Magnetic Resonance Imaging of AMS (Aneurysm of the Membranous Septum), review of the literature and case report

Ernesto Di Cesare[1*], Alessandra Di Sibio[1], Giuseppe Lanni[1], Antonio Gennarelli[1], Carlo Masciocchi[1]

1. Department of Radiology, University of L'Aquila, Italy

* **Correspondence:** Prof. Ernesto Di Cesare, Department of Radiology, University of L'Aquila, Via Vetoio 1, 67100 L'Aquila, Italy (*ernesto.dicesare@cc.univaq.it*)

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ABSTRACT

Aneurysm of the Membranous Septum (AMS) is a rare cardiac disease, mostly associated with other cardiac anomalies, very rare in the absence of other congenital heart defects. A prompt diagnosis is important, due to severe potential complications, but remain challenging. Most of the cases were earlier diagnosed using ventriculography, but, with the availability of echocardiography and cardiovascular magnetic resonance (CMR), this disease can be accurately assessed non-invasively. We report a case of a 62 years old female patient, without other cardiac congenital disease, who was incidentally diagnosed, by means of CMR with a true and isolated AMS. Our report underlines CMR usefulness in AMS diagnosis, thanks to accurate evaluation (both morphologic and functional) provided by this diagnostic tool, which is able to demonstrate clearly the presence of AMS (aneurysm of the membranous septum) and depict its features.

CASE REPORT

CASE REPORT

We describe a case in which a female patient, without other cardiac congenital defects, was incidentally diagnosed with a true and isolated Aneurysm of the Membranous Septum (AMS) by means of CMR (Cardiovascular Magnetic Resonance).

A 62 years old female patient underwent a CMR in our department after a clinic suspicion of an acute myocarditis. Acute myocarditis was confirmed by CMR and resolved in some weeks. Her medical history was moreover unremarkable.

CMR was performed using a 1,5T General Electric Medical System. Steady State Free Precession (SSFP) images were acquired in short axis and four chambers. Myocardial LE (Late Enhancement) images were acquired 10 / 15 min following the intravenous administration of contrast medium (BOPTA, MultiHance) in short axis and four chambers.

Imaging Findings:

Images showed a saccular formation (20 x 15 mm), with regular contours and a wide base, arising from membranous septum inferior to the aortic valve and protruding into the right ventricular outflow tract. SSPF sequences clearly demonstrated flow from the Left Ventricular Outflow Tract into the formation through the superior portion of the membranous septum, without any shunt flow between the ventricles. SSFP sequences showed also an outpouching or windsock appearance due to aneurysmal distention during ventricular systole. There was neither evidence of any thrombus nor evidence of hyperintensity in LE sequences.

On the basis of the above information, this structure was confidently referable to a true AMS.

Evaluation of short axis sequence through a post - processing software (Segment®) showed regular parameters: left EDV (end diastolic volume) 100 ml, left ESV (end

systolic volume) 35 ml, left EF (ejection fraction) 67%; right EDV 90 ml, right ESV 40 ml, right EF 55%.

Management and follow-up:

According to the guidelines of the European Society of Cardiology (ESC), our patient received no treatment but was planned a strictly CMR follow-up.

DISCUSSION

Etiology & demographics:

Aneurysm of the membranous septum (AMS) is a rare disease, mostly associated with other cardiac anomalies [1], that occurs in 0.3% of patients with congenital heart disease, in up to 19-22,4% with ventricular septal defect [2,3] and in 20% with perimembranous ventricular septal defects [3]. AMS is a very rare condition in the absence of ventricular septal defect [2], such in our case. Despite recent developments in the diagnosis of congenital heart disease, it is still difficult to determine the prevalence of these anomalies [3].

