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Congenital Subclavian Av Fistula Presented With Proptosis of Eye Mimicking Caratico Cavernous Fistula: A Case Report

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Abstract
Congenital subclavian artery aneurysm with AV fistula neck causing proptosis of eye in a high flow Caratico Cavernous Fistula (CCF) are rare entity. No such case trace being literature so far. CCF caused by subclavian artery aneurysm are very rare. Computed Tomography (CT) angiography, Contrast Enhanced Computed Tomography (CECT) of Neck and Eye, Digital Subtract Angiography (DSA) are key tools for establishing diagnosis of these case. Surgery is ultimate option for dealing such a high flow fistula. We present a case of suclavian artery aneurysm with Arteriovenous Malformation (AVM) neck leading to abnormal congenital pathway resulting in a high flow CCF&which present as grave proptosis of eye. After proper evaluation and management patient get significant relief in proptosis as well as improvement of vision.

Keywords: *Subclavial Artery Aneurysm, Caratico Cavernous Fistula, Arteriovenous Malformation, Proptosis*

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AUTHOR'S COMMENT
This is a case report so no need of ethical committee approval.

I. Introduction

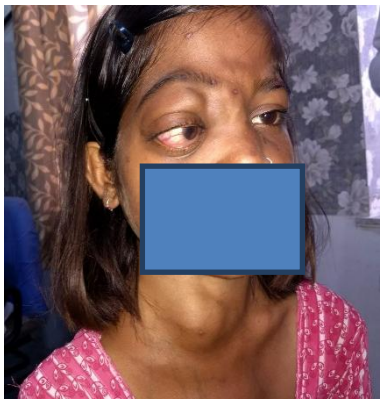
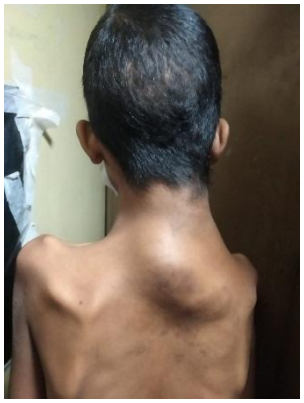
Arteriovenous Malformation of head and neck are defect of blood vessel. Blood vessels present themselves as tangled mass of artery and vein. AVM shares communication between arteries and veins (arteriovenous shunting) which lack normal capillary networks. The area with abnormal vasculature and shunting is called Nidus[1]. The cause of AVM head and neck are unknown, however, most developed during foetal development. They have wide range of clinical presentation and unpredictable course. Appearance of AVM depend on the size of blood vessel involved. Trauma is most common cause of neck region AVM. Spontaneous and congenital AVM in cervical region are rarely reported. Aneurysm of Subclavial artery aneurysm are extremely rare[2] (only 1% of all peripheral aneurysm)[3]. SCA aneurysm can be classified into two groups (i) Intrathoracic (ii) Extrathoracic.[4,5] 39% of SCA aneurysm involved proximal segment and most often cause by atherosclerosis, collagen disorders trauma, congenital or iatrogenic. [6]

Dural Carotico Cavernous Fistula may develop spontaneously in the setting of hypertension, atherosclerosis, collagen-vascular disease or childbirth, often low-flow in nature.**Dural arteriovenous shunts** are abnormal communications between the cavernous sinus and one or more meningeal branches of the internal carotid artery, external carotid artery or both. They tend to become symptomatic in middle-aged to elderly women. The pathogenesis of dural fistulas is unclear, but it appears that they originate from tiny arteriovenous shunts in the dura that are normal variants. "For some reason—possibly the effects of an angiogenesis factor—the shunts begin to proliferate and become abnormal.[7] Dural fistulas drain posteriorly first into the inferior petrosal sinus, basilar venous plexus or both and then anteriorly as the initial pathway becomes blocked. Posterior drainage usually produces no ocular symptoms, although some patients may experience a cranial neuropathy, such as facial paresis or ocular motor nerve paresis. Ocular signs and symptoms typically arise as the drainage shifts to an anterior route via the superior and inferior ophthalmic veins.

