Young Female with Acute Ischemic Stroke Secondary to Paradoxical Embolism from an Undiagnosed PAVM/HHT. Case Report and Literature Review

Ammar Ashraf*, Ahmed Mohamed Abdelrahman, Ibrahim Abdulhakim Ibrahim Al Shikmubarak, Osama Rizk and Marwan Abdulrahman Alshaikh Husain

Department of Medical Imaging, Ministry of National Guard Health Affairs, King Abdulaziz Hospital, Al-Ahsa, Saudi Arabia

*Corresponding Author: Ammar Ashraf, Department of Medical Imaging, Ministry of National Guard Health Affairs, King Abdulaziz Hospital, Al-Ahsa, Saudi Arabia.

Received: February 04, 2020; Published: February 17, 2020

Abstract

Pulmonary arteriovenous malformation (PAVM) is an abnormal direct capillary free communication between pulmonary arteries and veins which results in a right-to-left intrapulmonary shunt. Depending on the size and number of these vascular malformations and degree of the right-to-left shunt, PAVM can present with dyspnea, cyanosis or paradoxical embolism. Paradoxical embolism from a previously unknown isolated PAVM is a rare cause of ischemic stroke which is usually overlooked in most cases at the initial presentation. Transcatheter embolization (TCE) is the gold standard treatment for these PAVMs which decreases the risk of paradoxical embolism and other associated complications.

Here, we report a case of a young female presenting with acute ischemic stroke secondary to paradoxical embolism from an undiagnosed PAVM. We also review the related literature.

Keywords: PAVM; Capillary Free; Ischemic Stroke

Abbreviations


Introduction

Pulmonary arteriovenous malformations (PAVMs) are uncommon high-flow, low-resistance anomalous capillary free direct communication between the pulmonary artery and the pulmonary vein, and thus creating an anatomical extra-cardiac right-to-left shunt [1-5]. This extra-cardiac shunt bypasses the eminent normal “filter capacity” of the pulmonary capillary bed (Figure 1) [6], which predisposes to certain complications including cerebrovascular system and can cause stroke, transient ischemic attack and cerebral abscess [1,2,4,6]. Clinical presentation is inconstant and relies on the number and size of the AVMs as well as the degree of the shunt [3,4]. Transcatheter embolization is the established standard of care for pulmonary AVMs to impede paradoxical embolism, even in cases without any symptoms [2,4,6]. Currently, embolization is advocated for all PAVMs which are suitable for the procedure, as the risk of stroke does not depend on the size of PAVMs [2]. Surgical resection is reserved for larger lesions and lesions with multiple complex feeding vessels [1,4].

Citation: Ammar Ashraf, et al. "Young Female with Acute Ischemic Stroke Secondary to Paradoxical Embolism from an Undiagnosed PAVM/HHT: Case Report and Literature Review". EC Clinical and Medical Case Reports 3.3 (2020): 01-11.
Young Female with Acute Ischemic Stroke Secondary to Paradoxical Embolism from an Undiagnosed PAVM/HHT. Case Report and Literature Review

**Figure 1:** Drawing showing the main role of the pulmonary capillary bed that is omitted by the pulmonary arteriovenous malformations (red arrow) [6].

**Case Report**

A 36-years-old female presented to our hospital emergency department with a history of sudden onset of right-sided weakness, aphasia and decreased level of consciousness of 03 hours duration. There was no history of hypertension, hyperlipidemia, diabetes, smoking, epilepsy or any cardiac problem. Past gynecological history revealed two episodes of abortion. She had a strong family history of venous thrombosis. Upon arrival at the emergency department, the patient's Glasgow coma scale (GCS) score was 10/15. She had right-sided paralysis with 0/5 power in right upper and lower extremities, decreased sensations in right extremities and up going right plantar reflex. She had spontaneous eye-opening with bilateral reactive pupils, left-sided nystagmus and left upper motor neuron facial palsy. She was unable to talk (motor aphasia).