AMS involves the membranous portion of interventricular septum [4] and, although described as an aneurysm, doesn't appear to be a true aneurysm [5]. AMS, as in our case, is a well - developed, thickened fibrous - walled sac, arising from the right ventricular aspect of the membranous septum, beneath the septal leaflet of the tricuspid valve, sometimes perforated by one or many holes, bulging forward into the right ventricle [6,7]. Absence of myocardium leads to the aneurysm - like behaviour of this structure [8]; the characteristic outpouching or windsock appearance results from aneurysmal distention during ventricular systole [9].

AMS are generally classified as true, false and pseudoaneurysms [3]. AMS with regular contours and a wide base are termed true AMS, as in our case, while those with a narrow base and irregular shape are named false AMS [3]. A pseudo-AMS usually occurs as a complication of myocardial infarction, in which the necrotic muscular wall protrudes into the right ventricle, or as a complication of transaortic septal myotomy [3].

The etiology of this anomaly is poorly understood [9]. AMS may develop as a consequence of spontaneous closure, partial or complete, of a ventricular septal defect [2]; other theories suggest either abnormal embryologic development or weakness of the affected tissues [9].

Clinical & imaging findings

AMS may further enlarge [6] and patients may incur potential cardiac complications, such as aortic valve prolapse, right ventricular outflow obstruction, tricuspid valve insufficiency, arrhythmia, rupture, thromboembolism and bacterial endocarditis [2].

True AMS is difficult to identify angiographically, ultrasonographically and pathologically [3]. The first postmortem observation of AMS was reported by Laennec in 1826 [10], Steinberg made the first antemortem diagnosis using ventriculography in 1957 [11]. Conventional left ventriculography is a widely used method, that can well demonstrate aneurysm of the membranous interventricular septum but it bears limitation because of its invasive nature; in addition, detecting the thrombus within the aneurysm is not possible [12]. Nowadays AMS is often detected incidentally during non - invasive imaging (echocardiogram) [13]; nevertheless AMS could not be optimally visualized [13] and detailed morphological assessment is sometimes obscure [12], especially in patients in whom precordial windows are too poor to clearly visualize the cardiac anatomy [5]. According to Canale et al. the echocardiogram sensitivity is 57% in the subcostal four chamber view, 62% in the short axis view, 71% in the apical four chamber view, 87.5% in the long axis view [13]. CMR is a useful diagnostic tool both for morphological and functional assessment because it is capable, as in our case, of three-dimensional anatomical assessment and provides functional data about the blood flow into the aneurysm and integrity of the ventricular membranous septum [13]. CMR is able, as in our case, to demonstrating clearly AMS presence and depict its features (size, absence of holes and / or thrombus) without limitation due to poor acoustic window, that may impair a clear echocardiographic approach.

Phase-contrast MRI is able to quantify the severity of abnormal communication between the cardiac chambers by calculating the ratio between pulmonary and systemic flow (Qp/Qs) [14,15]. PC MRI was found to have 93% specificity and 100% sensitivity for description of shunts with a Qp:Qs greater than 1.5 in comparison with invasive oxymetry [15]. Transthoracic and transesophageal echocardiography usually allow exact localization and sizing of the defect but determination of shunt volumes by Doppler echocardiography has limitations [16]. In general, Doppler echocardiography can provide excellent assessment in smaller, younger children while MRI plays a more important role in the appraisal of the older, larger adolescent or adult with congenital heart disease, especially those that have undergone previous cardiothoracic surgery [16].

Multislice CT is another diagnostic tool, which has similar advantages in morphological and functional assessment compared to CMR, but CMR is preferable because able to provide functional data, without exposure of radiation [13].