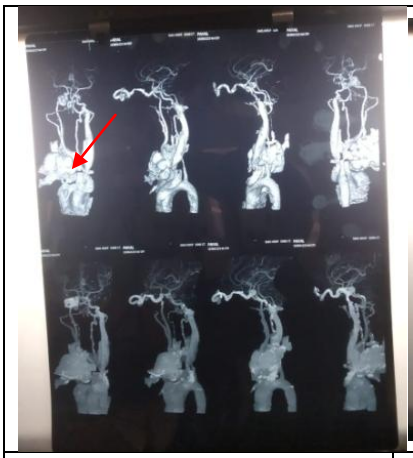
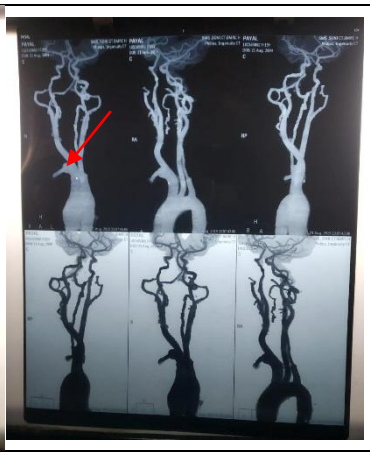
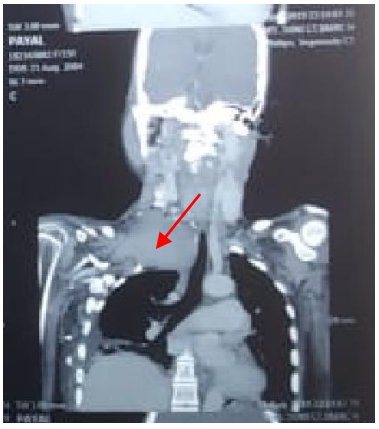
Elevated Intra Occular Pressure secondary to elevated episcleral venous pressure is also a common presenting sign in patients with CCFs.[8] Ophthalmoscopic abnormalities are more prevalent in patients with direct CCFs& include dilation of retinal veins, intraretinal haemorrhages, mild optic disc swelling and even non rhegmatogenous retinal detachments and choroidal detachments.

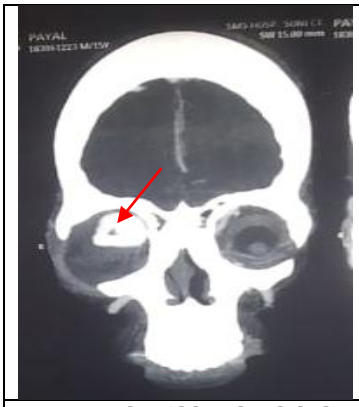
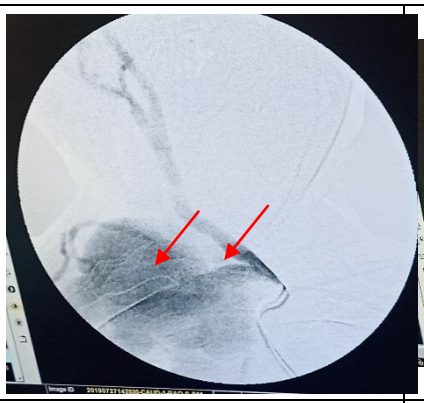
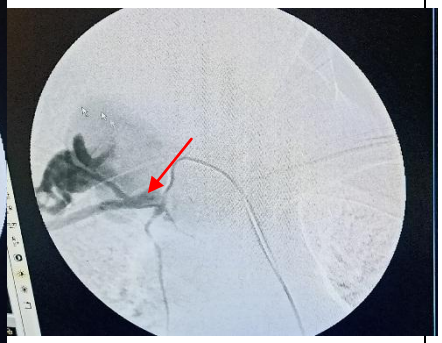
II. Case Report

15 year aged young female presented to neurology department with complain of gradual loss of vision in right eye progressing to blindness within 45 days, progressive painless protrusion of right eye from 1 year and another pulsatile massof gradual progression over front & right side of neck & upper back from 10 years. On examination right eye showed severe axial proptosis, chemosis& dilated &tortousepiscleral vessel and best corrected visual acuity was 0/0 with no light perception. Left eye examination was normal with 6/6 vision.


