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Case Report

Arteriovenous malformation of the external ear. A new case report

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Abstract
Arteriovenous malformations are seldom extracranial. External ear is a rare location, with sometimes an important esthetical prejudice. Its diagnosis is based on radiology. The management of this entity might be challenging. It may need multidisciplinary approach: Interventional radiologists and surgeons.
Our case concerns an arteriovenous malformation located in external ear, complicated by bleeding and managed with embolization

Keywords: Malformation, Ear, Embolization

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Arteriovenous malformation of the external ear. A new case report

Introduction:

An arteriovenous malformation (AVM) is a direct aberrant communication between arterial and venous systems, with absence of capillary connections. It is mainly located in the intracranial region, lung and kidney. External ear location is uncommon. Sometimes, the diagnosis and the management of this entity is challenging. Bleeding is the major complication. The effective treatment for an AVM is preoperative selective embolization and complete excision.

Case report

A 54-year-old male patient presents a right auricle reddish swelling tumefaction, since his young age, with no significant medical follow-up. His was hospitalized 5 days ago, in cardiology department for acute coronary syndrome. He was on

Coumadin anticoagulant therapy after a placement of two coronary stents. He was referred to our emergency department for abundant bleeding originating from his ear AVM, two days after debuting anticoagulation. The was no overdose of coumadin. The ladder was stopped and Calciparin-based anticoagulation was debuted. Vital signs were within normal limits. Otologic exam found three reddish centimetric formations on the helix of the right auricle. It was not pulsatile, with no inflammatory signs. Another 3 centimeter right retro auricular mass was found, visibly pulsatile. A bruit was heard on auscultation of the mass. These formations were bleeding on contact (Figure 1).

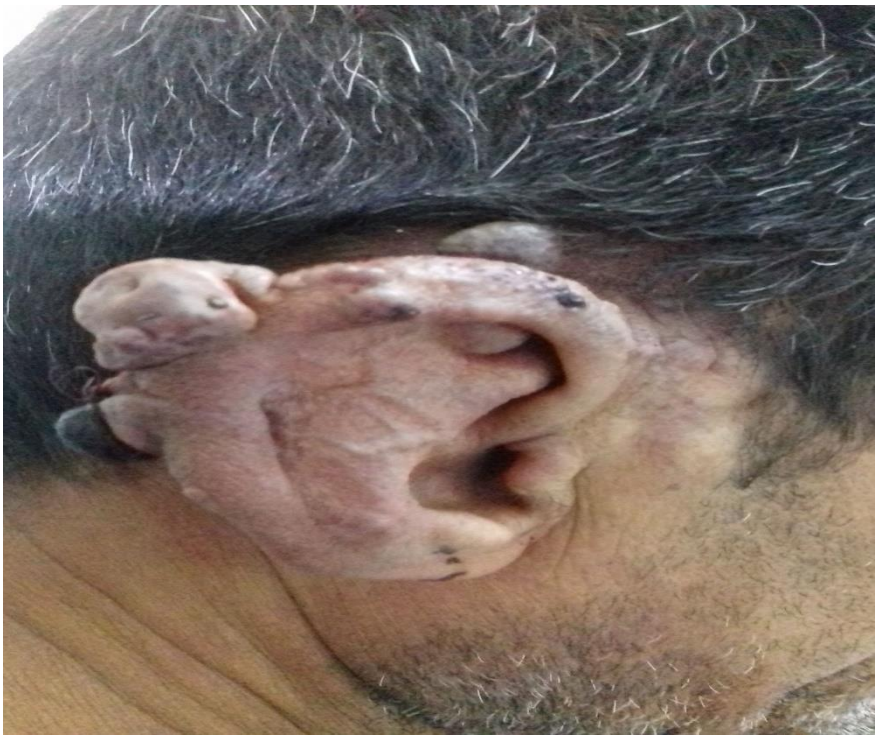

Figure 1: Pre embolization view of arteriovenous malformation of the external right ear

Otoscopic and general physical examination were unremarkable. Laboratory investigations showed no abnormalities. Clotting studies were unremarkable. Hemoglobin rate was at normal range. The patient was hospitalized in otolaryngology Department, and bleeding was controlled with a pressure bandage.

