Cirsoid aneurysm and MDCT - An excellent modality for mapping a rare complex extracranial pathology - A case series

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Authors: S. Arooj, S. Hamid, K. Baksh; Karachi/PK
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Learning objectives

Cirsood aneurysms are rare arteriovenous malformations of the scalp. They are challenging to treat because of their complexity and danger of potential profuse hemorrhage or cosmetic derangement. A well-rehearsed presurgery radiological mapping of the arteriovenous malformation with exact identification of the feeding artery/arteries and draining veins can substantially reduce these complications. Although conventional angiography is the gold standard for the diagnosis of arteriovenous pathologies, Multidetector computed tomography with three dimensional imaging is rapidly being accepted because of its high accuracy and non-invasive nature.

We propose to attend this:

- To project this significantly life threatening rare condition.
- To have an idea how to approach these cases from a radiological point of view.
- To provide a roadmap to the surgeon with a minimally invasive approach.
Background

Cirsoid aneurysms are vascular aberrations of the scalp. The term aneurysm is a misnomer, as these are actually fistulous connections between feeding arteries and draining veins first described by Brecht in 1833. These can be either congenital or traumatic in origin. They are thought to arise during the fetal period as a result of abnormal differentiation of the fetal vasculature with absent capillary bed. They have a characteristic variceal dilatation of the draining veins. Various terminologies have been used to describe these lesions, such as serpentinum aneurysm, aneurysm racemosum, arteriovenous aneurysm, plexiform angioma, arteriovenous fistula and arteriovenous malformation. These lesions usually present with spontaneous or post-traumatic, localised, pulsatile scalp swelling which may be associated with skin discoloration, tinnitus or haemorrhage. Cosmetic deformity secondary to these lesions is sometimes the leading cause of medical intervention.

In majority of the patients, the superficial temporal artery is the main supply to the fistula primarily due to its unprotected course and its consequent high risk of incurring trauma. The occipital artery is the second most affected vessel. The venous drainage is through the corresponding scalp veins, which become progressively dilated and tortuous with time. These lesions are usually asymptomatic resulting in enormous sizes at the time of initial presentation. They can present clinically as pulsatile, innocuous or monstrous swelling, with skin discoloration and rarely, profuse haemorrhage and intractable tinnitus. They are evenly distributed amongst the frontal, temporal and parietal regions with a slight right preponderance as observed by some researchers. A comprehensive depiction of the feeding artery (or arteries), draining vein(s) and arteriovenous communication is necessary in order to make an appropriate management decision. Whilst conventional angiography is the gold standard, Multidetector computed tomography (MDCT) is rapidly becoming the preferred method of imaging in the acute setting because of its minimally invasive nature.
Findings and procedure details

Here we are reporting a case series of three patients with arteriovenous malformations of the scalp who presented to our radiology department for CT angiography of brain. Two of them shared a common previous history of blunt head trauma resulting in progressive development of pulsatile masses on their scalp causing cosmetic deformity.

A 16 slice MDCT is used to perform exams. Protocol included intravenous Ultravist 100 ml at the rate of 3.0ml/ second. 3D CTA images were also acquired.

PATIENT 1: A 32 years old Caucasian female presented to us with a grotesque pulsatile swelling over her forehead. She has history of blunt head trauma after which she noted enlargement her of scalp vessels and pulsatile mass formation in the midline. Because of her remote geographical location she could not seek any emergent medical help. On clinical examination, there was a 6.0 x 4.5 cms soft compressible pulsatile swelling over her forehead. A bruit was detected over the lesion. There were no sensorimotor deficits or overlying skin discoloration. Her cerebral angiography identified multiple tortuous dilated contrast filled vessels forming a racemose baggy network originating from the left superficial temporal and supratrochlear artery with drainage into the subcutaneous cortical veins. [Figure - 1]. Three-dimensional volume rendering MDCT revealed the cirsoid aneurysm very elegantly with dilatation of the superficial temporal and supratrochlear arteries on the left of midline. [Figure - 2]. She was offered transarterial embolization by the neurosurgical team which she declined.

PATIENT 2: A 36 years old female patient presented with severe intermittent headache and five pulsatile lumpy swellings over her scalp within the hairline region but they were big enough to produce visible bulge on the overlying hair causing cosmetic issues. Largest one measured approximately 3.0 x 2.0 cms. These were first noticed by the patient eight years ago after which they progressively increased in size. She did not consult her physician due to the absence of any pain or discomfort. She was not able to recall any definite history of blunt head trauma. They were no comorbidities. She had no history of head surgery. She had refused any operative procedure offered by her consulting surgeons. Her contrast enhanced CT scan brain and four vessel cerebral angiogram were performed which demonstrated completely unremarkable cerebral circulation. However, multiple tortuous dilated contrast filled vessels were identified within the subcutaneous tissues of the scalp in the left parieto-occipital region with feeding artery from dilated left superficial temporal artery and drainage into the subcutaneous cortical veins with intracranial extension [Figure - 3]. Three dimensional CT images revealed aneurysmal dilatation of the vessels more clearly. [Figure - 4].

PATIENT 3: A 40-year-old man presented with a mass on his right preauricular region associated with pulsatile tinnitus. He had suffered blunt injury in this region during a road traffic accident followed by the appearance of this mass few months later. His
clinical examination demonstrated a pulsatile mass of approximately 4.5 x 3.0 cms on the right preauricular region. Cerebral angiography identified a high output fistula between the external carotid artery and superficial temporal vein that presented as large venous aneurysm. Three dimensional cerebral angiogram clearly demonstrated a large fistula with arterial supply from the external carotid artery and drainage into superficial temporal vein and some subcutaneous cortical veins. Surgical options were offered to the patient but he was too reluctant to opt them. He was discharged with an advice of regular follow-up.

Contrast medium administration and parameters affecting bolus geometry need to be carefully configured to match the arterial and venous enhancement pattern of cirrhotic aneurysms during MDCT. Identification of the recruited feeding vessels from the adjacent normal arteries, morphology of the nidus, and drainage of venous structures can be determined prior to intervention.

Primary differential diagnosis of a pulsatile scalp swelling is superficial temporal artery aneurysm. The indications for operative intervention include aesthetic concerns, relief of symptoms and prevention of complications such as haemorrhage. Management of the cirrhotic aneurysms of scalp is very challenging because of its high flow, complex vascular anatomy and cosmetic problems. There are various techniques and method of treatment for scalp AVMs. The treatment options include surgical excision, ligation of feeding vessels, transarterial and transvenous embolization, injection of sclerosant into the nidus and electrothrombosis. Vascular embolization is rapidly becoming popular as it obviates the need for surgery, which is associated with an increased risk of complications such as scarring, deformity and bleeding.
Fig. 1: Axial CT image of cirsoïd aneurysm with tortuous dilated contrast filled vessels forming a racemose baggy network originating from the left superficial temporal artery and subcutaneous cortical draining veins.

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**Fig. 2:** Three-dimensional volume rendering MDCT of the cirsoid aneurysm with dilatation of the superficial temporal and supratrochlear arteries on the left.

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Fig. 3: Axial CT image revealed multiple tortuous vessels within the subcutaneous tissues of the scalp in the left parieto-occipital region.

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Fig. 4: Three dimensional computed tomography angiograms demonstrated dilated tortuous network of vessels originating from the left superficial temporal artery.

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Conclusion

Evaluation of the angioarchitecture in cirsoid aneurysms is considerably improved with the use of Multidetector computed tomography with cerebral angiography. The facility of three dimensional image reconstruction and liberty to visualize the abnormal vascular network in any plane has rapidly increased its utilization.
References


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