Moya-Moya

Patient history

34-year old female with neurofibromatosis and Moya-Moya intracerebral arterial occlusive disease. Perfusion imaging was performed with 6 ml Gadavist at 4 ml/sec. MRA shows occlusion of the right PCA (yellow arrow) and left A1 segment (red) as well as a long segment significant stenosis of the right supraclinoid ICA (Figure 1) (blue arrow). Note the extensive collateralization (Moya-Moya) (black arrows). No acute stroke on MR images is observed but patchy ischemic changes throughout and a subtle old right occipital infarction are evident (Figure 2).

Initially, the left cerebellar lesion felt to represent a hamartoma, but it reveals hemorrhage, changing the diagnosis to infarction (Figure 2) (All 3).

Figure 1 MRA imaging

Figure 2 The right thalamic hamartoma (white arrow) consistent with neurofibromatosis FLAIR and T2-weighted cube FSE

Figure 3 Dynamic maps and ASL map. CBF and CBV maps within normal limits, TTP and TMAX are prolonged in right MCA and PCA territories. ACL shows failure in right PCA and delay in right MCA distribution
Post-Treatment and analysis

PWV images were processed using Olea Sphere® suite. Dynamic maps, such as CBV, CBF, Tmax, MTT, TTP were computed.

Imaging Findings

The CBF and CBV maps are within normal limits. However, TTP and Tmax are prolonged in right MCA and PCA territories. Note that ASL reveals tag failure in the right PCA and delay in the right MCA distribution.

Discussion

Perfusion imaging yields important insights as to the presence and extent of ischemia in cerebrovascular occlusive disease. This patient with neurofibromatosis and Moya-Moya has ischemia in the right MCA and PCA distributions that will eventually lead to a revascularization procedure.