CHALLENGES IN THE MANAGEMENT OF CAROTID CARVENOUS FISTULA IN GHANA; THE WAY FORWARD

[1,2]Awoonor-Williams, R., [2]Vowotor, K. R., [2] Frimpong, A. A., [2]Nketiah-Boakye, F., [2]Ampong, A., [2]Kwarteng, J., [2]Amankwah, P., [2]Leat, M.

[1]Department of Anatomy, School of Medicine and Dentistry, KNUST, Kumasi, Ghana

[2] Komfo Anokye Teaching Hospital, Kumasi, Ghana

INTRODUCTION

Carotid cavernous fistula (CCF) is an abnormal communication between two intracranial vascular structures namely; the carotid artery and the cavernous sinus [1,2] . Various configuration of the fistula can occur depending on which part is involved. The two main branches of the carotid artery or any of their branches can communicate with the cavernous sinus.[3] These fistulae are classified for the purpose of determining management modalities as well as prognosis. The classification takes into consideration the hemodynamic status, anatomical and etiological basis of the fistula.[2]

The haeomdynamic nature of the fistula can be low or high flow. Anatomically they are classified as direct or indirect and etiologically as spontaneous or traumatic.[1,2] Direct carotid cavernous fistula occurs as a result of an abnormal communication between the internal carotid artery and the cavernous sinus. Indirect carotid cavernous fistula occurs between the meningeal branches of internal carotid artery and external carotid artery and the cavernous sinus.[3-5]

On the basis of etiology, traumatic carotid cavernous fistula are by far commoner than spontaneous accounting for about seventy five percent of all cases.[6] Spontaneous carotid cavernous fistula is common in the elderly and female patients. They account for about a third of all carotid cavernous fistula.[1,7,8,9] We report a case of spontaneous Carotid cavernous fistula in a forty year old postpartum female and the challenges in the management of the case.

CASE REPORT

A forty year old woman was referred to the outpatient neurosurgical clinic from ophthalmology with a month's history of gradual protruding left eyeball and headache. She delivered via spontaneous vaginal delivery days prior to noticing the onset of the protrusion. The headache was hemi global on the left side, throbbing in nature with no relieving or aggravating factors. This had progressively worsened associated with pain and blurring of vision in the left eye. There was no history of trauma prior to the onset of the protrusion. She had a history of pregnancy induced hypertension during the last month of the pregnancy. Post-delivery, the Blood pressure dropped to normal range and was under monitoring at the postnatal clinic. The past medical history was negative for Diabetes or any connective tissue disease. Her first delivery was via spontaneous vaginal delivery with an uneventful puerperium with no history of hypertensive disorder. The second was via c/s on account prolonged second stage. On examination she was fully conscious. Vision was 6/6 (20/20) in the right eye, and 6/9 (20/30) in the left eye. Anterior segment examination of the right eye yielded normal findings. On examination of the left eye, there was proptosis of the left eyeball, with chemosis of the conjunctiva. Cornea was clear, anterior segment was quiet and deep, with no cells or flares, pupil was reactive, and the lens was clear. Examination of the fundus of both eyes yielded no abnormal findings. There was no ophthalmoplegia. Intraocular pressures were 16mmHg in the right eye, and 42mmHg (markedly raised) in the left eye. A bruit was auscultated over the left orbit. Her blood pressure was 150/90mmHg. All other neurological examination was essentially normal. She had no features of connective tissue disease. CT angiogram that was done showed an enlarged left carotid sinus and left superior ophthalmic vein and dilated left meningeal vessel in the left temporal fossa connecting to the left middle cerebral artery, a type D carotid carvenous fistula. There was no evidence of subarachnoid haemorrhage. Assessment by the ophthalmologist prior to referral revealed her headaches and vision of the left eye worsened from 6/9 (20/30) to 6/18 (20/60) over a three week period from the time of diagnosis and development of ophthalmoplegia. The patient was counseled for digital subtraction angiogram with trans-arterial embolization and referred abroad for the service.