An urgent unenhanced CT brain (Figure 2) was performed which showed hyperdense left middle cerebral artery (MCA) with evolving ischemic changes in its territory as well as ischemic changes in the left posterior cerebral artery (PCA) territory. No acute hemorrhage or significant mass effect/midline shift was seen. A chest radiograph (portable frontal view) was also done which showed an indeterminate nature, well-defined 2 cm opacity in right lower lung zone for which further evaluation with CT chest was suggested by the radiologist (Figure 3). The patient was admitted to ICU, the stroke pathway was activated and aspirin (antiplatelet) and atorvastatin (lipid-lowering) were started. Thrombolytics were not given due to large infarction and lapse of time (approximately more than 6 hours). Follow up CT brain at 24 and 48 hours interval (Figure 2) showed an interval increase in the size of left MCA and PCA territorial infarctions and mass effect with contralateral midline shift. There was also an interval development of a few small acute infarcts in right basal ganglia on follow up scans. No hemorrhagic transformation was noted. Due to infarctions in multiple vascular territories, the possibility of an undiagnosed right-to-left intra-cardiac shunt (like ASD) was blamed for this stroke. A transthoracic echocardiogram was performed which was, however, negative. Carotid Doppler ultrasound and venous Doppler ultrasound examination of bilateral lower extremities were performed which were also negative. Extensive laboratory investigations [anti-nuclear (ANA), anti-double-stranded DNA (anti-dsDNA), anticytome (ACA), antineutrophil cytoplasmic (ANCA) antibodies, rheumatoid factor (RF), complement C3 and C4] were done to exclude the
Young Female with Acute Ischemic Stroke Secondary to Paradoxical Embolism from an Undiagnosed PAVM/HHT. Case Report and Literature Review

suspicion of autoimmune vasculitis as the underlying etiology of this stroke; however, all these laboratory investigations were negative. Bleeding and clotting profile [prothrombin time (PT), partial thromboplastin time (PTT), international normalized ratio (INR), protein C and protein S] was also within normal limits. No mutation was seen in the factor V Leiden gene. Serum iron and fibrinogen levels were normal. Routine blood investigations (complete blood count, renal profile, liver profile) were also normal.

On day 5 of her admission, CT carotid angiogram was done which showed fetal left posterior communicating (P.COM) artery which was likely an anatomical variant; otherwise, no significant cerebral vascular pathology was seen. However, an incidental finding of bilateral pulmonary embolism was noted in the partially scanned upper chest on this carotid CT angiogram which was later on properly evaluated with a dedicated CT pulmonary angiogram. Extensive atelectatic changes were seen in both lower lobes and the radiologist overlooked the pulmonary opacity seen on baseline chest radiograph.

Heparin infusion followed by warfarin was started for pulmonary embolism. Due to acute infarcts in multiple cerebral vascular territories and extensive bilateral pulmonary embolism in a young female without any obvious cause, the vigilant cardiologist decided to go for a trans-esophageal echo cardiogram with contrast (air bubbles), despite normal transthoracic echocardiogram. No intra-cardiac shunt was seen on this trans-esophageal echo; however, it was positive for air bubbles in left heart chambers in 4th and 5th cardiac cycles which raised a high suspicion of (at least grade 2), extra-cardiac/intrapulmonary right-to-left shunt (Figure 3). After this echocardiogram, CT pulmonary angiogram scan was reviewed on cardiologist’s request and a pulmonary arteriovenous malformation (AVM) measuring 2 x 2 cm with a feeding artery measuring 3 mm, was diagnosed in the right lower lobe, which had initially been overlooked by the radiologist (Figure 3).

The patient underwent genetic analysis for hereditary hemorrhagic telangiectasia which was positive for mutations in the endoglin (ENG) gene and negative for mutations in the ACVRL1 gene. After the diagnosis of hereditary hemorrhagic telangiectasia (HHT), the patient underwent detailed systemic evaluation (ENT, dermatology and gastroenterology); however, no other associated findings of HHT were seen.

