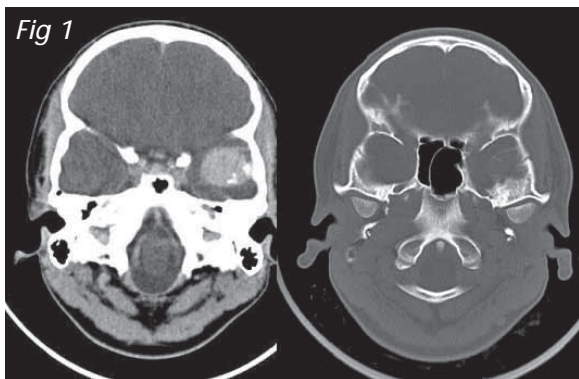
REFERENCES

1. Agnoli AL, Laun A, Schonmyr R. Enterogenous intraspinal cyst. J Neurosurg 1984;61:824-840.
2. Rodaci MA, Teixeira WR, Boer VHT, et al. Intradural extramedullary high cervical neurenteric cyst. Neuroradiology 1987; 29:588.
3. Shakudo M, Inoue Y, Ohata K, Tanaka S. Neurenteric Cyst with Alteration of Signal Intensity on Follow-up MR Images. AJNR Am J Neuroradiol 2001; 22:496-498.
4. Naidich TP, McLone DG. Growth and development. In Kricum ME. Imaging Modalities in spinal disorders, pp.1-19. Philadelphia, WB Saunders, 1988.
5. Brooks BS, Duvall ER, El Gammal T, et al. Neuroimaging features of neurenteric cyst: analysis of nine cases and review of literature. AJNR Am J Neuroradiol 2001 May 1993;14:735-746.
6. Martin AJ, Penney CC. Spinal neurenteric cyst. Arch Neurol 2001; 58:126-127.
7. Pierot L, Dormont D, Oueslati S, et al. Gadolinium-DTPA enhanced Mr imaging of intradural neurenteric cyst. J Comput tomogr 1988; 12(5):762-764.
8. Russell D, Rubinstein LJ. Pathology of tumors of nervous system. 5th edn. Baltimore: Williams et Wilkins; pp704-705, 690-695.
9. Elmadbouth H, Halpin SFS, Neal J et al. Posterior fossa epithelial cyst: case report and review of the literature. AJNR Am J Neuroradiol 2001 20:681-685.
10. Kallmes DF, Provenzale JM, Cloft H et al. Typical and atypical MR imaging features of intracranial ependymoid tumor. AJR Am J Roentgenol 169:883-887.
11. Ochi M, Hayashi K, Hayashi T, et al. Unusual CT and MR appearance of an epidermoid tumor of the cerebellopontine angle. AJNR Am J Neuroradiol 1998;19:1113-1115.
12. Tsuruda JS, Chew WM, Moseley ME, Norman D. Diffusion-weighted MR imaging of the brain: value of differentiating between extraaxial cysts and epidermoid tumors. AJR Am J Roentgenol 1990;155:1059-1065.

INTRACRANIAL DURAL CAPILLARY HEMANGIOMA: A CASE REPORT

DRS. MARIA CORTES, DONATELLA TAMPIERI, MARIE-CHRISTINE GUIOT, ANDRE OLIVIER & DENIS MELANÇON

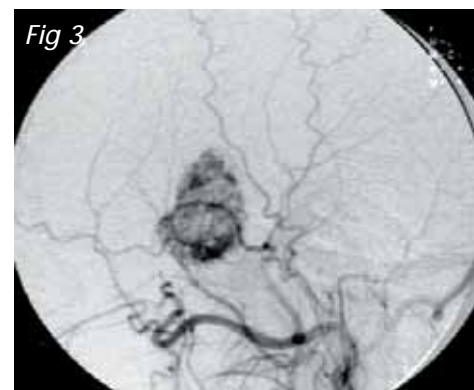
Capillary hemangiomas are more frequent in children and often locate in the skin, oral mucosa, orbits and scalp.³ They rarely involve the CNS^{1,2} and most cases affect the spinal cord, nerve roots or cauda equina.² We present the case of a young adult with an unusual hemangioma originating from the temporal dura.



CASE REPORT

A 21y.o. female presented with progressing stabbing headaches of three weeks, nausea, and vomiting. Her medical history and physical and neurological exams were unremarkable. Infused CT revealed a homogeneous enhancing lesion in the left middle cranial fossa. The greater sphenoid wing looked remodeled (Fig 1).

An enhanced MRI showed well-demarcated margins of the lesion and dural tail sign (Fig 2). Angiography revealed hypervascular lobulated tumoural blush, and persistent staining. The main arterial supply was arising from enlarged middle meningeal and accessory meningeal branches (Fig 3).



After successful endovascular embolization the tumour was completely resected surgically. Histopathology showed a capillary hemangioma.