Investigation such as CT Angio of carotid vessel s/o large saccular aneurysm from rt. Subclavian artery extending in supraclavicular region 78/53/57 mm, multiple tortous arterial channel in rt. Post paravertebral location at C6 – D3, multiple early draining vein in rt. Internal Jugular Vein & rt. Subclavian vein s/o High Flow Vascular Malformation with associated aneurysm in feeding artery (subclavian artery).CT Angiography of carotid vessel in venous phase &CECT NECK s/o bunch of vascular channel near rt. paraspinal muscle in lower cervical & thoracic region s/o AV Malformation. Filling defect also seen in vertebral & spinal arteries s/o thrombus. CECT Orbit s/o filling of cavernous sinus in arterial phase with marked dilatation of rt .Orbital Vein, however no obvious communication with Internal Carotid Artery is visualised. DSA s/o CCF on rt. side from inferior petrosal sinus, Subclavicular fistula filling/connected from rt. Subclavian artery is ending into large venous channel in lower part of Neck. This fistula also filled from anterior intercostal artery. There is a duralAVF supplied by rt. External Carotid Artery. There is a large channel filling/ connected from vertebral and spinal venous plexus on right upper side of back.

		
CT Angiography showing aneurysmal SCA with AV SAC	CT Angiography showing all neck vessel	CT Angio showing AV SAC

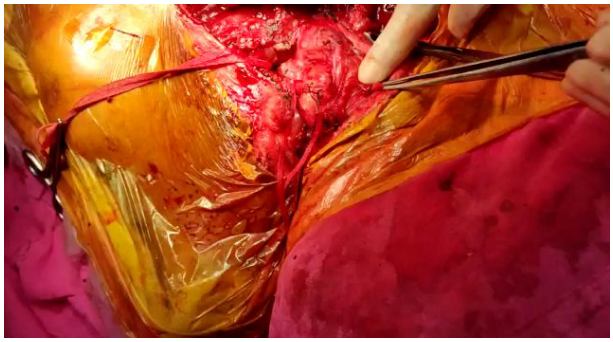
		
CECT Orbit (dilated ophthalmic vein and venules)	DSA showing aneurysmal SCA with AV SAC	DSA showing aneurysmal SCA

After relevant investigation, diagnosis of subclavian artery AVM with CCF is made and transferred to CTVS Department.

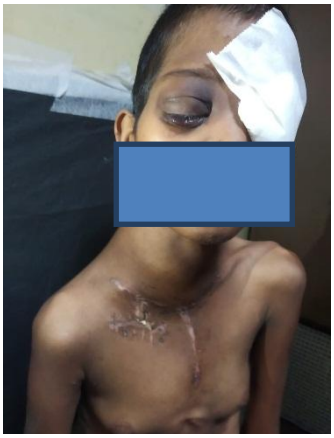
Surgery was planned, after taking prior consent, under General Anaesthesia. Upper midline partial sternotomy with extended incision to rt. supraclavicular region given.Rt.Subclavian artery found aneurysmally dilated and having an abnormal communication with a large sac on anterior part of neck behind carotid sheath.

Common Carotid Artery, ICA &ECA are looped, then aneurysmal part of subclavian artery excised along with ligation of communication path of AV sac. There was another communication between Rt. IJV& Rt. Brachiocephalic vein into posterior sac, this fistula/path was also ligated & excised.

Subclavian artery reconstructed using reverse saphenous graft.



Postoperatively, pt. recovered uneventfully & discharged. In 21 days follow up, there was significantly reduction of proptosis with improvement in vision 6/18 in right eye, reduction in size of neck & upper back swelling.



III. Discussion

AV malformations usually progresses over time. Initial quite lesion may progress to expansile mass during puberty or adolescence.[9]Once it progresses, it causes bleeding, ulceration, pain and cardiac volume overload.

Diagnosis of AVM was established by getting clinical history, performing physical examination,obtaining radiographic images & pathological results. CT arteriography revealed feeding arteries with dilated draining veins around a centralized nidus. Flow voids images suggestive of fast flow lesions. Multiple feeding arteries were present in each AVM.

Arteriovenous shunts may open, stimulating hypertrophy of surrounding vessels from increased pressure. Neovascularisation may be a primary stimulus for expansion. In this case subclavian artery aneurysm communicate with IJV, by an abnormal vascular channel and form a huge arteriovernoussac. This AV sac also has communication with cavernous sinuses. Probably back pressure of subclavian artery transmitted to cavernous sinus through abnormal AV sac.

According to current knowledge,the only curative treatment for AVM is complete removal or ablation of nidus, with recommended surgical treatment being complete radical resection. Radically resecting a vascular mass that may infiltrate normal structures in several tissue planes is difficult.[10]

A giant AV malformation was described by P.A. Dieng, etal and Senegal in both these cases venous drainage was in internal jugular vein but arterial supply was different.[11] It was external carotid artery in their case whereas in our case the arterial source was mainly from right subclavian artery.