She recovered well from the massive stroke. Due to eligibility issues and lack of interventional radiologist in our hospital, the patient was referred to her primary health care system for further evaluation (with possible embolization) by an interventional radiologist; however, the patient refused any such intervention. The patient opted for medical management (anticoagulation). She is on regular follow up with neurologist and hematologist. She is on oral warfarin 3 mg. She did not have any recurrent stroke or other HHT associated complications during the last five years follow up. She has flexion deformity of the right big toe, good memory and speech, 4/5 power in her right extremities and can do her daily domestic activities without any support. PAVM is stable on follow up chest radiographs.

Discussion

PAVM is an anomalous vascular communication between the pulmonary arteries and veins that result in an extra-cardiac intrapulmonary right-to-left shunting [1]. Pulmonary AVM is a rare disease entity that can cause stroke due to paradoxical cerebral embolism [1,4,7]. In the past it was considered a rare disorder with an estimated incidence of 2-3 per 100,000; however, recent literature shows a higher frequency with a prevalence of approximately 1 in 2,600 [6,8-11].

Most commonly PAVMs are congenital malformations but rarely, maybe acquired; causes of acquired PAVMs are chronic infections such as tuberculosis, schistosomiasis, actinomycosis, hepato-pulmonary syndrome (HPS) due to chronic liver disease, metastatic thyroid carcinoma, trauma and iatrogenic (due to cardiovascular intervention for congenital cyanotic heart disease) [3,5,6,12,13].

Approximately 80-90% of PAVMs are seen in association with hereditary hemorrhagic telangiectasia (HHT) or Osler-Weber-Rendu syndrome (OWRS) [1-5,7-9,11,12]. Remaining are sporadic cases (idiopathic/isolated), accounting for less than 10% of cases [1-5,9,11,12].

Citation: Ammar Ashraf, et al. "Young Female with Acute Ischemic Stroke Secondary to Paradoxical Embolism from an Undiagnosed PAVM/HHT. Case Report and Literature Review", EC Clinical and Medical Case Reports 3.3 (2020): 01-11.
Figure 2: Plain CT brain; day 1 (A and B) shows hyperdense left MCA sign with early ischemic changes. Follow up CT brain at day 3 (C) shows well established non-hemorrhagic acute infarction in left MCA territory with mass effect. There are additional findings of acute infarctions in the left PCA territory and right basal ganglia (which was not obvious on baseline scan). CT brain at day 10 (D) shows resolving stroke and its mass effect.

Figure 3: Baseline chest x-ray (A) shows a well-defined nodular opacity in the right lower lung zone. Reformatted coronal MIP* image from CT pulmonary angiogram (B) shows multiple filling defects in the right lower lobe pulmonary artery and an AVM in the right lower lung lobe (arrow). Trans-esophageal contrast echocardiography shows no intracardiac shunt in initial cardiac cycles (C) and appearance of air bubbles in left heart chambers after 4th and 5th cardiac cycles (D), suggestive of extra-cardiac intrapulmonary right-to-left shunt. *MIP: Maximum intensity projection.

Video 1: Trans-esophageal echocardiogram with contrast (air bubbles).
Young Female with Acute Ischemic Stroke Secondary to Paradoxical Embolism from an Undiagnosed PAVM/HHT. Case Report and Literature Review

Association of pulmonary AVM with HHT may be lower in the Asian population [4,14]. 36% of cases of single PAVM and 51 - 88% cases of multiple PAVMs are associated with HHT/OWRS whereas contrarily, only 15 - 50% of HHT patients have PAVMs [3,5,11,12]. HHT is transmitted as an autosomal dominant disorder [4,6,12]. Mutations in the endoglin (ENG) gene on chromosome 9 result in HHT 1 phenotype and mutations in activin receptor-like kinase 1 (ACVRL1) gene on chromosome 12 result in HHT 2 phenotype [3,5,6,8,15,19].

