CASE 1

Right atrial pseudotumor due to crista terminalis

Stefan L. Zimmerman

Imaging description

The crista terminalis is a smooth, vertically oriented, muscular ridge within the posterior wall of the right atrium (Figure 1.1). It is located at the site of embryonic fusion of the trabeculated and smooth-walled portions of the right atrium. The smooth-walled portion is also known as the sinus venosus. The crista terminalis is a critical structure for the cardiac conduction system, containing the sinoatrial node superiorly, and a frequent location of atrial tachyarrhythmias.[1] Most often the crista terminalis has low attenuation on computed tomography (CT) and is isointense to the right atrial wall on magnetic resonance imaging (MRI) (Figure 1.1).[2] In patients with lipomatous hypertrophy of the interatrial septum, the crista terminalis may also be enlarged and will include fat,[3] which will be low attenuation on CT (Figure 1.2) and high signal on bright blood and T1-weighted MRI images. In these cases, an etching artifact may also be recognized on bright blood steady-state free precession images, due to the interface of crista terminalis fat and the right atrial wall (Figure 1.3). The fat within an enlarged crista terminalis is contiguous with periatrial fat.

Importance

A prominent crista terminalis can be mistaken for a right atrial mass.[4] This could lead to inappropriate therapy and the associated risks of that therapy; for example, increased bleeding risk if anticoagulation is used in cases of suspected thrombus, or unnecessary open heart surgery if mistaken for myxoma.

Typical clinical scenario

The size of the crista terminalis has an average thickness of 4.5 mm; however, it is highly variable in size and can range from almost imperceptible to prominent and polypoid, projecting into the lumen of the right atrium.[1,4] It is often enlarged in patients with increased periatrial fat and lipomatous hypertrophy of the interatrial septum.

Differential diagnosis

A prominent crista terminalis should be distinguished from right atrial masses. In particular, right atrial thrombus or right atrial myxoma should be considered when an intraluminal polypoid mass is visualized attached to the right atrial wall. The crista terminalis can be distinguished by its characteristic location extending from the superior vena cava (SVC) to the inferior vena cava (IVC), smooth borders, and vertical orientation on coronal reformatted images. When present, fat within an enlarged crista terminalis will also exclude other entities. An atrial myxoma will demonstrate heterogenous enhancement, whereas crista terminalis will show mild enhancement identical to the adjacent atrial wall. Right atrial thrombus will often be found in the setting of a central venous catheter and will not have the characteristic shape or enhancement of the crista terminalis.

Teaching point

A prominent crista terminalis is common and can be identified by its characteristic location, smooth borders, and vertical orientation. It should not be mistaken for a right atrial mass.

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Figure 1.1 A. Axial contrast-enhanced CT image from a study performed in a 64-year-old male with lung cancer shows a prominent crista terminalis (arrow) that is low in attenuation due to fat content and located on the posterior wall of the right atrium. **B.** Multiple contiguous axial images in the same patient demonstrating the vertical course of the crista terminalis from superior to inferior along the posterior wall of the right atrium. **C.** Coronal oblique reformatted image shows the crista terminalis as a vertical, low-attenuation structure that courses superiorly to inferiorly (asterisks), just lateral to the connection of the superior vena cava (arrow) and inferior vena cava (arrowhead) to the right atrium. Note the partially visualized upper lung mass which is compressing the superior vena cava in this patient with known lung cancer.

