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CASE REPORT

Arteriovenous Malformation Pulmonary (AVM) in a Post-Cesarean Woman: Intensive Care and Urgent Surgery Operation

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The AVM is a rare congenital disease that often affects the brain and only rarely we find in other organs. In this pathology there is an alteration of the vascular system, with the arteries discharging directly into the veins, bypassing the capillary system; this puts the patient at high risk of sudden hemorrhage. Today we present the case of a young woman suffering from unknown pulmonary avm. The breakdown of this avm caused a hemothorax in the immediate post-cesarean, necessitating emergency surgery and subsequent intensive care.

Keywords: Arteriovenus Malformations; Pulmonary AVM; Post-cesarean; Hemotorax; Hereditary haemorrhagic telangiectasia

Introduction

Arteriovenous Malformations (AVMs) are malformations of the vascular system where the arteries, usually hypertrophic, discharge directly into one or more veins bypassing the capillary system (**Figure 1**). This process puts veins at higher pressures than the normal and increases the chance of a break with consequence hemorrhage (Shovlin & Gossage 2017). AVMs are rare, congenital, and often tardy disorders; they usually involve the brain, and rarely affect other organs.

80% of pulmonary MAVs are related to hereditary haemorrhagic telangiectasia (HHT) or Rendu-Osler-Weber Syndrome, an autosomal dominant pathology affecting 1–5/8000 people and whose main symptoms are dyspnoea and frequent epistaxis with possible multiple MAV to lungs, liver and digestive tract (Rotenberg et al. 2017).

Clinical case

We are presenting the case of a 33-year-old woman at the 39th week of gestation. In her anamnesis there was no former notable condition, except for a hormonal stimulation for infertility problems for 1 year. The patient came to ours observation from another hospital after a urgent caesarean section, because she displayed a severe dyspnea, hypoxia, and a significant chest pain on arrival at ER; those symptoms led colleagues to suspect pulmonary embolism and to treat her using heparin infusion, but we don't know the reason for this therapy.

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After she was transferred to our hospital, we performed chest CT which highlighted hemotorax with a subtotal atelectasia of the left lung (**Figure 2**), for breakdown of a pulmonary AVM of the basal area (**Figures 3** and **4**). Furthermore, we performed embolization procedure on AVMs arteries and alerted the thoracic Surgery Unit for to position chest drain, which resulted in an evacuation of approximately 1300 cc of blood.

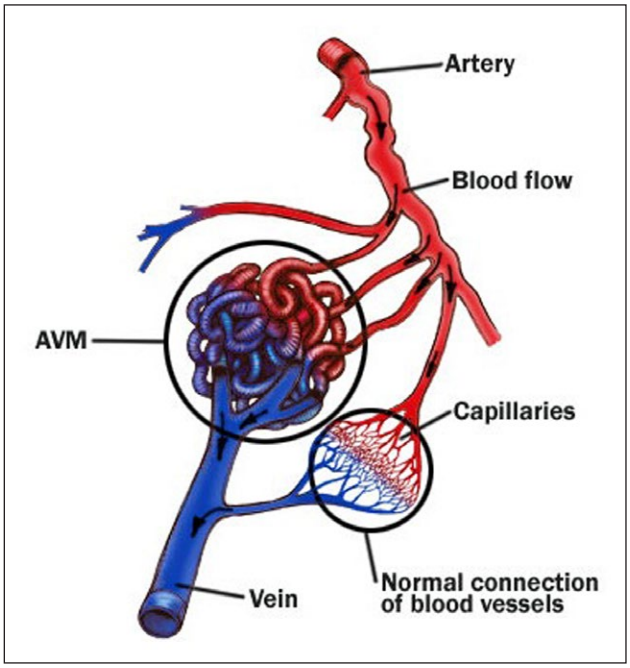

Figure 1: Structure of the AVM.