Prevalence of PAVMs is related to HHT genotype; one study showed PAVMs on CT scan in 58% of patients with ENG mutations whereas PAVMs were seen in only 18% patients with ACVRLI mutations (P < 0.001) [5,6].

PAVMs are categorized into simple and complex types [3,5,12]. 80% of PAVMs are of simple type whereas 20% is of complex type. Simple PAVM consists of one feeding artery, a non-septated aneurysmal sac, and one or more draining veins. Complex PAVM consists of two or more feeding arteries, a septated aneurysmal sac, and two or more draining veins. In the majority of PAVMs, the feeding artery originates from the pulmonary circulation; however, rarely, it can arise from the systemic circulation, like phrenic, intercostal, bronchial or internal mammary arteries. Draining veins are usually 1 - 2 mm larger than the feeding arteries [17,18]. Diffuse PAVMs and telangiectatic PAVMs are uncommon varieties of complex PAVMs [3,5,12]. Multiple and diffuse PAVMs are more frequently found in patients with HHT [3,6]. Approximately 53–70% of PAVMs are based in the lower lung lobes [1,3,5,12].

Majority of patients (~72%) present with manifestations akin to PAVM or underlying HHT. Clinical expressions of PAVM depend on the extent of right-to-left shunt and usually appear between the 4th - 6th decades of life when the load of shunted blood excels 25% of total blood volume. Exertional dyspnea is the commonest complaint seen in about 50% of PAVM patients. Other symptoms related to PAVM are clubbing (20%), cyanosis (18%), hemoptysis (10%), chest pain (6%), and systolic murmur (3%) [3,5,12].

Patients with HHT develop small telangiectatic vessels and AVMs at various locations, including nose, skin, lungs, liver, gastrointestinal, and brain and commonly present with repeated epistaxis, anemia, and/or problems related to previously unknown AVMs or PAVMs [6,12]. These patients are also prone to develop pulmonary hypertension [6].

Pulmonary AVMs vary in size from 1 to 5 cm [4]. Pulmonary AVMs smaller than 2 cm in size are usually asymptomatic [4,11].

Most of the patients with PAVMs remain asymptomatic throughout their lifetime; however, due to capillary free anomalous arteriovenous communication with paradoxical embolization, about 30% of patients have neurological complications including migraine, transient ischemic attack, stroke, and cerebral abscess [3,4,6,8,9].

In a recent series, the majority of PAVM-induced cerebral complications (ischemic stroke or brain abscess) occurred in patients with unknown PAVMs [6,19]. A median two-years delay is noted between cerebral event and diagnosis of PAVM, as PAVMs are often not considered in the etiological evaluation of these acute neurological complications [6,9].

A strong association exists between a single feeding artery diameter of >3 mm and various cerebral complications. The chances are higher in cases with multiple PAVMs [3].

There is a history of brain ischemia or abscess in 70% of patients with diffuse PAVMs. In the long term, stroke was reported in 30%, brain abscess in 10%, and pulmonary bleeding/hemothorax in 8 - 10% patients with pulmonary AVMs. Pulmonary arterial hypertension (PAH) was more frequent in patients with a positive family history of HHT [3].

The risk of paradoxical embolization rises with the patient’s age and quantity/perfusion of PAVMs with incidences of 10% below and 45% over age 30 and odds ratio of 4.5 comparing multiple to single lesions [9].

Citation: Ammar Ashraf, et al. "Young Female with Acute Ischemic Stroke Secondary to Paradoxical Embolism from an Undiagnosed PAVM/HHT. Case Report and Literature Review". EC Clinical and Medical Case Reports 3.3 (2020): 01-11.
Observational studies advocate that somewhat 25% of untreated cases of PAVM will suffer an ischemic stroke; hence, diagnosis and
definite treatment of these malformations amenable to endovascular therapy is crucial [8].

