In conclusion, although considered off-label, in case of severe ERIS determining NDFI with no evidence of vascular lesions on CT and little DWI lesion volume, a second ST might be considered.

Francesco Brigo, Tommaso Bovi
University of Verona, Italy

Giampaolo Tomelleri, Paolo Bovi, Giuseppe Moretto
DAI di Neuroscienze e Istituto di Neurochirurgia
Verona, Italy

REFERENCES

TO THE EDITOR

Intracranial Non-Occlusive Thrombus and Multiple Strokes in Giant Cell Arteritis

A 78-year-old woman presented with facial pain, scalp tenderness, jaw claudication and visual loss in her left eye. Her visual acuity was 20/25 in the right eye and 20/40 in the left eye. She had a left relative afferent pupillary defect (RAPD) and a constricted visual field in the left eye. Fundoscopy was consistent with anterior ischemic optic neuropathy. She had an elevated ESR at 50mm/hour, elevated CRP at 101mg/L and elevated platelet count at 621/mm³. She received methylprednisolone 1 gram IV and had temporal artery biopsies, which were consistent with giant cell arteritis (GCA). She was treated with prednisone 60 mg po daily.

Five days later, she developed aphasia followed by right arm weakness four days later, with right extensor plantar response on examination. Magnetic resonance imaging (MRI) of the brain showed multiple small strokes of varying ages in the left cerebellar, left parieto-occipital, bilateral centrum semiouale and right superior frontal regions (Figure A). Computed tomography (CT) angiogram of the head and neck showed luminal irregularity of both extracranial vertebral and internal carotid (ICA) arteries, and an intraluminal filling defect of the petrous segment of the left ICA consistent with an intracranial non-occlusive thrombus (iNOT) (Figure B, C). These occurred despite prednisone treatment; therefore, she was treated with methylprednisolone 1 gram IV daily, aspirin and heparin IV for five days. Repeat CT angiogram three days later showed resolution of the left petrous ICA iNOT (Figure D). After completion of her intravenous therapy, she was switched to prednisone 80 mg daily, aspirin and clopidogrel. Echocardiogram and Holter monitor did not reveal any cardiac source of embolus. On discharge from hospital, she had stable deficits in her vision and improvement of her language and motor functioning.

DISCUSSION

Giant cell arteritis is a systemic vasculitis that affects medium and large-sized arteries. The pathology shows granulomatous inflammation of the inner media, multinucleated giant cells and fragmentation of the internal elastic lamina. This leads to segmental necrosis, luminal disruption and thrombotic occlusion of the affected vessels. Giant cell arteritis has a predilection for the vertebral arteries, but can also affect the anterior circulation.1

Figure: Radiologic findings. Axial diffusion weighted image (1A) shows multiple small acute infarcts in bilateral centrum semiovale. Initial CT angiogram source image shows an intraluminal filling defect (arrows) in the left cavernous ICA (1B). Curved planar reformat from the CTA (1C) shows multiple intraluminal filling defects (arrows). Follow-up CT angiogram (1D) performed three days later shows complete resolution of these filling defects suggesting intraluminal thrombi.
Ischemic strokes occur in 3-4% of patients with GCA, and the causes are multi-factorial. Inflammation of intra- and extracranial vasculature lead to intimal thickening, luminal irregularities, stenoses, and occlusions, which can cause borderzone hypoperfusion and infarction. In situ thrombosis of inflamed vessels can also result in occlusion and distal embolization.

It would be ideal to treat both inflammatory and thrombotic mechanisms. High-dose corticosteroids are the mainstay of therapy in GCA, but small observational studies have explored the addition of antithrombotics. Use of low-dose aspirin was associated with lower rates of visual loss and strokes. Another study looked at antithrombotics in patients with GCA and found they could be treated with baseline antiplatelet or anticoagulant agents, without an increased risk of bleeding. Several case reports have described mixed success with use of anticoagulation in patients with GCA, but none have documented the presence and resolution of iNOT while on anticoagulation. One study looked at iNOT on CT angiography in patients with acute ischemic events (non-arteritic) and found a non-significant trend to resolution of iNOT with dual or triple compared to single antithrombotic therapy.

Our patient developed new ischemic infarcts despite high dose corticosteroids. On neuroimaging, there was evidence of luminal irregularities as well as an iNOT. We used IV steroids and a limited course of IV heparin and aspirin to target inflammation and thrombosis. We observed the interval resolution of the iNOT after several days of combination therapy, without evidence of bleed. Due to limited prospective studies on use of antithrombotics in GCA, the risks and benefits of such therapy is unknown. In our patient, the small size of her strokes and the progression of disease despite treatment prompted a more aggressive approach. Well-designed prospective studies are necessary to explore combined therapy for treatment-refractive GCA.

To the Editor

Plaque-Type Blue Nevus with Meningeal Melanocytomas

Pigmented primary neoplasms of the central nervous system are uncommon. Differential diagnosis of such lesions is limited and includes pigmented meningioma, melanocytic schwannoma, and primary versus metastatic melanoma.

Primary melanocytic tumors of the meninges remain among some of the rarest neoplasms. Again, a variety of entities fall under such a category, including: pigmented meningioma, melanocytic schwannoma, pigmented primitive neural-ectodermal tumor, diffuse melanosis, and meningeal melanocytoma. Of interest, is the entity of purely melanocytic origin that arises from the dura, that being meningeal melanocytomas (MM).

Meningeal melanocytomas are scarcely found in the literature. First ever described by Virchow in 1959, Pigment und diffuse Melanose der Arachnoides, these pigmented dural based neoplasms were definitively classified with the help of electron microscopy. As of 2000, only about 110 cases were present in the literature. The WHO estimates a prevalence approaching one per 10 million, in the population.

The usual MIB index of these lesions is below 5%, making them typically a benign entity. Those MM’s with MIB indices in the intermediate range, 5-10%, are uncommon. In addition, melanocytic dermatological manifestations such as nevus of Ota, have been associated with ipsilateral intracranial MM, with only seven cases being described as of 2009. Even rarer is the association of ipsilateral benign blue nevus and MM, with only two cases previously being described in the literature.

Here we describe a new and rare case report of a patient with a plaque-type blue nevus, ipsilateral meningeal melanocytomas, and an intermediate pathological grade (MIB index of 5-10%).