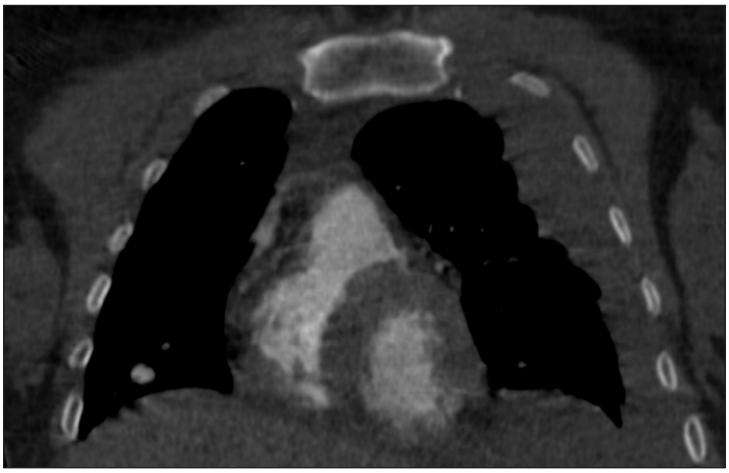

Figure 2: CT image of the hemothorax.

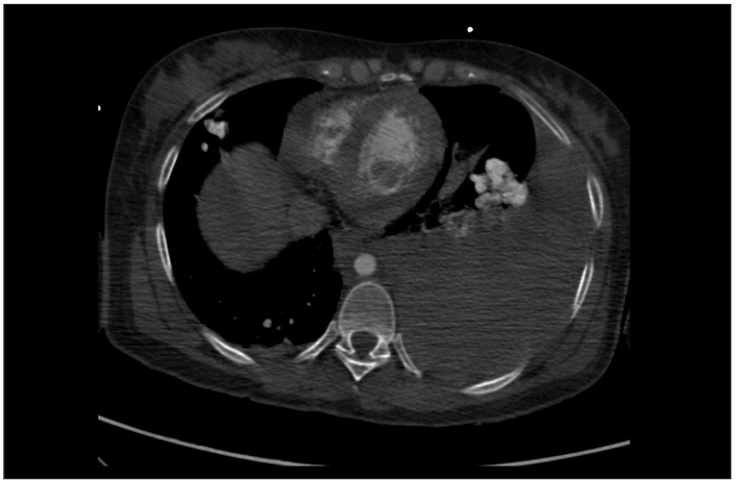

Figure 3: CT image of the AVM, transversal scan.

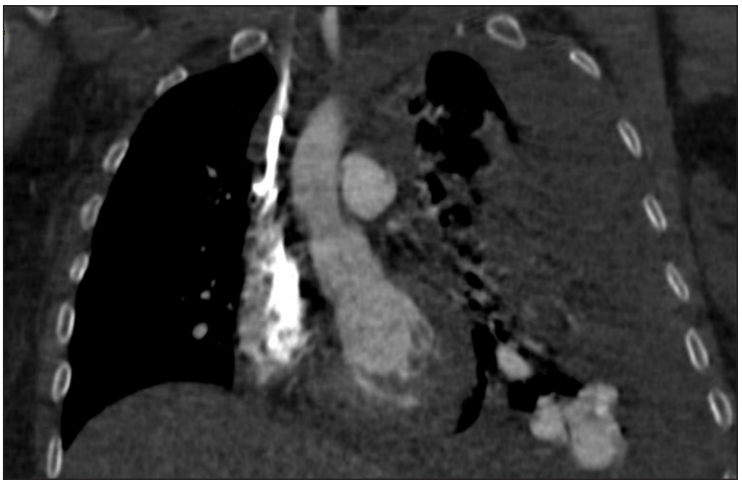

Figure 4: CT image of the AVM, front scan.

The gynecological unit is also alerted for probable hysterectomy, since uterine bleeding is impossible to stop. In addition, blood tests, serial blood gases and TEG are performed for patient monitoring (**Figures 5** and **6**). However, during her hospitalization in Intensive Care she started to bleed again from chest drain, which resulted in a hemodynamic instability.

According to the clinical presentation, chest surgeons immediately performed emergency thoracotomy in intensive care in order to produce hemostasis, and then an atypical resection of the left-lung lower lobe was performed in the operating room, together with gynecologists for hysterectomy. The operation required numerous transfusions of concentrated blood cells and plasma.