Low serum iron, high serum fibrinogen, and low pulmonary artery pressure and are the strongest hazardous aspects for ischemic
stroke up till now [6,9,20]. Iron deficiency and fibrinogen excess promote thrombus formation and low pulmonary pressures facilitate
paradoxical embolism [6,9]. Traditional venous thromboembolism can persuade to ischemic stroke after paradoxical embolization, which
is, however, uncommon in cases of PAVMs [6,20].

Antiplatelet agents are recommended for secondary prevention of PAVMs induced ischemic strokes according to the 2011 AHA (American Heart Association) Stroke Guidelines [6]. Based on these AHA recommendations, the newly reclaimed ebullient platelet aggregation in iron deficiency seems to be a more authentic logical explanation [6,20].

The risk of stroke also depends on the degree/severity of the shunting of PAVM [6]. CT evident PAVMs, as well as paradoxical embolic
events, are more prevalent in patients with high-grade shunts on contrast echocardiogram [6,21]. One study reported a 10.4-fold upsurge
in the risk of stroke/cerebral abscess in patients with grade 3 shunts (PPV of 92.5% for CT-evident PAVMs) and no increased risk for such
complications in patients with grade 1 shunts (PPV of 13.4% for CT-evident PAVMs) [6,21,22].

Conventional risk factors, like diabetes, hypercholesterolemia, hypertension, or cardiac arrhythmias are not blameworthy for ischemic
strokes in patients with PAVM [6,20].

The diagnosis of hereditary hemorrhagic telangiectasia (HHT) is affected by geological and socioeconomic factors and may be chal-
lenging [6,23]. With the increasing understanding of HHT, the described percentage of PAVM cases associated with HHT boosted from
approximately 70% (before 1998) to 93.6% (in 2008) in one series, with 59% of patients having no prior diagnosis of HHT [6]. Transthoracic
contrast echocardiography (TTCE) is a minimally invasive, safe, simple, universally available modality with high sensitivity and low false-
negative rate and is recommended as an initial screening modality for PAVM by international HHT guidelines committee [3,5,6,24,25].
It has a sensitivity of 95 - 100% [3]. It is considered positive for intra-cardiac shunt when the injected bubbles are seen in the left heart
chamber after one or two cardiac cycles and positive for extra-cardiac/intrapulmonary shunt when these microbubbles are seen in the left
heart chamber after 3-8 cardiac cycles. A positive TTCE with a normal CT chest is virtually diagnostic of a microscopic PAVM. Depending
on its grading system; TTCE can predict the risk of cerebrovascular complications. Grade 1 shunt (<30 microbubbles) does not have any
associated increased risk of CNS complications whereas grade 2 (30 - 100 microbubbles) and 3 (>100 microbubbles) pulmonary shunts
are independent predictors of CNS complications (ischemic stroke and abscess) [3,26]. Echocardiography results also depend on the type
of HHT; in one study, 85% of patients with HHT type 1 had positive contrast echocardiography (CE) whereas 35% of patients with HHT
type 2 had positive CE [6]. In a combined Dutch–Italian study, grade 3 shunt had a positive predictive value (PPV) of 92.5% for CT evident
PAVMs in comparison to a PPV of only 13.4% for the grade 1 shunt [6,22].

Radiological investigations evaluate PAVM nature, size, site, number, and appropriateness for possible endovascular embolization
[6,27]. Chest radiography is an economical, basic, quick, widely available and comparatively low radiation imaging modality for initial
screening of a PAVM. PAVM may be easily seen on chest x-ray; however, its sensitivity is low in picking up small lesions and even can miss
10 - 40% of clinically significant PAVMs [3,6,18]. Radiographically a typical PAVM presents as rounded well-delineated nodular opacity
of variable size (usually of 1 - 2 cm) with branching feeding and draining vessels from lesion towards the hilum whereas a complex PAVM
usually appears as an ill-defined lesion with dilated, branching and tortuous feeding and draining vascular channels [3,5].