Differential Diagnoses:

The differential diagnosis of AMS includes anatomically related aneurysms and aneurysmal-like structures arising in and around the left ventricular outflow tract, such as sinus of Valsalva aneurysm [17]. AMS arises from the right ventricular aspect of the membranous septum, beneath the septal leaflet of the tricuspid valve, and bulging forward into the right ventricle [6,7]. It is possible to make a definitive diagnosis only by demonstrate flow from the Left Ventricular Outflow Tract into the AMS through the superior portion of the membranous septum [17]. On the contrary, the criteria for diagnosing a Valsalva sinus aneurysm include an origin above the aortic annulus, a saccular shape and normal dimensions of the adjacent aortic root and ascending aorta [18]. Although angiography is considered the reference standard for

confirming the presence of a Valsalva sinus aneurysm, most are initially seen at color Doppler echocardiography [18]. Multiplanar MR imaging with sequences also allows accurate assessment of the origin and size of Valsalva sinus aneurysms and the status of the surrounding cardiac and mediastinal anatomy [18]. The advantages of performing MR imaging in the setting of a known or suspected Valsalva sinus aneurysm include the ability to evaluate the left ventricular hemodynamic pattern, identify aortic regurgitation, and quantify any aortocardiac shunt or turbulent or fistulous blood flow [18]. CT provides detailed anatomic depiction of Valsalva sinus aneurysms and surrounding cardiac structures but MR imaging can be performed without exposing the patient to ionizing radiation or iodinated contrast material, which is a further fundamental advantage [18].

Treatment & prognosis:

Patients with AMS who are asymptomatic should be followed closely in terms of potential cardiac complications [2]; European Society of Cardiology (ESC) states that follow-up at 3 to 5 year intervals may be reasonable [19]. Surgery is usually considered in AMS cases complicated with either thrombus and related systemic emboli, rupture or accompanying significant VSD [13].

Thus, as in our case, information provided by CMR are useful for planning patient care and follow - up, also thank to high reproducibility of CMR data.

TEACHING POINT

Aneurysm of the Membranous Septum is a thickened fibrous - walled sac arising from the right ventricular aspect of the membranous septum and bulging forward into the right ventricle; absence of myocardium leads to the aneurysm like behaviour of this structure. It is possible to make a definitive diagnosis by assessing its origin and demonstrating flow from the Left Ventricular Outflow Tract into the aneurysm through the superior portion of the membranous septum.

