Meningioangiomatosis

Meningioangiomatosis (MA) is an uncommon cerebral cortical mass lesion characterized histologically by cortical meningiovascular proliferation that may extend to involve the overlying meninges [1].

Classification

There are two populations of patients with MA: (1) symptomatic children and young adults who present with headaches or seizures and (2) asymptomatic individuals (mostly those with neurofibromatosis) in whom MA is discovered as an incidental finding

Nearly 50% of the reported cases showed no other stigmata of neurofibromatosis [2] [3] [4] [5] [6] [7] [8] [9] [10] [11] [12] [13] [14] [15].

Pathogenesis

Its pathogenesis is uncertain, but the favored hypothesis is that MA represents either an occult vascular malformation, which is later accompanied by meningioendothelial cell proliferation without evidence of malignancy, or a congenital hamartomatous malformation [16] [17] [18].

Histology

MA has many of the characteristics of an endotheliomatous meningioma that is located within the cerebral cortex, but it may extend to the leptomeninges along the Virchow-Robin spaces associated with abnormal blood vessels [19] [20] [21] [22].

Clinical

MA exhibits a wide range of clinical, imaging, histopathological and electrophysiological features, making the diagnosis difficult [23].

Free et al., reported two cases who presented headache and vision change, somewhat suggestive of migraine [24].

Diagnosis

The combination of magnetic resonance imaging (MRI) and computerized tomography (CT) can establish the diagnosis of MA [25].

MRI findings in MA correspond to the histological picture. However, the appearance on imaging is non-

specific and may suggest cystic atrophy, angioma and tumours. Several abnormalities have been found in close proximity to MA lesions, i.e. meningioma, oligodendroglioma, arteriovenous malformation, encephalocoel and orbital erosion. In spite of histopathological diversity, MA lesions are either predominantly cellular or vascular. Immunohistochemical results are inconsistent among cases, add little to the diagnosis, and do not support a meningeal origin. Electrocorticographic recordings from the surface and within MA lesions revealed a spectrum of electrophysiological expressions. Intrinsic epileptogenicity of MA lesions was documented in some cases. Epileptogenicity was confined to the perilesional cortex in some patients and it was complex (extralesional, multifocal, generalized) in others [26].

Differential diagnosis

MA should be considered in the differential diagnosis of cortical lesions, particularly in patients with NF2 [27].

Outcome

In symptomatic patients, complete or partial resection is curative.

In the series of Wiebe et al., became seizure-free postoperatively compared with 68% previously reported, and >70% of our patients and those in the literature continued to require antiepileptic drugs. This is in keeping with the diverse electrophysiology of MA and suggests a less optimistic postoperative outcome than previously recognized [28].

For Feng et al., MA is curable and the prognosis is excellent since most patients became free of seizure and recurrence after surgical treatments [29].

Seven patients with MA at the Department of Clinical Neurological Sciences, University of Western Ontario, London, Canada were investigated and their outcome was assessed. A review of the literature is included. MA exhibits a wide range of clinical, imaging, histopathological and electrophysiological features, making the diagnosis difficult. Sporadic MA cases are not associated with neurofibromatosis and the two disorders are genetically distinct. Medically refractory, localization-related epilepsy is the commonest presentation in sporadic cases, but atypical presentations also occur. Unlike sporadic cases, MA with neurofibromatosis is often found incidentally, does not produce seizures, occurs less frequently (ratio of 1:4), and is multifocal. MRI findings in MA correspond to the histological picture. However, the appearance on imaging is non-specific and may suggest cystic atrophy, angioma and tumours. Several abnormalities have been found in close proximity to MA lesions, i.e. meningioma, oligodendroglioma, arteriovenous malformation, encephalocoel and orbital erosion. In spite of histopathological diversity, MA lesions are either predominantly cellular or vascular. Immunohistochemical results are inconsistent among cases, add little to the diagnosis, and do not support a meningeal origin. Electrocorticographic recordings from the surface and within MA lesions revealed a spectrum of electrophysiological expressions. Intrinsic epileptogenicity of MA lesions was documented in some cases. Epileptogenicity was confined to the perilesional cortex in some patients and it was complex (extralesional, multifocal, generalized) in others. Only 43% of our patients became seizure-free postoperatively compared with 68% previously

reported, and >70% of our patients and those in the literature continued to require antiepileptic drugs. This is in keeping with the diverse electrophysiology of MA and suggests a less optimistic postoperative outcome than previously recognized [30].