DISCUSSION

Capillary hemangiomas (CH) are frequent tumours in children and in 50% of the cases affect the head and neck. The CNS is a rare location for these tumours, and most cases reported involve the spinal cord of adults.² The descriptions of isolated intracranial CH are scarce. They may involve the brain parenchyma or the dura. Our case adds to the short list of intracranial dural CH. The CT and MRI features in dural CH are similar to those of meningioma, and the literature does not seem to identify characteristics that would allow imaging differentiation between these lesions. In our patient the presence of a dural tail favored meningioma, however, the bone remodeling and the angiographic appearance (Figs. 1,3) lead us to consider hemangiopericytomas, hemangiomas, and angiosarcomas in the differential diagnosis. Histopathology confirmed the diagnosis of CH.^{1,2,3} (Photomicrograph Fig 4)

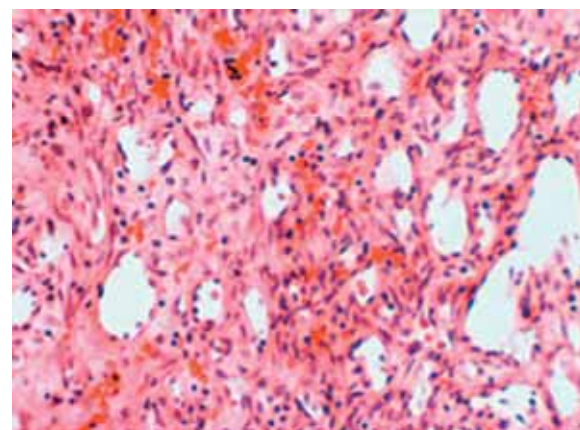


Fig 4: H&E High power photomicrograph: Dilated and flattened blood vessels lined with endothelium.

CONCLUSION

Intracranial dural capillary hemangioma is rare. Its radiological appearance may be misleading due to the resemblance to meningioma. Careful correlation of CT, MRI, and Angiography may provide clues to the presence of hypervascular tumours, among them, capillary hemangioma.

REFERENCES

1. Simon SL, Moonis G, Judkins AR, Scobie J, Burnett MG, Riina HA, et al. Intracranial capillary hemangioma: case report and review of the literature. *Surgical Neurology* 2005;64:154-159
2. Abe M, Tabuchi K, Tanaka S, Hodozuka A, Kunishio K, Kubo N, et al. Capillary hemangioma of the central nervous system. *J Neurosurg* 2004;101:73-81
3. Willing SJ, Paye-Petersen O, Aronin P, Faiths S. Capillary Hemangioma of the Meninges. *AJNR* 1993;14:529-536

NEW NEURO ANGIOGRAPHY SUITE AT THE MNH/MNI

DR DONATELLA TAMPIERI, MD

The new Neuro-Angiographic suite has opened at the Montreal Neurological on June 13th, 2005. The equipment is a GE LCN bi-plane unit that enables 3D rotational angiography and MRI/Angiography Fusion.

The 3D rotational angiography is extremely helpful in the treatment planning for intracranial vascular lesions, particularly aneurysms and arterio-venous malformations (AVMs). A basilar artery aneurysm prior to endovascular treatment is shown in Fig 1A. Fig 1 B demonstrates the mass of coils (in yellow) within the aneurysm completely occluded.

The length of the diagnostic and interventional procedures is significantly decreased.

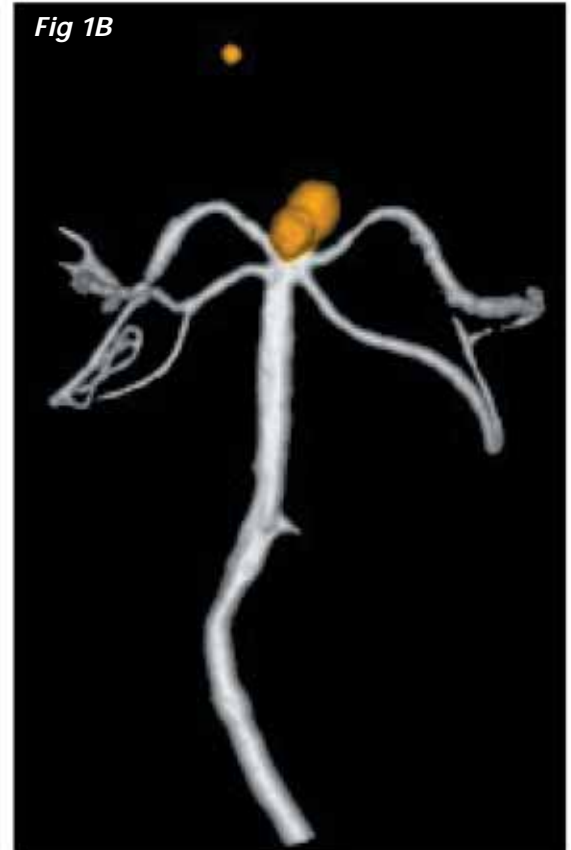
The 3D rotational angiography enables a more detailed demonstration of the aneurysm and its relationship with the parent artery.

The software of this equipment allows fusion of images obtained with different modalities, therefore an angiogram may be superimposed to a MRI image to demonstrate the relation of the aneurysm with the surrounding brain.

Fig 1A



Fig 1B



THIS NEWSLETTER IS SPONSORED BY

Department of Radiology
MUHC/MNH

BERLEX CANADA INC.

Boston Scientific
Delivering what's next.™

NEURIKON

Volume 21 – number1 – Bibliothèque nationale, ISSN 1180-0844 National Library of Canada, Production – Denis Melançon – Neurikon Inc.
Layout, Graphic Design by J.P. Acco at the Department of NeuroPhotography at the Montreal Neurological Hospital (9-2005)