	05/04/17 40400823	04/04/17 50519721	04/04/17 50519572	04/04/17 40400534	03/04/17 50519503	03/04/17 50519405	03/04/17 40400269	02/04/17 50519317	02/04/17 50519252	02/04/17 50519219	02/04/17 50519153	01/04/17 50519106	01/04/17 50519038	01/04/17 5051899
Esame emocromocitometrico						Postoperative							Preoperative	
CBC+DIFF (POLI)														
Eritrociti	2.78	2.69	2.49	2.41	2.62	2.84	2.79	2.85	3.03	2.36	2.80	2.74	2.02	3.15
Emoglobina	8.4	8.1	7.5	7.2	8	8.4	8.6	8.8	9.3	7.2	8.6	8.4	6	9.5
Ematocrito	25.4	24.3	22.8	21.8	23.6	25.2	24.9	25.6	27	21	24.8	24.5	17.8	27.8
Volume Corpusculare Medio	91.2	90.6	91.5	90.3	89.9	88.6	89.2	90.0	89.3	89.1	88.5	89.4	88.2	88.1
Emoglobina Corpusculare Media	30.2	30.1	30.2	29.9	30.5	29.7	30.7	30.9	30.6	30.6	30.5	30.7	29.6	30.2
Concentrazione di Emoglobina Cor...	33.1	33.3	33.0	33.1	33.9	33.5	34.5	34.3	34.3	34.3	34.5	34.3	33.5	34.2
RDW-CV	13.8	13.4	13.7	13.5	13.7	13.7	13.7	13.4	13.5	13.4	13.3	13.4	13.5	13.7
Piastrine	110	92	90	97	89	88	69	69	66	58	59	44	55	88
Volume Medio Piastrinico	9.8	9.4	10.0	10.1	10.0	10.1	11.1	10.5	10.1	10.1	9.9	9.8	9.7	9.9
Leucociti	8.96	10.58	11.55	10.90	12.87	11.55	9.63	9.89	9.36	8.02	11.28	11.84	12.89	23.22
Neutrofili %	94.3	83.9	86.7	83.7	90.7	89.8	83.7	84.0	84.5	84.2	82.6	86.5	88.7	86.9
Linfociti %	4.0	11.5	9.6	12.3	6.1	5.8	9.9	9.1	8.2	9.1	10.0	6.7	5.8	6.1
Monociti %	1.6	3.8	3.5	3.6	3.1	4.2	6.3	6.8	7.1	6.7	7.3	6.7	5.5	6.9
Eosinofili %	0	0.6	0.1	0.3	0	0.1	0.1	0	0.1	0	0	0	0	0
Basofili %	0.1	0.2	0.1	0.1	0.1	0.1	0	0.1	0.1	0	0.1	0.1	0	0.1
Neutrofili #	8.44	8.88	10.02	9.11	11.68	10.36	8.05	8.31	7.91	6.75	9.31	10.25	11.43	20.15
Linfociti #	0.36	1.21	1.10	1.34	0.78	0.67	0.96	0.90	0.77	0.73	1.13	0.79	0.74	1.42
Monociti #	0.15	0.40	0.41	0.40	0.40	0.49	0.61	0.67	0.66	0.54	0.83	0.79	0.71	1.61
Eosinofili #	0.0	0.07	0.01	0.04	0.0	0.01	0.01	0.0	0.01	0.0	0.0	0.0	0.0	0.01
Basofili #	0.01	0.02	0.01	0.01	0.01	0.02	0	0.01	0.01	0	0.01	0.01	0.01	0.03

Figure 5: Serial Blood counts, pre and post operative.

	07/04/17 40401352	06/04/17 40401082	05/04/17 40400823	04/04/17 50519721	04/04/17 50519572	04/04/17 40400534	03/04/17 40400269	02/04/17 50519317	02/04/17 50519252	02/04/17 50519219	02/04/17 50519153	01/04/17 50519106	01/04/17 50519045	01/04/17 5051899
Esame della coagulazione						Posoperative							Preoperative	
Profilo PT														
PT - INR	1.18	1.09	1.07	1.15	1.24	1.22	1.22	1.28	1.23	1.21	1.24	1.31	1.55	1.34
PT - Ratio	1.18	1.1	1.07	1.15	1.24	1.23	1.23	1.28	1.23	1.22	1.24	1.31	1.56	1.35
APTT - Secondi	38	32	34	35	35	33	32	29	28	30	33	41	49	34
APTT - Ratio	1.22	1.03	1.09	1.14	1.14	1.05	1.03	0.94	0.9	0.95	1.06	1.34	1.57	1.09
Fibrinogeno di CLAUSS	282	384	506	425	335	365	261	246	227	203	149	121	104	116
ATIII - Antitrombina	93	101	101	75	87	83	101	102	82	79	48	57	59	64
D-Dimero	679	429	809	954	822	1014	242	337	368	473	360	450	813	1130

Figure 6: Coagulation profile, pre and post operative.