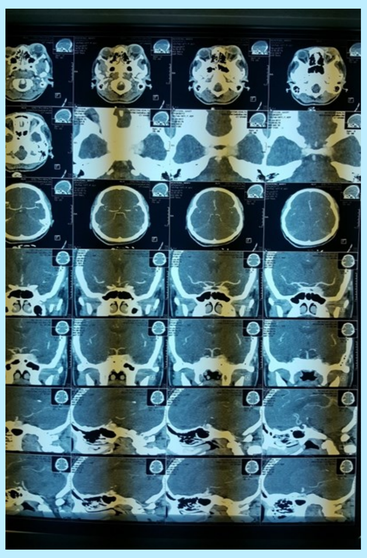


Fig 1a: Contrast enhanced CT scan of brain parenchyma and vasculature

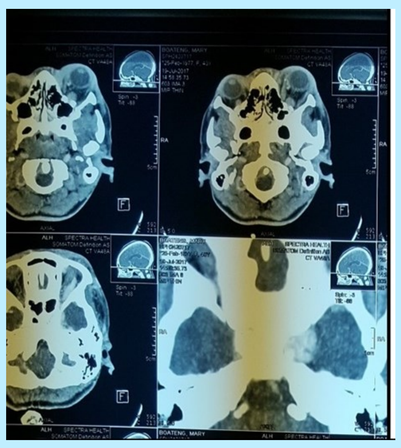


Fig 1b: Axial view of CT scan showing proptosis of left eye ball

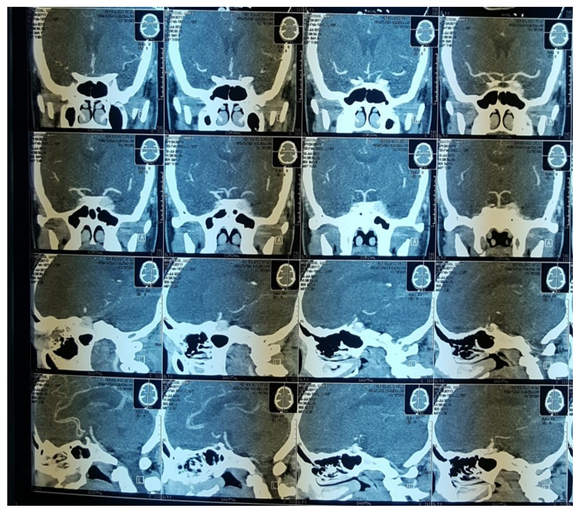
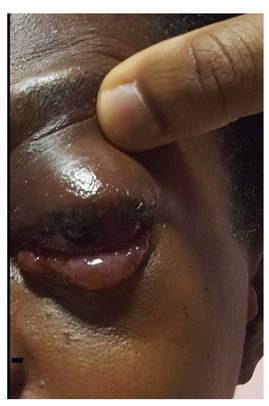


Fig 1c: Sagittal and coronal views of brain window showing left dilated cavernous sinus, as well as dilated left superior ophthalmic vein



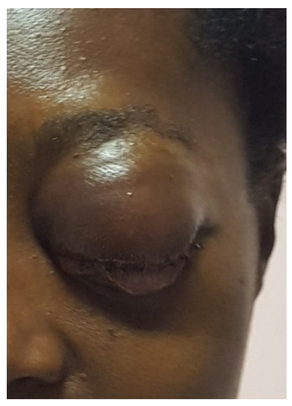


Fig 2a: showing chemosis of left conjunctiva Fig 2b: showing proptosis of left eye ball

DISCUSSION

The Barrow *et al.,* (1985) classifies carotid cavernous fistula (CCF) into four types, A to D. This anatomic classification provide essential information on the treatment options.[1,3]

Type A fistula is defined as a connection or communication between the intracavernous part of the internal carotid artery and the cavernous sinus. They are high flow in nature and are direct.[2,3] Type B is an abnormal connection between meningeal branches of the internal carotid artery and the cavernous sinus[2,3]. Type C fistulae are abnormal connection between the meningeal or dural branches of the external carotid artery and the cavernous sinus.[2,3] Type D occurs between the meningeal or dural branches of both the internal carotid artery and the external carotid artery and the carvenous sinus.[1,3]

Type A fistula are usually high flow fistula which occur as a result of trauma or iatrogenic injury.[2,3,10]

Indirect fistula or dural fistula refers to dural or meningeal branches of the internal carotid artery and or the external carotid artery communicating with the cavernous sinus.[1-3] These fistulae; type B to D are usually low flow commonly seen in elderly women.[3] Predisposing factors to these types include systemic arterial hypertension, trauma, pregnancy, collagen vascular disease and artherosclerosis.[10-14]

