Case Report

Rare co-occurrence of dural arteriovenous fistula and arteriovenous malformation with bilateral subcortical and basal ganglia calcification

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Abstract

The present study describes the imaging findings in a patient with dural arteriovenous fistula (AVR) and arteriovenous malformation (AVM) with bilateral subcortical and basal ganglia calcification.

A 29 year old male patient presented with chief complaint of recent onset of generalized tonic clonic seizures and mild disorientation. The imaging studies on MCT demonstrated diffuse, symmetric calcification in the bilateral basal ganglia and subcortical white matter. MR imaging and angiography revealed AVM in parieto-occipital region with supply predominantly from left posterior cerebral and middle cerebral arteries. Multiple dural feeders from meningeal branches of occipital and superficial temporal branches of bilateral external carotid and right internal carotid arteries. Calcification is proposed to be due to chronic reflux into the parenchymal veins or vascular steal phenomenon. This rare co-occurrence of subcortical calcification in a patient with a dural AVF and AVM is being reported.

Keywords: Dural arteriovenous fistula, Arteriovenous malformation, Subcortical calcification.

Introduction

Calcification of intracranial arteriovenous malformations (AVMs) are not uncommon, however these are usually limited to the vessel wall of AVM and occasionally to the surrounding gliotic parenchyma.[1,2] However calcification at distant site from the AVM which is extensive, bilateral and diffuse has been less often seen. Bilateral subcortical and basal ganglia calcification has been rarely reported in literature to be associated with dural areteriovenous fistula (AVF).[3,4] Dural AVFs are abnormal arteriovenous connections that are located within the dura mater and involve a dural sinus and/or cortical veins and constitute 10-15% of all intracranial arteriovenous shunts. The dural AVF have been found to be associated with chronic venous reflux which has been postulated to result in subcortical calcifications.[5] A case of characteristic calcification in the cortico-medullary junction at the bottom of cerebral sulci and basal ganglia in a patient with both AVM and dural AVF is reported.

Case Report

A 29-year-old male patient with history of mental retardation developed recent onset generalized tonic clonic seizures; he also became unconscious and was admitted in a public hospital where he regained consciousness after 3 to 4 days. His CT scan was performed which demonstrated bilateral subcortical and basal ganglia calcification. Multiple punctate areas were seen in right parietooccipital region raising the possibility of AVM. As CT or MR angiogram was not performed because of financial constraints, therapeutic option of angioembolization was planned directly. He was started on antiepileptics and sent to angiographic department of our hospital. Initial angiogram showed a large Grade-IV Spetzler AV malformation AV malformation in the left parieto-occipital region. The nidus measured 7.5 X 4 X 3.5 cm with supply predominantly from multiple feeders from hypertrophied left posterior cerebral artery and left middle cerebral artery. Multiple dural feeders were also seen arising from meningeal branches of occipital and superficial temporal branches of both external carotid and right internal carotid artery. Drainage was peripherally in to superior sagittal sinus. AVM was embolized progressively in three sessions.

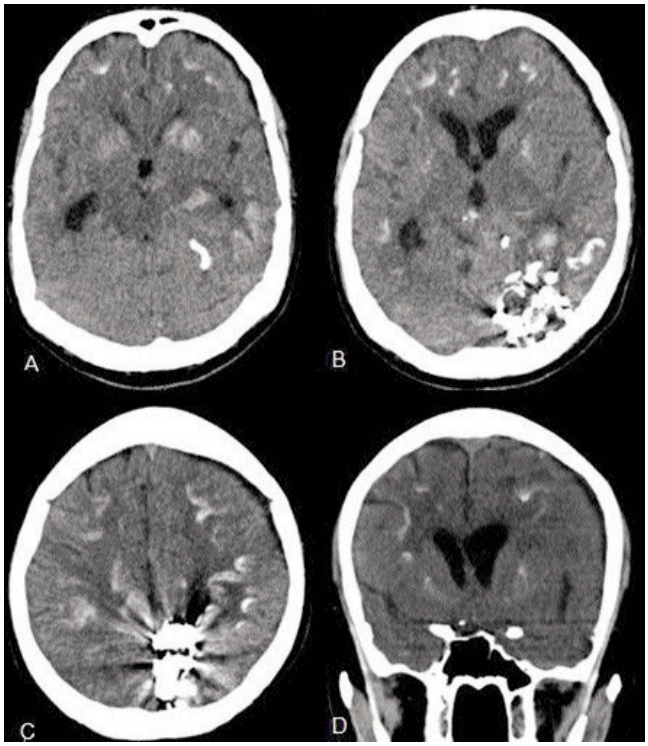

Figure-1: Axial and reconstructed coronal images of unenhanced CT study showing bilateral symmetrical diffuse subcortical and basal ganglia calcifications. High density in left parieto-occipital region is due to glue deposition from previous AVM embolization.

