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Case Report

Dural Arteriovenous Fistula Presenting with Exophthalmos, Hemiparesis and Seizures: Resolution after Endovascular Treatment - Case Report

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Abstract

A concomitant focal seizure with left hemipaeresis and exophthalmos in the context of an aggressive cavernous dural arteriovenous fistula (dAVF) is a variable neurological presentation. Here, we report a 73 years old female who presented with a 5-months history of progressive painless exophthalmos with conjunctival chemosis of right eye without any visual impairment. But now she had admitted to the hospital due to new onset of focal seizures with mild left hemipaeresis. Emergency MDCT scan brain showed suspected small malignant dural ateriovenous fistula of right sphenoparietal sinus with cerebral venous congestion and tiny hemorrhagic venous infarction at right operculum and right exophthalmos. Cerebral angiography demonstrated right cavernous dAVF fed by branches from the right ascending pharyngeal artery and right middle meningeal artery, right accessory meningeal artery, bilateral arteries of foramen rotendum, dural branch of left vertebral artery with drainage to the right superior ophthalmic vein, right sphenoparietal sinus and right frontal cortical vein which was consistent with Cognard Type IIb dAVF. Successful transvenous embolization of the right cavernous via the right inferior petrosal sinus was done by using fibre coils and NBCA. Post- embolization control angiogram showed complete obliteration of right cavernous dAVF feeding vessels. Subsequent clinical follow up revealed that she had dramatic improvement of proptosis, chemosis and neurological symptoms along with cessation of seizures.

Keywords: *Dural Arteriovenous Fistula; Exophthalmos; Hemipaeresis; Seizures; Endovascular Treatment*

Abbreviations

dAVF:DuralArteriovenous Fistula; OPD: Outpaient Department; NBCA: N-butyl-2-cyanoacrylate

Introduction

Intracranial dural arteriovenous fistulas (dAVF) are pathologic shunt situated between dural arteries and dural venous sinuses, meningeal veins or an adjacent cortical vein which can have profound detrimental effects on the underlying brain and cranial nerves [1]. It may also present in the spinal compartments of the central nervous system [2]. In intracranial, most frequently dAVF are affecting the region of the transverse and cavernous sinuses but uncommonly involve the dura of the craniocervical junction [3]. Intracranial dAVF account for 10-15% of all intracranial artriovenous lesions [4]. The incidence of dAVF at various locations as reported in literature is as follows: transverse sinus 50%, cavernous sinus 16%, tentorium cerebelli 12%, and superior sagittal sinus, 8% [5]. Dural AVF can affect a variety of cerebral venous structures, which can present at various clinical stages, ranging from a simple irritat-

ing pulse-synchronous bruit, tinnitus to a disabling neurological deficit as a result of ischaemia from impaired venous drainage to life threatening intracranial hemorrhage from venous hypertension [5,6]. In our case report on emphasize on the neurological presentations in intracranial dAVF along with treatment.

Case Report

A 73 years old hypertensive and diabetic female was attended to the outpatient department (OPD) with the history of left hemipaeresis with focal seizure on the left upper limb without generalization for 1 week. But initially she noticed that her right eye was congested and gradually painless proptosed without any visual impairment. She had no complaints of tremulous hand, weight loss, palpitation, thyromegaly, fever or head trauma for last 5 months. Despite of initial symptoms she was not consulted to any physicians or even the ophthalmologist within that time. After appearing of neurological features she was admitted into a tertiary level hospital and from their sent to our institute. Her general physical examination was revealed normal and neurological examination was found well orientated, upper motor neuron lesion in right side (MRC Grade 4) without any cranial nerve involvement. Laboratory investigations including thyroid, renal and liver function tests were normal. Emergency MDCT scan brain showed suspected small aggressive dAVF of right sphenoparietal sinus with cerebral venous congestion and tiny hemorrhagic venous infarction at right operculum and right exophthalmos (Figure 1). With above clinical findings along with neurological involvements and suspicious aggressive dAVF lesion in MDCT so we went for specific cerebral angiography inspite of Magnetic resonance imaging (MRI) and MR angiography (MRA).