Aneurysmal dilatation of the cavernous portion of the internal carotid has been demonstrated as a major underlying condition predisposing to spontaneous fistula[15-18]. Some connective diseases which predispose to weakening of the vascular wall with attendant risk of developing fistula in the presence of an appropriate precipitating factor such as coughing have been noted in the etiology of spontaneous CCF fistula.[2]Another important pathophysiology is an increase in intracavernous sinus pressure results in damage of the delicate vessels of the cavernous sinus in the presence of certain underlying conditions. Systemic arterial hypertension, artherosclerosis, pregnancy, diabetic, vasculopathy and minor trauma may possibly be underlying causes.[1,6,11,14,25-26] In pregnancy rise in blood pressure can lead to rupture of a preexisting aneurysmal dilatation and subsequently fistula formation.[25]

In the index case, no clinical features of connective tissue disease were present. However the history of pregnancy induced hypertension or straining during labour could have resulted in a rupture of a preexisting aneurysm. The anatomic configuration of her fistula was consistent with the presentation of that of a spontaneous indirect fistula. Traumatic carotid cavernous fistula is the most common type accounting for about two thirds of cases. [6] Head injury associated with basal skull is a major cause and this is by far common in males.

Vascular injury as a result of the direct tear from bony fragment or shear forces may lead to carotid cavernous fistula. Another mechanism is an increase intraluminal pressure in the ICA with simultaneous distal compression leading to vascular tear. [2,27] The management of CCF depends on the type, in combination with the nature of the flow, either high or low flow. The haemodynamic nature of the fistula which is helpful in understanding the clinical features and planning management is largely subject to bias. It is operator dependent.[3] The anatomic classification still serves as the key basis since it can be objectively assessed. Nonetheless the haemodynamic nature is still used to help classify the fistula and determine management options and give an idea with respect to prognosis.[1-3,29]

CLINICAL PRESENTATION

The clinical features depend on the site of the fistula, the extent of venous congestion and hypertension, the rate of flow and pressure gradient.[2] The features include proptosis, chemosis, diplopia, pain and visual loss. [28,30] These were demonstrable in the index case. The increased flow from the cavernous sinus into the orbital veins produces venous hypertension. This leads to oedema of the extraocular muscles manifesting as proptosis. The limited movement of the extraocular muscles as a result of oedema leads to diplopia.[3]The increased venous pressure results in impaired venous return of the conjunctiva presenting as chemosis.[31] Visual loss occurs as a result from decreased retinal blood flow due to stasis. Headache also occurs as a result retrograde cortical venous drainage. Pain is another symptom which occurs from possible involvement of the dural walls of the cavernous sinus in the direct type.[3,32] Sudden onset of symptoms associated with audible bruit on auscultation are consistent with spontaneous CCF arising from ruptured aneurysmal dilatation.[28]

RADIOLOGICAL EVALUATION

The radiological investigations that are beneficial in the diagnosis of this condition include; computed tomography (CT) scan, Magnetic resonance imaging (MRI) and cerebral CT angiogram.[1-3,29] This delineates the characteristics of the fistula and identifies features including; proptosis, extra ocular muscle enlargement, superior ophthalmic vein dilatation, venous drainage patterns, flow pattern and associated injuries such as fractures. The gold standard is cerebral CT angiogram.[1-3,29] In the index case the contrast enhanced was done to identify the anatomical configuration of the fistula. The standard angiography should be able to assess the common carotid and its bifurcation as well as the contralateral common carotid to exclude possible pseudo aneurysms, dissection or stenotic anatomy. [3] Open surgical intervention still has role in the management of carotid cavernous fistula. The Indications include; failure of trans-arterial or trans -venous endovascular intervention, and compromised proximal vascular access hindering endovascular approach.[3] Image guided intervention has become the mainstay of treatment. These approaches include trans arterial balloon occlusion technique, trans arterial coil occlusion technique largely for direct high flow Carotid-cavernous fistula. Indirect fistulas are managed via trans arterial or trans venous embolization techniques.[3] The absence of facilities for such intervention in our setting is a limiting factor to the management of such cases necessitating referral abroad to patronize the service. Carotid-cavernous fistula is an uncommon condition which requires the use of advances in interventional radiology for management. It is important for developing countries like Ghana to make such facilities available for the management of such cases.