Three patients, age range of 2-16 years old, presented with episodes of seizure. The patients demonstrated no family history or stigmata of neurofibromatosis type II. Electroencephalogram (EEG) was unremarkable for epileptiform activity. Magnetic resonance imaging (MRI) revealed enhancing lesions within the frontal gyrus, the anterior cingulate gyrus, and the parietal lobe. Incomplete resection led to recurrence in one patient, and later, intraoperative ultrasound was used to achieve total resection in another patient. Each patient was seizure free on follow-up, and managed with anti-epileptic medication. Resection is the only curative treatment in 85% of MA cases. Complete resection is necessary for symptomatic treatment in cases of MA, as recurrence has been documented in this lesion. Intraoperative ultrasound is an effective imaging modality to ensure gross total resection of MA [31].

Case reports

Roux et al., report the surgical management of a lesional drug resistant epilepsy caused by a meningioangiomatosis associated with a type IIIc focal cortical dysplasia located in the left supplementary motor area in a young male patient.

A first anatomically based partial surgical resection was performed on an 11-year-old under general anesthesia without intraoperative mapping, which allowed for postoperative seizure control (Engel IA) for 6 years. The patient then exhibited intractable right sensatory and aphasic focal onset seizures despite 2 appropriate antiepileptic drugs. A second functional-based surgical resection was performed using intraoperative corticosubcortical functional mapping with direct electrical stimulation under awake conditions. A complete surgical resection was performed, and a left partial supplementary motor area syndrome was observed. At 6 months postoperatively, the patient is seizure free (Engel IA) with an ongoing decrease in antiepileptic drug therapy.

Intraoperative functional brain mapping can be applied to preserve the brain function and networks around a meningioangiomatosis to facilitate the resection of potentially epileptogenic perilesional dysplastic cortex and to tailor the extent of resection to functional boundaries [32].

A case report of periventricular meningioangiomatosis associated with meningioma [33].

Free et al., from the Sanford Clinic, Sioux Falls, South Dakota reported two cases of sporadic meningioangiomatosis (MA) - a rare condition of the central nervous system known to cause headaches, seizures and other focal neurologic deficits. Both patients presented with headache and vision change, somewhat suggestive of migraine. The combination of magnetic resonance imaging (MRI) and computerized tomography (CT) can establish the diagnosis of MA [34].

Non-meningothelial meningeal tumours with meningioangiomatosis-like pattern of spread [35].

Jamil et al., report the first case, to their knowledge, of multifocal meningioangiomatosis in a child. This unique case highlights therapeutic challenges associated with these lesions and demonstrates that multifocality is possible in the pediatric population. This finding has implications for diagnosis and follow-up for children afflicted with these tumors [36].

A 37 year-old man with a long history of frontal headache. In suspected sinusitis, the patient underwent cerebral MRI that showed hypointense lesion in the right frontal lobe with heterogeneous contrast enhancement after gadolinium administration. There were no stigmata or family history of neurofibromatosis. A right pterional approach with a supraorbital craniotomy was performed. The lesion was removed with complete remission of the headache in the postoperative time. MA enters into differential diagnosis with several other diseases and a correct diagnosis is mandatory. The total surgical removal is the treatment of choice, and the prognosis after surgery is usually excellent for the absence of recurrence in sporadic cases [37].

MA has been reported infrequently in association with NF2. Omeis et al., report 2 cases of MA associated with NF2 in one family, and we add the cerebellum to possible locations of occurrence. MA should be considered in the differential diagnosis of cortical lesions, particularly in patients with NF2 [38].

Park et al., report the CT and MR findings in two patients with multifocal meningioangiomatosis, neither of whom had a family history or stigmata of neurofibromatosis. All lesions were located in the cortical and subcortical areas and had round dense calcifications with eccentric cysts. The masses were associated with surrounding edema and gliosis [39].

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