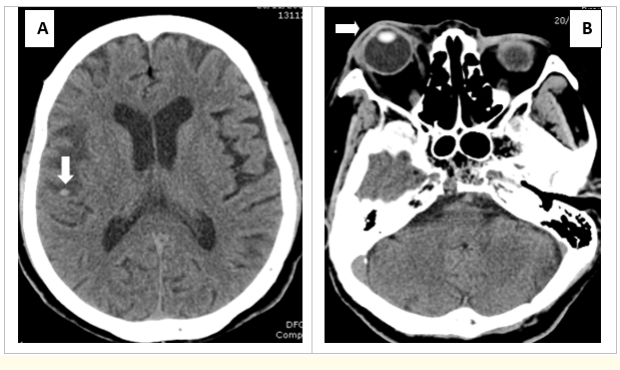

Figure 1: MDCT showed suspected small aggressive dAVF of right sphenoparietal sinus with cerebral venous congestion and tiny hemorrhagic venous infarction at right operculum (Figure: A-arrow head) and right proptosis (Figure: B -arrow head).

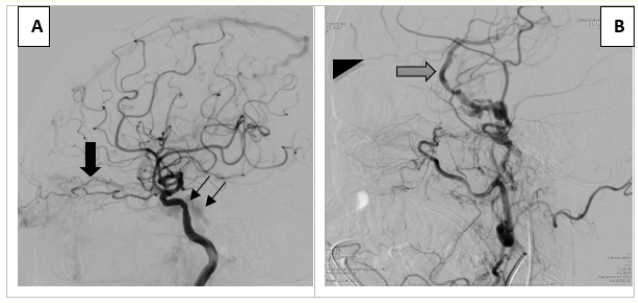

Figure 2: (A+B) Cerebral angiography revealed right cavernous dAVF (double arrow) with drainage to the right superior ophthalmic vein (bold black arrow), right sphenoparietal sinus (bold white arrow) and right frontal cortical vein (black arrow head).

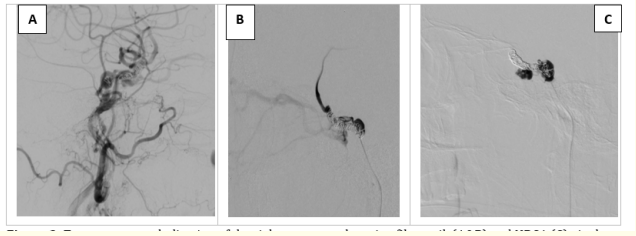

Figure 3: Transvenous embolization of the right cavernous by using fibre coils(A&B) and NBCA (C) via the right inferior petrosal sinus.

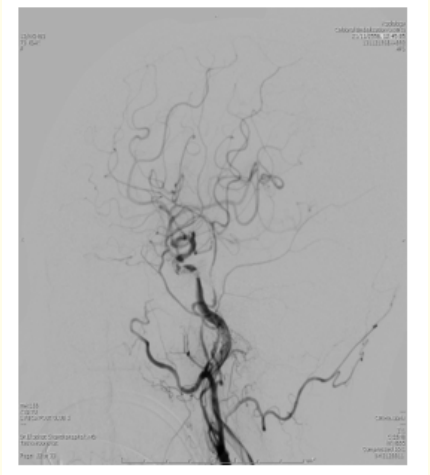

Figure 4: Post-embolization control angiogram shows complete obliteration of right cavernous dAVF feeding vessels.

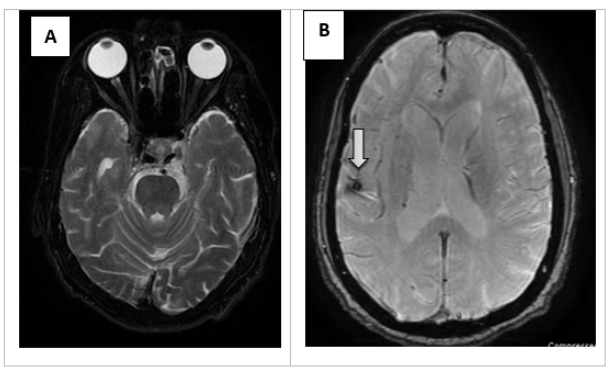

Figure 5: Follow up MRI and MRA after 3 months of post embolization found tranavenous embolization coils and glue without residual dAVF with resolution of venous congestion at anterior right temporal lobe and right frontal operculum without proptosis (A), leaving only a few tiny hemorrhagic spots and superficial siderosis (arrow head-B).

Cerebral angiography revealed right cavernous dAVF which was fed by branches from the right ascending pharyngeal artery and right middle meningeal artery, right accessory meningeal artery, bilateral arteries of foramen rotendum, dural branch of left vertebral artery with drainage to the right superior ophthalmic vein, right sphenoparietal sinus and right frontal cortical vein (Figure 2) which was consistent with Cognard Type IIbdAVF.