REFERENCES

Barrow, L. D., Spector, H. R., Braun, F. I., Landman, J. A., Tindall, S. C. and Tindall, G. T. (**1985**). Classification and treatment of spontaneous carotid- cavernous sinus fistulas. *J Neurosurg* **62**: 248–256

Ellis, A. J., Goldstein, H., Sander Connolly, E. Jr., Meyers, M. P.(**2012**). Carotid-cavernous fistulas. *Neurosurg Focus* **32** (5): 310-312.

Ringer, A. J., Salud, L. and Tomsick, T. A. (**2005**). Carotid Cavernous Fistulas : Anatomy , Classification , and Treatment, *Neurosurg Clin N Am* **16**: 279–295

Peeters F. L. M., Kroger R. (1979). Dural and direct cavernous fistulas. *AJR Am J Roentgenol* 132:599–606.

Connors, J. J., Wojak, J. C. (1999). Treatment of carotid cavernousfistula. In: Interventional neuroradiology: strategies and practical techniques. *Philadelphia: WB Saunders*; 215–26.

Yoo, K., Krisht, A. F. (2000). Etiology and classification of cavernous carotid fistulas, Eisenberg MB, Al-Mefty O (eds): **The Cavernous Sinus: a Comprehensive Text.** Philadelphia: Lippincott Williams & Wilkins: 191–200

De Keizer R. (2003). Carotid-cavernous and orbital arteriovenous fistulas:ocular features, diagnostic and hemodynamic considerations in relation to visual impairment and morbidity. *Orbit* **22:**121–142

Debrun, G. M., Viñuela, F, Fox, A. J., Davis, K. R., Ahn, H. S. (1988). Indications for treatment and classification of 132 carotid-cavernous fistulas. *Neurosurgery* **22:**285–289

Viñuela, F., Fox, A. J., Debrun, G. M., Peerless, S. J., Drake, C. G. (1984). Spontaneous carotid-cavernous fistulas: clinical, radiological, and therapeutic considerations. Experience with 20 cases. *J Neurosurg* **60:**976–984

Lin, T. K., Chang, C. N., Wai, Y. Y. (1992). Spontaneous intracerebral hematoma from occult carotid-cavernous fistula during pregnancy and puerperium. Case report. *J Neurosurg* **76:**714–717,

Newton, T. H., Hoyt, F. W. (1970). Dural arteriovenous shunts in the region of the cavernous sinus. *Neuroradiology* **1:**71–81, **Raskind R., Johnson N., Hance D. (1977)** Carotid cavernous fistula in pregnancy. *Angiology* **28:**671–676,

Slusher, M. M., Lennington, B. R., Weaver, R. G., Davis, C. H. Jr. (1979). Ophthalmic findings in dural arteriovenous shunts. *Ophthalmology* **86:**720–731,

Toya, S., Shiobara, R., Izumi, J., Shinomiya, Y., Shiga, H., Kimura, C. (1981). Spontaneous carotid-cavernous fistula during pregnancy or in the postpartum stage. Report of two cases *J Neurosurg* **54:**252–256.

Parkinson D. (1967). Transcavernous repair of a carotid cavernous fistula: a case report. *J Neurosurg* 26:420–4.

Kobayashi, N., Miyachi, S., Negoro, M., Suzuki, O., Hattori, K., Kojima, T., Yoshida, J. (2003). Endovascular treatment strategy for direct carotid-cavernous fistulas resulting from rupture of intracavernous carotid aneurysms. *AJNR Am J Neuroradiol* 24**:**1789–1796

Kupersmith, M. J., Stiebel-Kalish, H., Huna-Baron, R., Setton, A., Niimi, Y., Langer, D., Berenstein, A. (2002). Cavernous carotid aneurysms rarely cause subarachnoid hemorrhage or major neurologic morbidity. *J Stroke Cerebrovasc Dis* 11**:**9–14

Linskey, M. E., Sekhar, L. N., Hirsch, W. Jr., Yonas, H., Horton, J. A. (1990). Aneurysms of the intracavernous carotid artery: clinical presentation, radiographic features, and pathogenesis. *Neurosurgery* **26:**71–79.