Dural AV Fistula Presenting as Enhancing Tumor like Lesion

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Abstract :

A case of dural AV Fistula with atypical clinical and imaging presentation is being presented here. A 76 years old male patient presented with headache for about 4 days with limb weakness and unsteadiness. A CT raised the possibility of cerebellar infarct with hemorrhagic transformation or a left cerebellar mass. Presence of dilated tortuous vessels in the vicinity of lesion raised the possibility of Dural AV fistula, which was later confirmed by a catheter angiogram. Endovascular embolization was performed and complete obliteration of fistula was achieved.

Keywords: Dural AV fistula, MRI

Introduction :

The adult type intracranial dural arterio-venous fistulae (DAVFs) are rare lesions occurring in 0.16 per 100 000 adults per year. [1] These can be located anywhere in the skull, along the dura. Most commonly, these are the results of cerebral venous thrombosis. [2, 3] There is subsequent collateral revascularization, which permits an abnormal fistulous connection to form between arteries and veins residing in the walls of a dural sinus or involving an adjacent cortical vein. This can be demonstrated by catheter angiography showing multiple arterial feeders draining into single draining vein, which has early filling compared to other veins in the brain. In this article, we describe a rare case of intracranial DAVF that manifested as an enhancing lesion in the cerebellum, giving the misleading appearance of a neoplasm. The DAVF was confirmed with catheter angiography and treated with endovascular embolization.

Case Report :

A 76 years old male patient presented at our hospital with history of loss of consciousness and weakness of limbs. Initially CT of head with and without contrast showed mass effect within the left cerebellum and two foci of hemorrhage. There was also non specific enhancement in the lesion. Brain MRI re-demonstrated abnormal signal intensity lesion involving the left cerebellar hemisphere with the foci of small hemorrhages within the lesion causing mass effect in fourth ventricle. Post contrast images revealed enhancing lesion. (Fig.1) The differential varied from sub acute infarct to the primary neoplasm of the brain. In addition, there were several tortuous veins in posterior fossa (Fig.2) which raised a concern for an underlying dural AV fistula. Contrast CT revealed the same findings alone with heterogeneous enhancement-giving a tumor like appearance. Catheter angiography was performed and revealed enlarged posterior meningeal branch of the right vertebral artery feeding a dural fistula. The tangle of the blood vessels was present, which subsequently drained into the posterior fossa via an early filling vein, extending inferiorly and anteriorly. Subsequently it extended more cranially and then likely into the transverse sinus.

Figure 1: Plain non contrast axial CT – showing a hypodense lesion in the left cerebellum with foci of hemorrhages.