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FIGURES

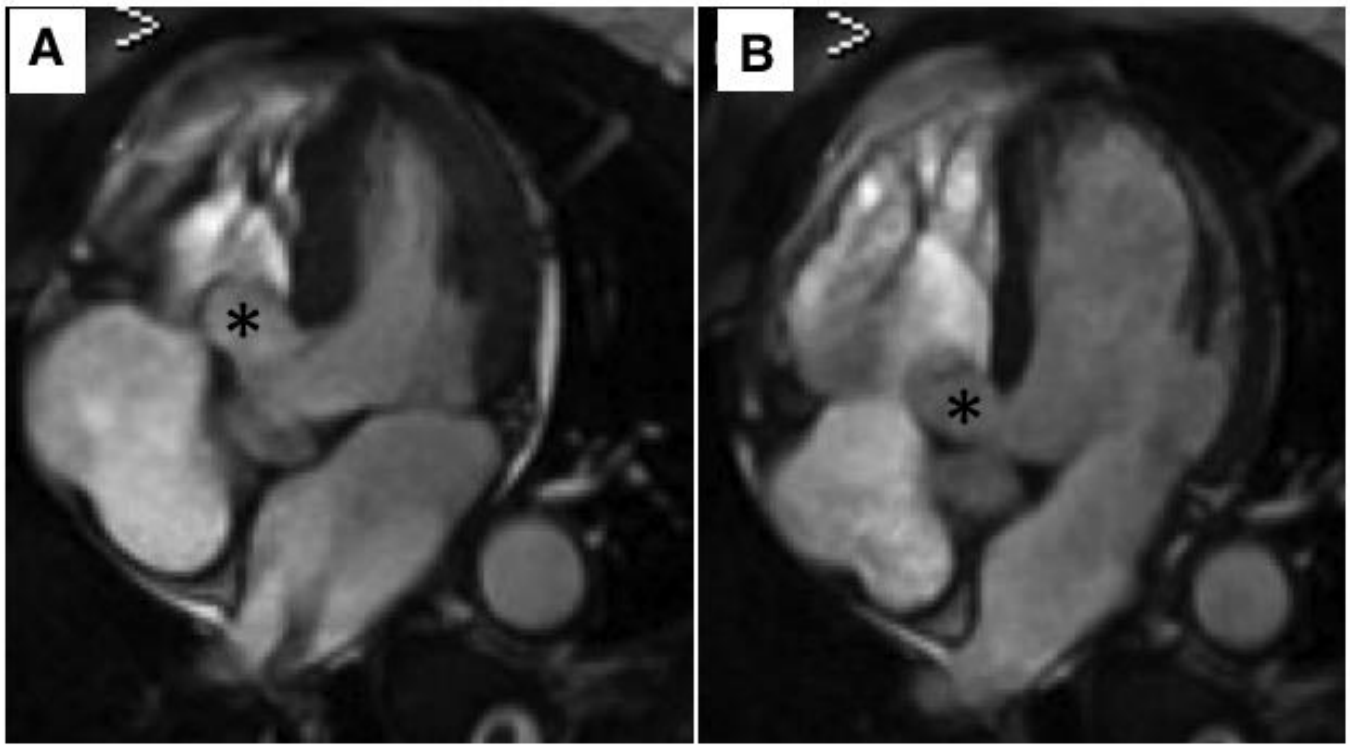

Figure 1: 62 year old female with aneurysm of the membranous septum.
Technique: MRI 1,5T; Steady State Free Precession (SSFP) four chambers view; TR (repetition time) 3,6 msec, TE (echo time) 1,6 msec, Flip angle 65°, Slice thickness 4mm, Image matrix 256 x 256.
Finding: Steady State Free Precession (SSFP) four chambers view in systole (a) and diastole (b) show an AMS (20 x 15 mm) arising from membranous septum beneath the septal leaflet of the tricuspid valve, bulging into right ventricle, without any thrombus or shunt flow between the ventricles (asterisk).