At present, the thin section contrast-enhanced multidetector CT (MDCT) chest is the imaging modality of choice in diagnostic confir-
mation and management planning of PAVM [3,24]. On CT scan, a simple classical PAVM presents as a peripheral well-circumscribed spher-
Young Female with Acute Ischemic Stroke Secondary to Paradoxical Embolism from an Undiagnosed PAVM/HHT. Case Report and Literature Review


Precise shunt quantification is possible with a technetium perfusion scan; however, it is no more used in the detection or surveillance of PAVMs [6].

Currently suggested diagnostic, screening and management protocol is summarized in figure 4.

**Figure 4:** Flow chart for screening, diagnosis, management, and follow-up of pulmonary arteriovenous malformation [6].
Young Female with Acute Ischemic Stroke Secondary to Paradoxical Embolism from an Undiagnosed PAVM/HHT. Case Report and Literature Review

Imaging differential diagnosis of PAVM

Pulmonary artery aneurysm, Pulmonary vein varix, Calcified granuloma, Bronchocele [3].

Treatment

Definitive treatment is indicated in all patients, except for those with no evidence of HHT and asymptomatic cases with small PAVMs [3]. Before the 1980s, surgical resection was the only available treatment option for AVMs [4]. Due to the recent developments in angiographic techniques, currently, transcatheter embolization is the first-line treatment for all PAVMs who can undergo embolization [4,6,29]. The risk of stroke does not depend on the size of PAVMs and thus, transcatheter embolization (TCE) is suggested for all PAVMs [3].

Transcatheter embolization (TCE) using different techniques including coils, balloons, or Amplatzer vascular plugs, is a secure and efficient strategy in impeding the hemorrhagic and embolization-related complications [2,3]. Most isolated small-to-moderate size PAVMs can be completely occluded with coils and large size PAVMs with Amplatzer vascular plugs [1,12,30]. Several reports of successful transcatheter embolization of large pulmonary AVMs are present in the literature [2,4,30].

Amplatzer vascular plug is a relatively modern embolization device made of a self-expanding cylindrical nitinol mesh that is especially suitable for the embolization of large vessels with high flow or vessels with a short landing zone. Several recent reports have described its use for the treatment of PAVMs. Different types (AVP1, AVP2, AVP3, and AVP4 devices) varying in size and configuration are available. The diameter of the elected AVP should be mildly larger (~1.5-2 times) than the size of the feeding artery [2,12].

Amplatzer vascular plugs have advantages of complete occlusion of large feeding vessels with single device (thus decreasing radiation exposure and procedure time), easier occlusion of the neck of the sac, decreased requisite for accurate sizing of the AVP, and occlusion of a shorter length of vessel, thereby decreasing the chances of occlusion of normal lung vascularity and are now preferred over coiling for embolization [2,6,8,27]. PAVMs should be embolized as close as possible to the arteriovenous communication to avoid recurrence and revascularization of PAVMs [2,4,6].

Successful embolization occludes the feeding vessels and leads to the regression of pulmonary AVMs, the reversal of size of former feeding and draining vessels, resolution of the right-to-left shunt, and prevention of embolic complications [4,6,8,9,11,31]. More recent literature shows diminution in migraines [6], strokes [6,20], and decrease in ventilatory [6,31], cardiac [6,31,32], and hematologic (hemoglobin) [6,33] demands. The risk of infective embolic complications also decreases; however, the lifelong obligation of prophylactic antibiotics before dental and surgical treatment persists [2]. After successful embolization, the PAVM does not develop neo-vascularity from adjacent blood vessels; however, revascularization has been mentioned in the literature with variable rates, which is the reason for life long follow up [6,8,9,27].