Because of the risk of bleeding, some surgeons proceeding with 2 steps for the surgery.1ststage to reduce the size and flow byembolisation using gelfome etc. then surgical resection in 2nd stage. But in our case embolisation was tried by interventional neurologists but they abandoned the procedure due to very high flow AVM. Secondly due to pulmonary embolismas reported by M.E. Lidsky etal in 2012 after using gelfome and ethanol for embolisation.[12]

In our case there are 4 more sources of arterial supply to AV malformation which were not tackled may cause problem in future.

The localization of arteriovenous malformation on the neck induces surgical difficulties. The complete excision of the mass without nerve or vascular injury or major bleeding is a surgical challenge.

IV. Conclusion

A thorough evaluation of proptosis cases must be done before deciding on treatment.Decision on the mode of treatment of AVM should be taken carefully after weighing the pros & cons. Prompt diagnosis facilitates early and most appropriate management and therapy. Early intervention would not only salvage the eyes but also prevents patients from more disaster morbidity or fatality commonly due to intracranial haemorrhage or congestive heart failure.

Abbreviations:

AVM : Ateriovenous Malformation, SCA : Subclavian Artery, ICA : Internal carotid artery, ECA : External Carotid Artery, CCA : Common Carotid Artery, IJV : Internal Jugular Vein, CCF : Carotico Cavernous Fistula, CECT : Contrast Enhanced Computed Tomography, DSA : Digital Substraction Angiography, GA : General Anesthesia, AVF : Arteriovenous Fistula, IOP : Intraocular Pressure

References

[1]. YoungAE,Mulliken JB.Arteriovenous malformation.In:MullikenJB,edVascular Birthmark:Haemangioma and Malformation.Philadelphia:WBSaunders,1988;228-45.

[2]. Dent TL, Lindenauer SM, Ernst CB, Fry WJ. Multiple arteriosclerotic arterial aneurysms. Arch Surg 1972; 105: 338-44.

[3]. Dougherty MJ, CalligaroKD, Savarese RP, De Laurentis DA. Atherosclerotic aneurysm of the intrathoracicsubclavianartery : A case report and review of the literature. J VascSurg 1995; 21: 521-9.

[4]. Vierhout BP, Zeebregts CJ, Van Den DungenJJ; Rejjnen MM. Changing profiles of diagnostic and treatment options in subclavian artery aneurysms. Eur J VascEndovascSurg 2010; 40: 27-34.

[5]. SaloJA, Ala-Kulju K, Heikkinen L, Bondestam S, Ketonen P, Luosto R. Diagnosis and treatment of subclavian artery aneurysms. Eur J VascSurg 1990; 4: 271-4.

[6]. Mohan IV, Stephen MS. Peripheral arterial aneurysms. Open or endovascular surgery. ProgCardiovasc Dis 2013;56: 36-56.

[7]. Erdmann MW, Jackson JE, Davies DM, Allison DJ. Multidisciplinary approach to the management of head and neck arteriovenous malformations.AnnRCollSurg Engl.!((%;77(1):53-9.

[8]. Chaudhary IA,Elkhamry SM, Al-Rashed W, Bosley TM .Carotid Cavernous fistula;opthalmological implication.Middle East African Journal of Opthalmology,2009;16(2):57-63{PMC free article} {Pubmed} {Google Scholar}.

[9]. Holman E, Reflections on arteriovenous fistula,Ann ThoracSurg 1971;11(2):176-86.

[10]. Kohout MP,HansenM,PribazJJMulliken JB. Arteriovenous malformations of head & neck:natural history & management,PlastReconstrSurg 1998:102(3);643-54.

[11]. P. A. Dieng, P. S. Ba, M. Gaye, S. Diatta, M. S. Diop, E. Sene, A. G. Ciss, A. Ndiaye, and M. Ndiaye. Giant Arteriovenous Malformation of the Neck. Case Rep Vasc Med. 2015; 2015: 124010. PMCID: PMC4546949. PMID: 26347847. Published online 2015 Aug 10. doi: 10.1155/2015/124010.

[12]. M. E. Lidsky, J. N. Markovic, M. J. Miller Jr., and C. K. Shortell, "Analysis of the treatment of congenital vascular malformations using a multidisciplinary approach," *Journal of Vascular Surgery*, vol. 56, no. 5, pp. 1355‑1362, 2012.View at: Publisher Site | Google Scholar

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