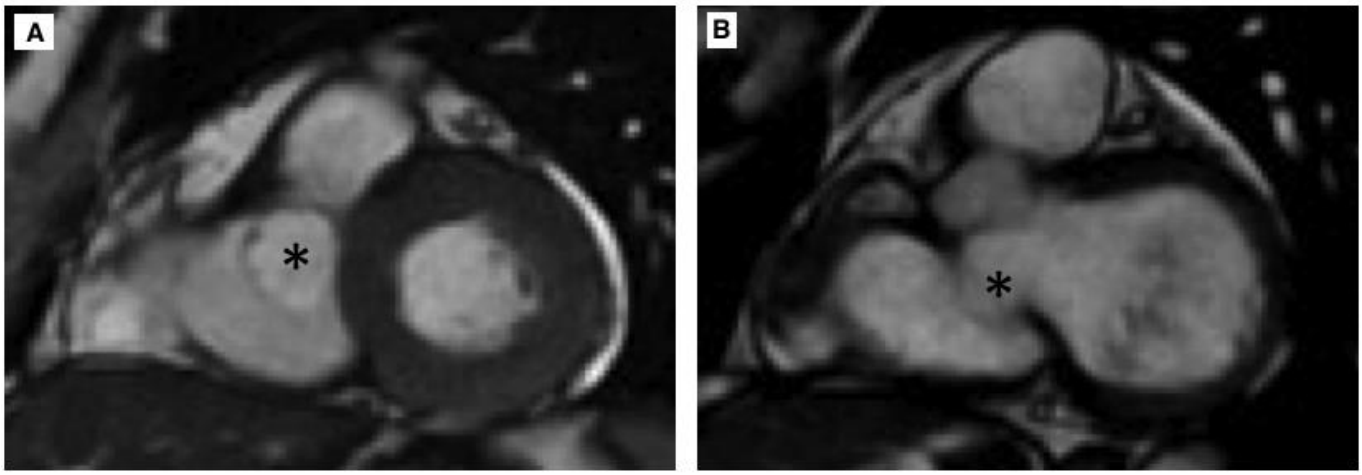

Figure 2: 62 year old female with aneurysm of the membranous septum.
Technique: MRI 1,5T; Steady State Free Precession (SSFP) short axis view; TR (repetition time) 3,6 msec, TE (echo time) 1,6 msec, Flip angle 65°, Slice thickness 4mm, Image matrix 256 x 256.
Finding: Steady State Free Precession (SSFP) short axis view in systole (a) and diastole (b) show AMS (asterisk) adjacent to interventricular septum, inferior to the aortic valve, protruding in right ventricular chamber.

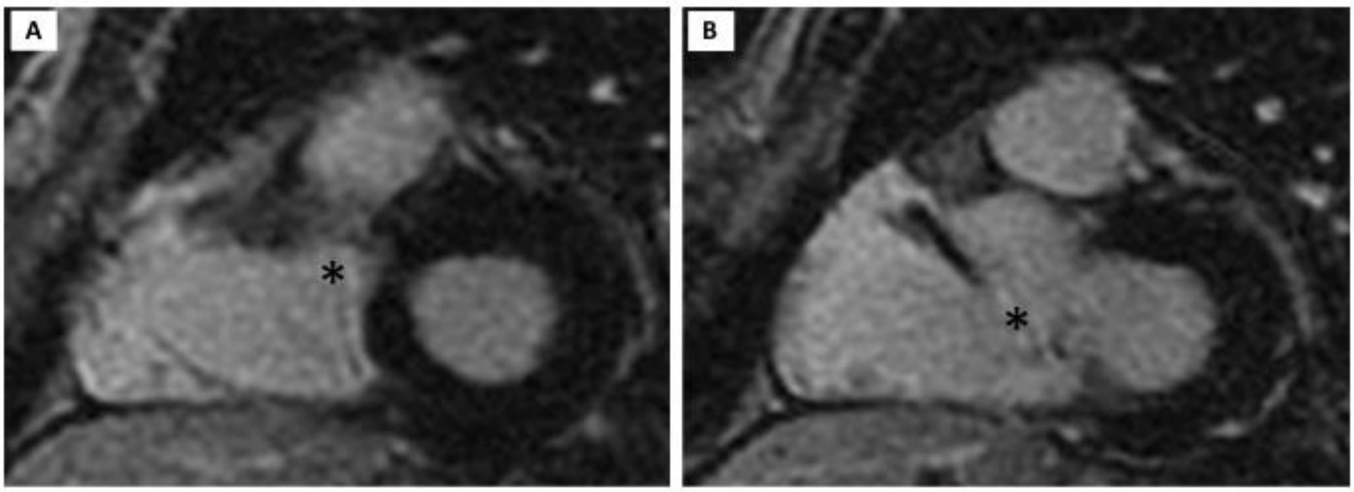

Figure 3: 62 year old female with aneurysm of the membranous septum.
Technique: MRI 1,5T; myocardial LE (Late Enhancement) short axis view acquired 10 / 15 min following the intravenous administration of contrast medium (BOPTA, MultiHance); TR (repetition time) 3,6 msec, TE (echo time) minimum, TI (Inversion Time) 180 msec, Flip angle 20°, Image Matrix 256 x 256.
Finding: myocardial LE (Late Enhancement) short axis view in systole (a) and diastole (b) show AMS (asterisk) without evidence of hyperintensity.