A dynamic contrast-enhanced cerebra-cervical MRI was performed. It showed a external right ear AVM, supplied by a branch of homolateral external carotid artery. It was drained by a developed subcutaneous veinous network issued from internal table, towards the superior longitudinal sinus (Figure 2).

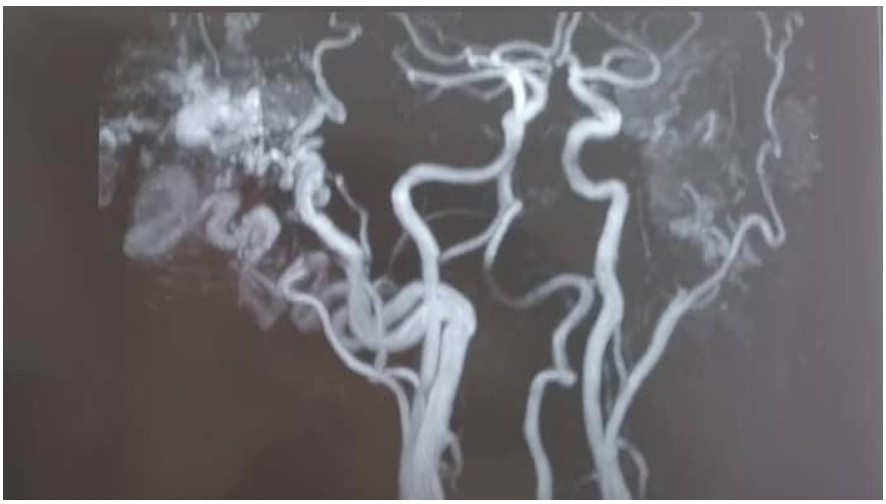

Figure 2: Cervical enhanced MRI showing external right ear arteriovenous malformation, drained by a branch of external right carotid

During his hospitalization, the patient presented recurrent spontaneous bleeding from the AVM. Hemoglobin rate dropped from 14 to 8 g/dl, requiring the transfusion with two packed red blood cells.

The Dilemma in the management of this patient was in the balance between the risks and benefits of debuting an anticoagulation, considering the history of cardiac disease with recent stents placement, and, on the other side, the acute problem of recurrent bleeding. It was decided to treat this haemorrhage conservatively due to anaesthetic risk.

The catheterisation was performed from the right femoral artery. The angiography objected an important AVM of the right ear supplied by two dilated and tortuous arteries: Posterior branches of the superficial temporal artery and the right auricular artery. The embolizing product used was Onyx: 11 ml of Onyx were injected during 40 minutes allowing the exclusion of 90% of the malformation. Immediate post-embolization angiographic control showed almost complete exclusion of the AVM.

Given the absence of recurrence of bleeding, our patient was discharged 7 days post-embolization after adapting his anticoagulant treatment (Figure 3).