So we planned for embolization immediately as this was aggressive dAVF. Successful transvenous embolization of the right cavernous via the right inferior petrosal sinus was done by using fibre coils(Nester) initially but there was no complete obliteration of dAVF (Figure 3A and 3B). Then we further embolized by 30%NBCA mixing with lipiodol (Figure 3C) and post-embolization control angiogram showed complete obliteration of right cavernous dAVF feeding vessels (Figure 4). Her postoperative course was uneventful and neurologic deficits gradually improved. She underwent aggressive rehabilitation for 0.232 weeks after embolization and was conversant and ambulatory with minimal assistance at discharge with significant improvement of neurological symptoms. She had advised to attend the clinic for follow-up examinations 1 month, 3 months, and 5 months after endovascular treatment. At her 3-months follow-up we had done MRI and MRA and found tranavenous embolization coils and glue and no evidence of aforementioned residual dAVF with resolution of venous congestion at anterior right temporal lobe and right frontal operculum without proptosis (Figure 5A), leaving only a few tiny hemorrhagic spots and superficial siderosis (arrow head -Figure 5B). Subsequent clinical follow up revealed that she had dramatic improvement of proptosis, chemosis and neurological symptoms along with cessation of seizures.

Discussion

Approximately 10 to 15% of all intracranial arteriovenous malformations consist of adult-type DAVFs [7]. There is no correlation of dAVF between age, sex and frequency of aggressive neurologic symptoms [8,9]. The incidence of dAVFis a higher in women and mean age is between 40 and 60 years [10,11] but dAVFs can occur at any age. In our reported study showed that the woman was 73years which was similar to the previous studies.

Although the mechanism of underlying the formation of fistulas is not yet fully understood but there are two hypotheses. Fistulas are thought to develop when normal physiologic pathways become pathologic arteriovenous shunts, or they are attributed to the release of other angiogenic factors that cause the shunts to develop [12]. The dAVFs are usually considered to be acquired lesions. Trauma is a well-known cause of acute onset dAVF attributed to damage of nearby dural arteries and veins. The DAVFs also have been reported by subacutely after trauma, infection and surgery. Thrombosis of a vein is also a common feature of dAVFs [13]. Our patient had no past history of trauma, infection, or surgery.

A wide spectrum of symptoms exists, ranging from the benign to the more aggressive; depend on the specific location of the lesion, extent of arterial supply and specific pattern of venous drainage. Drainage of a petrous region dAVF to the transverse or sigmoid sinus commonly produces pulsatile tinnitus; sometimes in association with an audible bruit [7]. Ocular manifestations (e.g.; opthalmoplagia, proptosis, chemosis, decreased visual acuity, bilateral papilloedema and atrophic optic disc) are commonly seen with carotid-cavernous dAVF [13,15]. Our reported case symptoms' began with ocular manifestations those were matched with the fistula in carotid-cavernous site.

More aggressive dAVF may manifest as focal neurological deficits, seizure, headache, progressive dementia-type of syndrome or cerebral haemorrhage, including subarachnoid, subdural or intraparenchymal bleeds and also brain edema and/or ischemia due to local venous congestion [13,16-18]. Such features are usually considered to be due to venous hypertension, although neurological deficits may be secondary to arterial steal [13]. In a meta-analysis of 360 dAVFs the tentorial incisura was the most ominous location, with 31 out of 32cases associated with haemorrhagic or non

haemorrhagic stroke [13,16]. So the pattern of venous drainage was considered of paramount importance in predicting aggressive behavior. This case study had the manifestation of neurological symptoms due to long standing presentation as a result of venous hypertension.

CT, MRI and cerebral angiography all have roles to play in the investigation of patients with a possible DAVF. The differentiation of high- and low-risk DAVFs is based on venous drainage patterns: Increased cortical venous reflux is consistent with a more aggressive course [10,19]. Because the clinical and imaging features can be non-specific, the diagnosis of a DAVF is often delayed or missed, but for detection of these fistulas, cerebral angiography is the method of choice, allowing the dynamic assessment of cerebral circulation [20]. Benign diseases, without cortical venous reflux that can be missed using both CT and MRI, if there is a strong clinical suspicion of a fistula. If haemorrhage is suspected, non-enhanced CT is a pre-requisite. Venous congestion may appear as an area of low density on CT. Our studied case had showed in the Multi-detector CT (MDCT) with suspected small aggressive dAVF of right sphenoparietal sinus with cerebral venous congestion and tiny hemorrhagic venous infarction at right operculum and right exophthalmos.