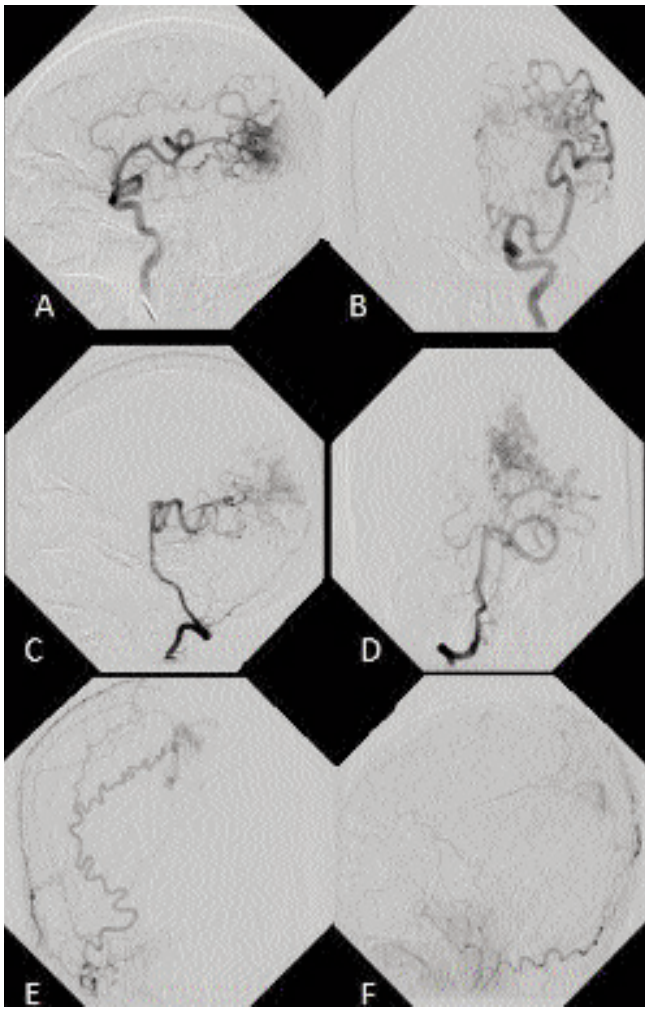

Figure-2: A,B- Left internal carotid angiogram AP and Lateral views demonstrating large feeder vessels from middle cerebral artery and C,D- Left vertebral angiogram AP and Lateral views demonstrating large feeder vessels E,F-.Dural feeders seen from meningeal branches of occipital and superficial temporal branches of external carotid arteries supplying the AVM.

Discussion

Intracranial calcifications may be physiological or pathological, diffuse or focal. Calcium deposition may normally be seen in the choroid plexus, pineal body, falx and Pacchionian bodies: many pathological conditions may also result in calcification.[6] Differential diagnoses of intraparenchymal-calcified lesions include metabolic disorders (hyperparathyroidism, lead poisoning), infectious disorders (cytomegalovirus infection, toxoplasmosis, tuberculomas), neoplasms (ependymoma, oligodendroglioma), and vascular malformation, including Sturge-Weber syndrome and pial arteriovenous malformation.[7]

Subcortical calcification is a nonspecific imaging finding and is commonly bilateral and symmetrical.

Frequent causes include Sturge-Weber syndrome,[8] tuberous sclerosis, Fahr disease,[8] post chemoradiotherapy change, and metabolic disorders secondary to parathyroid or thyroid gland abnormalities. Similar calcification changes in corpus striatum and dentate nucleus are also commonly seen in these disorders.

Although there have been few reports in literature describing association of subcortical calcification with dural AVF,[9] the exact mechanism leading to calcification is poorly understood. Metoki et al[5] reported 3 cases of dAVF with corticovenous reflux presenting with characteristic calcification in the cortico-medullary junction. They believed that in their patients focal hypoperfusion due to steal phenomenon resulted in dystrophic subcortical calcification. In a similar case report, Lai et al[3] postulated calcification was due to persistent venous congestion or steal phenomenon.

Chen et al[10] reported cerebral and basal ganglia calcification in a child with bilateral sigmoid sinus atresia resulting in anomalous intracranial venous drainage. CT scan demonstrated that a vascular scalp mass filling through the emissary foramen.The reason of calcification was again due to steal phenomenon. The reports by Yu et al[1] also support the postulation that parenchymal ischaemia due to steal of blood leads to calcification in patients with AVMs.

As reported by Yang et al,[4] MR imaging including the angiogram and venogram may be useful to determine the arterial supply as well as venous drainage.

In our patient the drainage was however into the venous sinuses and proposed venous congestion could not be demonstrated. This type of co -occurrence of dural AVF and AVM with subcortical and basal ganglia calcification has not be reported to best of our knowledge and further literature review and correlation with other imaging modalities as Iodine 123-labeled N-isopropyl-p-iodoamphetamine (123I-IMP) and single-photon emission tomography (SPECT) as utilized by T. Metoki el al may be helpful to determine the exact reason for the calcification.

References

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