Multiple reports have shown good immediate results and remote benefits; however, post-embolization PAVM recurrence or sac persistence can exist in up to 25% of cases [2,6]. Post-embolization persistent PAVM sacs may be either secondary to reopening of previously occluded feeding arteries (more commonly) or maybe due to the evolution of new arterial feeders from pulmonary or uncommonly systemic circulation [6,27]. These new feeders are less feasible than re-canalized feeding arteries to successive re-embolization [6,27]. According to one study recanalization was more likely to occur if coils were used singly or placed >1 cm from the PAVM sac [6]. There are also reports in the literature showing higher recanalization rates in childhood [6,24].

Transient pleurisy is the commonest complication of TCE and is seen in about 10% of patients [1,2,6,34]. Pleurisy may occur without pulmonary infarction and is more frequently seen in patients with peripheral or diffuse types of PAVMs [2,6]. Other potential complications are paradoxical embolism of devices or thrombi, air embolism, device migration, spontaneous balloon deflation, and hemoptysis [1,6,34].

Citation: Ammar Ashraf, et al. “Young Female with Acute Ischemic Stroke Secondary to Paradoxical Embolism from an Undiagnosed PAVM/HHT. Case Report and Literature Review”. EC Clinical and Medical Case Reports 3.3 (2020): 01-11.
Peculiar attention should be paid in case of a history of severe pulmonary hypertension as PAVM embolization is more precarious and can cause an acute lethal increase in pulmonary artery pressure [6]. Severe pulmonary hypertension persists a relative contraindication to elective embolization; however, it needs further evidence, as these are more likely to get a symptomatic benefit from PAVMs embolization [6,33].

Surgery (local excision, lobectomy, or pneumonectomy) is occasionally recommended in patients with large pulmonary AVMs having multiple complex large feeding arteries, which are not amenable to transcatheter embolization; however, there is no definite threshold for the surgical indication [4]. In general, all those cases who have contraindication of embolization should undergo surgery [1,8,11,35-37].

Follow up

After embolization, HHT patients need long term follow-up and continued surveillance to detect recanalization or growth of PAVM [3,5,8,25]. Current international guidelines advocate MDCT chest angiogram 6 - 12 months after embolization and then every 3 years thereafter [3,5,6,8,25]. In patients with small untreated and microscopic PAVMs; follow up period and frequency is decided on a case-by-case basis with a follow-up CT chest about every 1 - 5 years [3,5,6,25]. TTCE can be falsely positive in patients with treated PAVMs and is no longer used for follow up screening [8].

Patients who already have an ischemic stroke secondary to PAVM and do not undergo embolization, need secondary prevention with long term anticoagulation, although supportive evidence is lacking [1,8].

Conclusion

Pulmonary arteriovenous malformations (PAVMs) are rare, anomalous direct communications between the pulmonary artery and the pulmonary vein, thereby resulting in an extra-cardiac intrapulmonary right-to-left shunt. Paradoxical embolism leading to stroke or brain abscess is the major complication of this intrapulmonary shunt and the majority of these complications occur in patients with previously undiagnosed PAVMs. About 15% of ischemic strokes are cryptogenic, and rare causes of stroke are often overlooked, particularly in young patients without any evidence of more common etiologies. Our case is a Hellenic example of a rare cause of stroke and highlights the importance of considering PAVM in the differential diagnosis of cryptogenic stroke, particularly in young individuals. Transthoracic contrast echocardiography (TTCE) is the preferred screening investigation, contrast-enhanced computed tomography (CT) chest is the gold standard imaging technique to establish the diagnosis of PAVM, and endovascular embolization of the feeding artery is the treatment of choice of PAVMs.

Compliment for the Cardiologist

"Our appreciation is extended to Dr. Anwar Jelani (consultant cardiologist) for providing this awesome echocardiogram."

Bibliography


Citation: Ammar Ashraf., et al. "Young Female with Acute Ischemic Stroke Secondary to Paradoxical Embolism from an Undiagnosed PAVM/HHT. Case Report and Literature Review". EC Clinical and Medical Case Reports 3.3 (2020): 01-11.