In the most institutions CT is more readily available and cheaper than MRI and so becomes the first-line investigation of patients presenting with tinnitus, headache or other vague neurological symptoms. MDCT angiography (MDCTA) can now provide high resolution detail of vascular anatomy [9]. T2 weighted MRI is more sensitive to the white matter changes of venous congestion or infarction when compared to CT. It has the drawback of being less sensitive to the changes of acute haemorrhage. If dilated cortical veins are present they may be seen on conventional spin echo sequences and visualized using MR angiographic techniques such as phase contrast venography or contrast enhanced MR angiography [9,21]. Time-of flight MRA (TOF MRA) is now widely available test for primary screening tool for when DAVF is suspected [22]. However, studies have reported that CTA has reduced sensitivity versus MRA for the detection of dAVFs (15.4% versus 50%) [23]. Cerebral angiography is the gold standard for detection of cerebral dAVF [24]. So we studied cerebral angiography and found right cavernous dAVF which was fed by branches from the right ascending pharyngeal artery and right middle meningeal artery, right accessory meningeal artery, bilateral arteries of foramen rotendum, dural branch of left vertebral artery with drainage to the right superior ophthalmic vein, right sphenoparietal sinus and right frontal cortical vein which was consistent with Cognard Type IIb classification.

Treatment is dependent on the clinical picture, temporal progression and the grade of fistula [25]. High risk radiographic features, such as cortically diverted venous drainage, patient with visual loss, hemorrhage or infarction require prompt therapy. A multidisciplinary approach involving an interventional neurologist, neurosurgeon and neuroradiologist is required. The goal of treatment of high risk fistulas should be complete obliteration. Treatment modalities include conservative management, endovascular intervention (i.e. transvenous transarterial embolization), microsurgery, and stereotactic radiosurgery [17]. Although endovascular embolization has evolved to become the first line treatment option for most dAVFs, it can be limited by inadequate access to the fistula point, non-target embolization, and the potential for recanalization [26]. In these instances, definitive cure can be achieved by surgical interruption of leptomeningeal venous drainage [26].

The low risk fistulas can be treated conservatively. Transarterial embolization (TAE) of feeding artery through external carotid branches with particle can easily performed, which can reduce shunt flow. TAE with n-butyl-2-cyanoacrylate has been applied to complex dural AVFs that are not accessible with percutaneous transvenous catheterization. However, complete cures are difficult from this method because of the existence of feeding arteries that cannot be catheterized and the recruitment of a blood supply from collateral arteries [27]. Therefore, this method is generally used to relieve symptoms or in combination with other procedures such as transvenous embolization (TVE), irradiation or surgery [7,28].

TVE with coils must be considered in patients with fistulas of the transverse and sigmoid sinuses. For curative purposes TVE with coils is used and many studies have reported it to be very useful (complete occlusion in 80%-100% of cases) [7,29]. But this procedure sometimes associated with, serious complications like; vessel injury and intracranial haemorrhage have also been reported [30]. Inadequate embolization leads to a worsening of symptoms. Critical assessment of diagnostic images and clinical conditions is also important for successful procedures. Previously this procedure was performed by direct surgical or transfemoral introduction of thrombogenic material into fistulae [31].

The use of detachable coils before liquid adhesives (onyx or glue) injection slows and decreases flow in the fistula and provides secure anchoring to the onyx or glue cast [32]. Other options such as surgical approaches and a combination of TAE and radiosurgery should also be considered when treating complex dAVFs [33]. Our case report demonstrated the aggressive dAVF, the presence of neu-

rological features with cortical venous reflux. Therefore our goal of treatment was to complete obliteration of fistula. We planned initially transvenous coil embolization with fibre coils (nester) of the right cavernous sinus via the right inferior petrosal sinus. But after embolization of the right cavernous sinus there was still presence of reflux in the right sphenoparietal sinus. So we further embolized by 30% NBCA mixing with lipiodol successfully and found complete obliteration dAVF in post-control angiography. Follow up MRI and MRA after 3 months of post embolization found tranavenous embolization coils and glue without residual dAVF with resolution of venous congestion at anterior right temporal lobe and right frontal operculum without proptosis (Figure 5A), leaving only a few tiny hemorrhagic spots and superficial siderosis (arrow head- Figure 5B). Subsequent clinical follow up revealed that she had dramatic improvement of proptosis, chemosis and neurological symptoms along with cessation of seizures.

Conclusion

Prognosis of high grade dAVFs greatly depends on early diagnosis and treatment. Significant advancements in the management of dAVFs have been made due to the development of recent pre- and intraoperative imaging methods with endovascular techniques. So the aggressive dAVFs need treatment immediately to prevent the further dreadful complications.

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