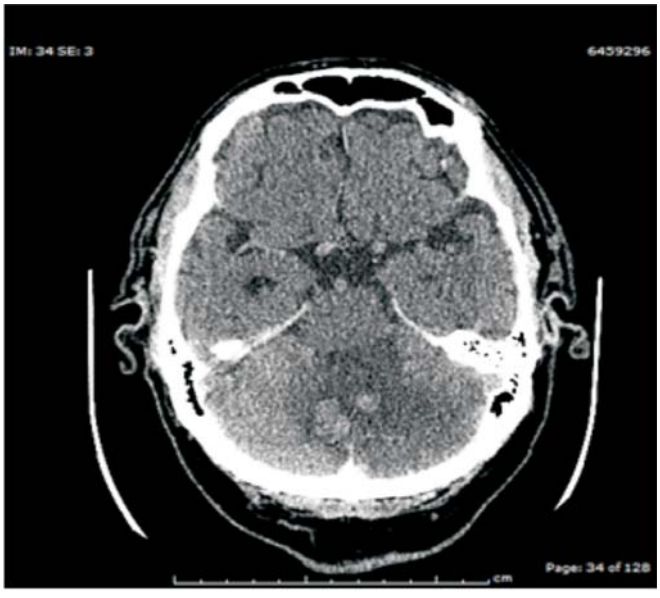

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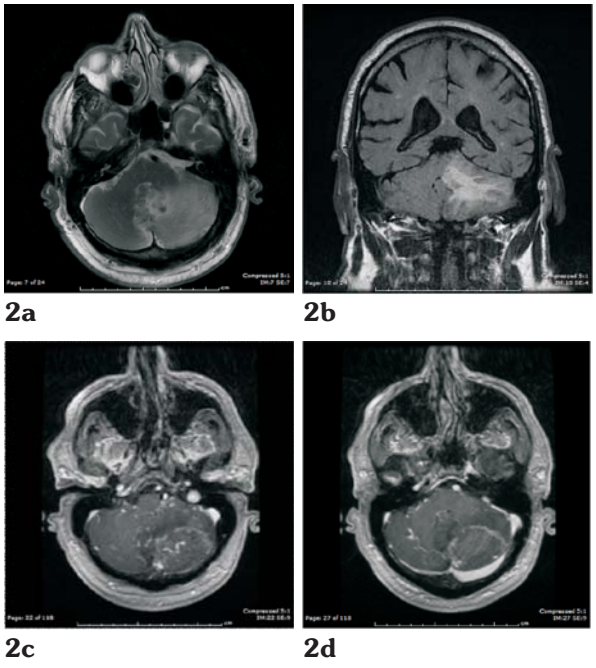
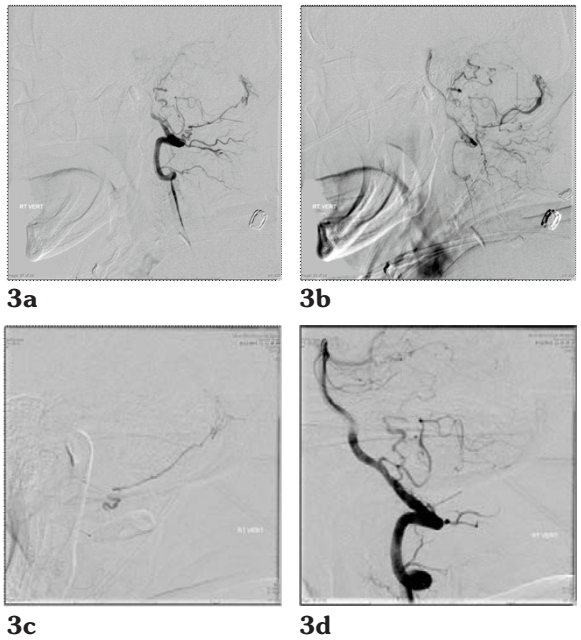
Figure 2: Axial T2(2a) and coronal STIR(2b) image depicting a left cerebellar hyperintense lesion; the possibility of a left cerebellar mass could not be ruled out. MRI Brain with GD-(2c and 2d) enhancing lesion along with multiple dilated tortuous veins in periphery- findings raise the possibility of Dural AVF, although tumor cannot be certainly ruled out.



2a **2b**

2c **2d**

Figure 3: (3a) Cerebral Angiography in the right vertebral artery showing enlarged posterior meningeal branch of the right vertebral artery which was seen extending along the floor of the posterior fossa and feeding a dural fistula. (3b) Early filling vein is visualized which subsequently extends likely into transverse sinus and dilated large tortuous veins. (3c and 3d- Embolization of fistula) (3c) Selective catheterization of the dorsal meningeal artery was done and (3d) complete embolization was achieved as shown by the stump in the figure.



3a **3b**

3c **3d**

Discussion :

Dural AVFs are rare vascular lesions, which can be found anywhere in the dura along the calvaria. Possible causes are trauma, previous craniotomy and idiopathic. However, most commonly, they are the result of cerebral venous thrombosis (typically transverse venous sinus thrombosis). [2 3] There is subsequent collateral revascularization with formation of innumerable small microfistulae connecting the arteries with the veins residing in the walls of a dural sinus or involving an adjacent cortical vein. Arterial supply is from the same branches that supply the meningeal arteries. According to various regions in the brain the arterial feeders are as follows: [10]

Supratentorial region: Middle meningeal artery (ECA)

Anterior cranial fossa: ethmoidal branches of the ophthalmic artery (ECA)

Posterior cranial fossa: dural branches from the vertebral arteries, branches from occipital and ascending pharyngeal arteries

Cavernous sinus: dural branches from the ICA and accessory meningeal branch of the maxillary artery (via foramen ovale) - branch of ECA.

The posterior fossa lesion in our case was being supplied by the dural branches of the right vertebral artery.

The typical clinical presentation depends upon the location - which may include tinnitus (most common- due to most common involvement of transverse sinus), orbital symptoms (retroocular pain, headaches, pulsatile proptosis and chemosis in caroticocavernous fistula), symptoms of venous hypertension (headache, vomiting, papilloedema and focal neurological deficits) may be seen. [4,7]

Cross sectional imaging is not always successful in picking up a dural AVF. Plain CT may not show any changes. Dilated trans-osseous feeding vessels can often been seen in these patients. This finding can also be observed on MRI images. CTA and MRA are more helpful in predicting dural AVFs. Indirect visualization by observing the presence of multiple tortuous vessels can help in picking up a dural AVF. Most sensitive and specific study study is DSA. DSA can help to identify retrograde venous flow or cortical venous drainage, presence of either of which can put the AVF into a higher Grade. [4, 5, 6, 7]

Grading of Dural AVF can be done by two systems.