Incidence	Rare, although it is still difficult to determine its prevalence. AMS occurs in 0.3% of patients with congenital heart disease, in up to 19-22,4% with ventricular septal defect and in 20% with perimembranous ventricular septal defects.
Etiology	AMS develops, more often, as a consequence of spontaneous closure of a ventricular septal defect.
Findings on imaging	• AMS appears as a saccular formation arising from membranous septum inferior to the aortic valve and protruding into the right ventricular outflow tract. • AMS has regular contours and an outpouching appearance during systole. • AMS is often detected incidentally during echocardiogram but morphological assessment may be difficult. • Doppler echocardiography may identify shunt flow between the ventricles. • SSPF MRI sequences demonstrate AMS presence and describe its feature, also show flow from the Left Ventricular Outflow Tract into the AMS through the superior portion of the membranous septum. • PC MRI is able to quantify the severity of abnormal communication between the cardiac chambers and AMS. • CT is able to demonstrating clearly AMS presence and features but bears limitation due to exposure of radiation. • Ventriculography can well demonstrate AMS but it is an invasive procedure and is not able to demonstrate a thrombus within the aneurysm.
Prognosis	AMS may further enlarge. Patients may incur potential cardiac complications, such as aortic valve prolapse, right ventricular outflow obstruction, tricuspid valve insufficiency, arrhythmia, rupture, thromboembolism and bacterial endocarditis.
Treatment	Patients with AMS who are asymptomatic should be followed closely while surgery is usually considered in cases complicated with concurrent heart diseases, hemodynamic abnormalities and / or symptoms.

Table 1: Summary table for AMS (Aneurysm of the Membranous Septum)

	Aneurysm of Membranous Septum (AMS)	**Valsalva Sinus Aneurysm (VSA)**
US	AMS is often detected incidentally during echocardiogram but morphological assessment may be difficult. Doppler echocardiography may identify shunt flow between the ventricles.	Most VSA are initially seen at color Doppler echocardiography but global assessment may be difficult.
CT	CT is able to demonstrating AMS presence and features but bears limitation due to exposure of radiation	CT is able to demonstrating VSA presence and features but bears limitation due to exposure of radiation.
MRI SSPF	SSPF MRI sequences demonstrate AMS presence and describe its feature. Also SSFP sequences show flow from the Left Ventricular Outflow Tract into the AMS through the superior portion of the membranous septum.	MRI SSFP sequences allow accurate assessment of the origin and size of VSA. Also, SSFP sequences identify any aorto-cardiac shunt or fistulous blood flow.
MRI PC	PC MRI is able to quantify the severity of abnormal communication between the cardiac chambers and AMS.	MRI PC quantifies any aorto-cardiac shunt or fistulous blood flow.
MRI LE	In some cases, AMS shows hyperintensity in LE sequences.	In some cases, VSA shows hyperintensity in LE sequences.
Ventriculography	Ventriculography can well demonstrate AMS but it is an invasive procedure and is not able to demonstrate a thrombus within the aneurysm.	Ventriculography is considered the reference standard for confirming the presence of a VSA but it is an invasive procedure.

Table 2: Differential diagnoses for AMS (Aneurysm of the Membranous Septum)

AMS TYPE	Characteristics
True	Is a congenital disease. Has regular contours and a wide base.
False	Is a congenital disease. Has irregular shape and a narrow base.
Pseudoaneurysm	Is an acquired disease occurring as a complication of myocardial infarction or of transaortic septal myotomy.

Table 3: Classification of AMS (Aneurysm of the Membranous Septum) types

ABBREVIATIONS

AMS = Aneurysm of the membranous septum
CMR = Cardiovascular Magnetic Resonance
EDV = End Diastolic Volume
EF = Ejection Fraction
ESV = End Systolic Volume
FIESTA = Fast Imaging Employing Steady State Acquisition
IVS = Interventricular Septum
LE = Late Enhancement
PC = Phase Contrast
SSFP = Steady State Free Precession
TE = Echo Time
TR = Repetition Time
VSA = Valsalva Sinus Aneurysm

KEYWORDS

aneurysm of the membranous septum; interventricular septum; cardiovascular magnetic resonance

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