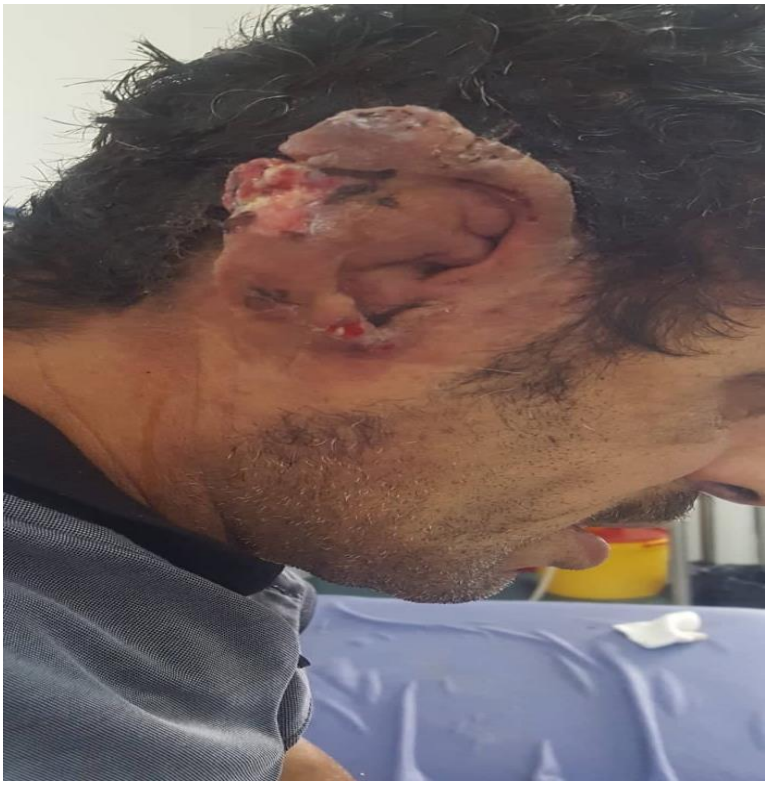

Figure 3: Post embolization view of arteriovenous malformation of the external right ear

At 2 month follow up, the patient had no further episodes of bleeding. He was referred to plastic surgery department for esthetic excision of the remaining blackish formation of the helix generated by embolization.

Discussion:

An AVM is the result of failure of regression of arteriovenous channels in the primitive retiform plexus between the fourth and sixth weeks of gestation. An AVM is present at birth but usually is not clinically significant, except for a cosmetic problem [1]. It has also been postulated that local ischemia plays a role in pathogenesis. This high blood flow is responsible for changes in the vessel wall which may explain their fragility with a significant risk of rupture. Unlike infantile haemangiomas, adult AVMs have no invasive evolution. It continues to evolve to the detriment of the surrounding healthy tissues due to the lack of specific vascularization and reduced perivascular regulation [2].

AVMs are congenital but may not become apparent until several years of life, even in adulthood. They may be visceral or superficial. Superficial AVMs are rare and they are the most aggressive [1].

Superficial AVMs have a polymorphic clinical presentation. They may present as a warm, pulsatile, subcutaneous reddish mass [3].

The diagnosis of AVMs is radiological. Indeed, physical examination only allows suspicion of the diagnosis. Doppler ultrasound can orientate the diagnosis. Magnetic resonance imaging (MRI), especially T2-weighted MRI and MRI angiography are the cornerstone of radiologic diagnosis. It can confirm the hyper vascular nature of the mass, the size of the nidus, the extension of the AVM and its relationship with the adjacent tissues [4].

Biopsy is not recommended or even contraindicated due to the risk of hemorrhage, especially as it can lead to a relapse of the AVM [3].

Auricular AVM had been the subject of single case or a few case reports and, thereby, there are no standard guidelines for the management of this entity [4]. Most authors recommend If the auricular AVM is small and asymptomatic, the treatment is not usually needed. However, the treatment for the large symptomatic auricular AVM is often difficult [4,5].

Optimal treatment for the auricular AVM is a combined approach of selective embolization and complete excision. If possible, complete excision should be performed within 48 h after embolization because of collateralization. Other authors suggest a period between 5 days and 6 weeks to avoid re-expansion of the vascular malformation [4]. treatment with laser, steroid or irradiation has not been proven to be effective [3,5]. Partial excision and proximal ligature are not curative and should be avoided because revascularization and new collateral circulation will lead to recurrence [1,4]. Super selective embolization alone was used for palliation, but its use is seldom successful because of the formation of new collateral vessels [4,5].

New perspectives are currently being tested such as mTOR inhibitors (Mammalian target of rapamycin inhibitors) whose efficacy in AVMs is debatable [1].

Conclusion:

AVMs are congenital anomalies that can occur at any age. They can be life threatening due to their high risk of bleeding. Partial excision is not possible in this type of anomaly. Interventional radiology is increasingly finding its place in the management of AVMs. The cervico-facial location of these anomalies is not uncommon and requires multidisciplinary intervention to have the best possible vital and esthetic outcomes.

Ethics approval and consent to participate: The approval of the current study has been granted by the medical committee of research ethics of Military Hospital. Written informed consent was obtained from the patient for publication of this study. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Consent for publication: Written informed consent was obtained from the patient for publication of this study. A copy of the written consent is available for review by the Editor on request.

Competing interests: The authors declare that they have no competing interests

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