Grading of dAVFs [4, 5, 8]

1. Cognard Classification [9]

Grade 1 : In sinus wall; normal antegrade venous drainage (low risk; benign clinical course)

Grade 2A: In sinus; reflux to sinus, not cortical veins

Grade2B: Reflux (retrograde drainage) into cortical veins (10-20% hemorrhage)

Grade 3: Direct cortical venous drainage; no venous ectasia (40% hemorrhages)

Grade 4: Direct cortical venous drainage + venous ectasia (65% hemorrhages)

Grade 5: Spinal perimedullary venous drainage

2. Borden Classification

Type I: Dural arterial supply with antegrade drainage into venous sinus

Type Ia: Simple dAVF with single meningeal arterial supply

Type Ib: Complex dAVF with multiple meningeal arteries

Type II: Dural supply + ↑ intrasinus pressure → antegrade sinus, retrograde cortical venous drainage

Type III: Dural arteries drain into cortical veins

Both classifications revolve around the knowledge that venous drainage pattern correlates with increasingly aggressive neurological clinical course. The Borden classification is a simplified version of the Cognard system, but loses in granularity. [4] The presentation of our patient was radiologically misleading. The initial CT and MRI findings were most consistent with neoplastic process such as lymphoma, or a hemorrhagic transformation of an infarction. The diagnosis of vascular anomaly was suspected due to presence of dilated tortuous vessels around the area of hemorrhage. Prognosis of the AVF generally depends on the location and the grade of the AVF. More than 95 percent of AVFs have a benign course. Those with reflux or cortical venous drainage have a malignant course. These aggressive dAVFs can cause hemorrhage and recurrent neurological symptoms. [4, 6, 7] This patient's AVF was later embolized using TruFill mixture and complete embolization was achieved. As of today, the patient is doing well and following up on an outpatient basis.

References :

1. Al-Shahi R, Bhattacharya JJ, Currie DG, Papanastassiou V, Ritchie V, Roberts RC, Sellar RJ, Warlow CP; Scottish Intracranial Vascular Malformation Study Collaborators. Prospective, population-based detection of intracranial vascular malformations in adults: the Scottish Intracranial Vascular Malformation Study (SIVMS). Stroke. 2003 May;34(5):1163-9. Epub 2003 Apr 17. PubMed PMID: 12702837.
2. Mullan S. Reflections upon the nature and management of intracranial and intraspinal vascular malformations and fistulae. J Neurosurg. 1994 Apr;80(4):606-16. PubMed PMID: 8151338.
3. Terada T, Higashida RT, Halbach VV, Dowd CF, Tsuura M, Komai N, Wilson CB, Hieshima GB. Development of acquired arteriovenous fistulas in rats due to venous hypertension. J Neurosurg. 1994 May;80(5):884-9. PubMed PMID: 8169629.
4. Gandhi D, Chen J, Pearl M, Huang J, Gemmete JJ, Kathuria S. Intracranial dural arteriovenous fistulas: classification, imaging findings, and treatment. AJNR Am J Neuroradiol. 2012 Jun;33(6):1007-13. doi: 10.3174/ajnr.A2798. Epub 2012 Jan 12. Review. PubMed PMID: 22241393.
5. Gomez J, Amin AG, Gregg L, Gailloud P. Classification schemes of cranial dural arteriovenous fistulas. Neurosurg Clin N Am. 2012 Jan;23(1):55-62. doi: 10.1016/j.nec.2011.09.003. Review. PubMed PMID: 22107858.
6. Gross BA, Ropper AE, Du R. Cerebral dural arteriovenous fistulas and aneurysms. Neurosurg Focus. 2012 May;32 (5):E2. doi: 10.3171/2011.12.FOCUS11336. PubMed PMID: 22537128.
7. S.Y. Ha, Y.S. Kwon, B.M. Kim, D.I. Kim, D.J. Kim. Clinical and angiographic characteristics of multiple dural arteriovenous shunts. AJNR Am J Neuroradiol 33:1691–95 (1691-95).
8. Geibprasert S, Pereira V, Krings T, Jiarakongmun P, Toulgoat F, Pongpech S, Lasjaunias P. Dural arteriovenous shunts: a new classification of craniospinal epidural venous anatomical bases and clinical correlations. Stroke. 2008 Oct;39(10):2783-94. doi: 10.1161/STROKEAHA. 108.516757. Epub 2008 Jul 17. PubMed PMID: 18635840.
9. Cognard C, Gobin YP, Pierot L, Bailly AL, Houdart E, Casasco A, Chiras J, Merland JJ. Cerebral dural arteriovenous fistulas: clinical and angiographic correlation with a revised classification of venous drainage. Radiology. 1995 Mar; 194(3):671-80. PubMed PMID: 7862961.
10. Dural AV Fistula Dr. Henry Knipe and Dr. Frank Gaillard et al http://radiopaedia.org/articles/dural-arteriovenous-fistula