Scalp cirsoid aneurysm

They are also referred to as aneurysma serpentinum, aneurysm racemosum, or plexiform angioma.

Cirsoid aneurysm of the scalp is one of the rarest occurrences in neurosurgery. It is an aneurysmal tumor formed by arteriovenous fistula of the arteries and veins of the scalp [1)]

They can be easily misdiagnosed and mistreated.

Historically their unusual portly appearance led to various synonyms being coined for the entity [2)].

Epidemiology

Scalp AVM (SAVM) is a rare condition [3) 4)] and infrequently encountered by the neurosurgeon [5)].

Classification

Yokouchi et al . classified scalp arteriovenous fistulae into three types: Type A: has a single fistulous connection from the proximal portion of the artery, Type B: has a single fistulous connection but from multiple distal portions of the arteries and Type C: where there are multiple fistulae constituting the plexiform feeding type.

Sometimes the drainage can be intracranial with an associated cerebral arteriovenous malformationYokouchi et al . classified scalp arteriovenous fistulae into three types: Type A: has a single fistulous connection from the proximal portion of the artery, Type B: has a single fistulous connection but from multiple distal portions of the arteries and Type C: where there are multiple fistulae constituting the plexiform feeding type.

Sometimes the drainage can be intracranial with an associated cerebral arteriovenous malformation

Etiology

Its origin can be congenital or traumatic.

Congenital arteriovenous malformations (AVMs) of scalp are rare. They are usually not symptomatic at birth and are often misdiagnosed as haemangiomas. To date, only two cases of symptomatic neonatal scalp AVM have been reported in literature. Pathophysiology of congenital AVM is not completely understood but genetic and acquired causes are implicated. Diagnosis and management are often difficult and require multidisciplinary approach. Hussain et al. report a rare case of symptomatic congenital scalp AVM in a 10-day-old neonate who was successfully managed [6)].

Clinical

The clinical picture presents with complaints of increased scalp, scalp disfigurement, pain and neurological symptoms.

They can present a subcutaneous scalp lump or a large, pulsatile mass with a propensity to skin erosion and massive haemorrhage [7) 8)].

The symptoms associated with cirsoid aneurysm of scalp vary according to the size of the fistula. Common clinical manifestations include loud bruit, pulsatile scalp mass, headache, and tinnitus. If left untreated, there is an increased risk of developing life-threatening complications such as aneurysmal hemorrhage or scalp necrosis [9) 10) 11)].

Diagnosis

Since cerebral blood flow may decrease depending on the progression of the lesion, the cerebral blood flow should be evaluated. Considering the treatment modalities depending on the lesion can provide treatment with less recurrence and higher patient satisfaction [12)].

Treatment

Scalp cirsoid aneurysm treatment.

Outcome

Even after complete surgical resection, a case of recurrence after 18 years has been reported [13)], which is why regular follow-up is advised.

Systematic Review

Sofela et al. conducted a PUBMED, SCOPUS, OVID SP, SciELO, and INFORMA search using the keywords; “cirsoid,” “aneurysm,” “arteriovenous,” “malformation,” “scalp,” “vascular,” and “fistula.” They identified 74 pertinent papers, reporting 242 cases in addition to our reported index case.

Median age at presentation was 25 yr (range 1-72 yr); male to female ratio was 2.5:1. The most common symptoms were a pulsatile mass (94% of patients), headaches (25%), and tinnitus (20%). The median duration of symptoms was 3 yr (6 d to 31 yr), with 60.2% occurring spontaneously, 32.23% traumatic, and the rest iatrogenic. A total of 58.5% of cases were managed with surgical excision only, 21.6% with endovascular embolization only, and 14.5% with a combination of both methods. The complication rate observed in the endovascular embolization treatment cohort (55.8%)

was significantly higher than that observed in the surgical excision only cohort (9.9%) (P < .00001) and in the combined therapy cohort (0%) (P < .00001). There is a low recurrence rate after treatment irrespective of modality: surgical excision only (6.3%), endovascular embolization only (8.3%), and combined therapy (0%).

Scalp cirsoid aneurysms are associated with good prognoses when recognized and managed appropriately. They suggested combining surgery with endovascular embolization as the optimum treatment modality [14)].

Case series

Scalp cirsoid aneurysm case series.

Case reports

Scalp cirsoid aneurysm case reports.

Unclassified

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