In the post-operative period, RX Chest control and Brain MRI are performed, which excludes cerebral MAV, but highlights areas of cortical distress, necessitating neurological counseling with an EEG examination; the result was coma from diffuse encephalopathy. Therefore medical assistance is continued in collaboration with the neurology unit with progressive improvement of the clinical picture, which makes it possible on the twelfth day to proceed to respiratory wearing with extubation of the patient.

Unfortunately, focal seizure to bilateral diffuction complicated post-operative hospitalization in intensive care, and were likely to be a consequence of the initial hypoxia in ER, but they are promptly treated with appropriate medical treatment.

We were finally able to transfer the patient, who showed a total neurologic recovery, to the Neurology ward after 26 days of intensive care hospitalization (**Figures 7** and **8**). Therefore, on a clinical basis, neurologists hypothesized a Rendu-Osler-Weber Syndrome, which is in an ongoing process of assessment in a specialized center.

Discussion

Hereditary Hemorrhagic Telangiectasia, also known as Osler-Weber-Rendu syndrome, is an autosomal dominant disorder that causes abnormal blood vessel formation. The diagnosis of hereditary hemorrhagic telangiectasia is clinical. Genetic mutations that have been identified include ENG, ACVRL1/ALK1, and MADH4/SMAD4, among others. At least 50% of patients with HHT develop pulmonary AVMs (Kritharis, Al-Samkari & Kuter 2018).

Then pulmonary AVM is a rare and often asymptomatic disease that can break and cause haemorrhage during pregnancy. This is due to the particular physiology that characterizes the gestation itself; increase of blood volume, cardiac output and venous distensibility (Di Crescenzo 2015). All factors that determine a rapid growth of MAV.

Moreover, to what extent the heparin infusion negatively impacted the case?

Due to the lack of literature of similar case studies of lung AVMs related to hormone therapy and heparin infusion, we are not able to ascertain how these procedures impacted the evolution of the clinical presentation.

Esame emocromocitometrico						
Eritrociti	3.54	↓	10^6 /µL	3.70	-	5.10
Emoglobina	10.5	↓	g/dL	11.7	-	16
Ematocrito	32.4	↓	%	36	-	48
MCV	91.7		fL	80.0	-	99.0
MCH	29.6		pg	26.0	-	33.5
MCHC	32.2		g/dL	31.0	-	35.6
RDW-CV	16.0	↑	%	11.5	-	14.5
Piastrine	272		10^3 /µL	150	-	400
MPV	8.5		fL	7.2	-	11.1
Leucociti	6.66		10^3 /µL	5.20	-	12.40
Neutrofili %	59.1		%	40.0	-	74.0
Linfociti %	27.6		%	19.0	-	48.0
Monociti %	8.8		%	3.4	-	9.0
Eosinofili %	3.6		%	0	-	7
Basofili %	0.9		%	0	-	1.5

Figure 7: Last blood count before transfer.

Esame della coagulazione					
P-Tempo di protrombina (INR)	1.1		0.8	-	1.2
P-Tempo di protrombina (Ratio)	1.1		0.8	-	1.2
P-APTT	30	sec	24	-	36
P-APTT (Ratio)	0.96		0.8	-	1.2

Figure 8: Last coagulation profile before transfer.

The only certain thing is the importance of collaboration between the various medical disciplines for the resolution of these cases, almost always characterized by respiratory failure and emergency caesarean section (Sood, Sood & Dhawan 2011).

Conclusion

From the acquired experience we can state how the MAV of the lung represents a serious eventuality, whose resolution is purely surgical, especially in those cases in which the embolization has not proved efficient (Biçakçioğlu 2017). But even as we do not immediately think of rare diseases, such as hereditary hemorrhagic telangiectasia, often unknown.

Competing Interests

The authors have no competing interests to declare.

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