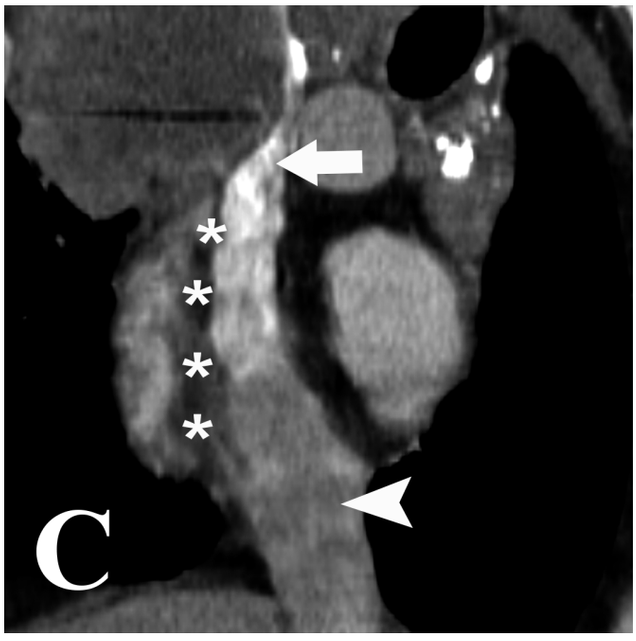

Figure 1.1 (*cont.*)

Figure 1.2 Axial image from contrast-enhanced CT in a 56-year-old woman with cardiac CT performed to evaluate chest pain demonstrates a prominent crista terminalis with a somewhat polypoid appearance (arrow). There is associated lipomatous hypertrophy of the interatrial septum (asterisks), which can be often seen in patients with a prominent crista.

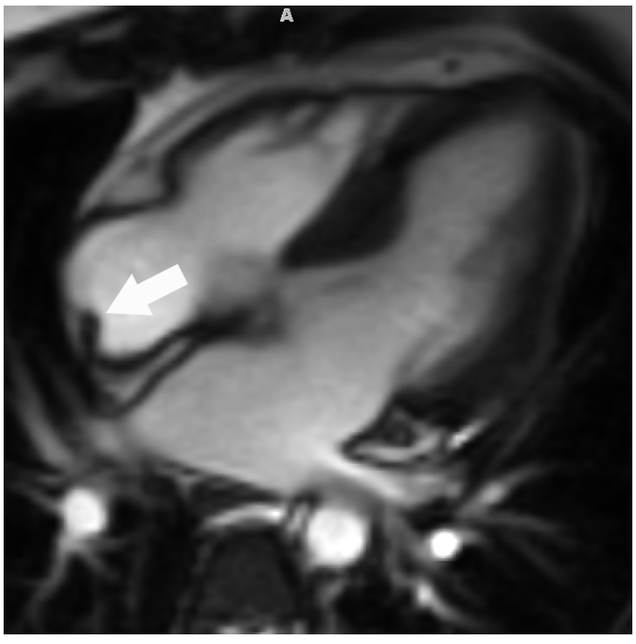

Figure 1.3 Horizontal long-axis bright blood steady-state free precession image from a cine cardiac MRI performed in a 35-year-old female with sarcoidosis shows a prominent crista terminalis (arrow), which has an internal high signal and a rim of low signal etching artifact due to the interface of fat with the right atrial wall. The fat of the crista is contiguous with fat within the interatrial septum, also surrounded by etching artifact.

CASE 2

Cardiac pseudotumor due to lipomatous hypertrophy of the interatrial septum

Pejman Motarjem and Stefan L. Zimmerman

Imaging description

Lipomatous hypertrophy of the interatrial septum (LHIS) is a benign process of the heart characterized by fatty infiltration of the interatrial septum.[1] The diagnosis is made when fat in the interatrial septum measures greater than 20 mm in thickness and it is usually an incidental finding at cardiac imaging.[2]

At echocardiography, LHIS is recognized by echogenic thickening of the interatrial septum. On multiple detector computed tomography (MDCT) (with or without contrast) LHIS is a low-attenuation, < 0 Hounsfield units, bilobed mass with smooth margins that spares the fossa ovalis. It is this sparing of the fossa ovalis which gives this entity its characteristic bilobed or dumbbell-shaped morphology (Figure 2.1). Often, there is cranial extension to the level of the cavoatrial junction and fat may surround the distal superior vena cava (Figure 2.2).

On MRI the morphology of LHIS is similar to MDCT. The LHIS demonstrates hyperintensity on T1-weighted imaging with homogenous signal drop out on a fat-suppressed T1 sequence characteristic of macroscopic fat (Figure 2.2). On post-gadolinium sequences no enhancement is seen.[3]

FDG uptake within the atrial septum at positron emission tomography (PET) examinations may be seen, and is attributed to the variable presence of brown fat within LHIS (Figure 2.3).[4] It is important to note that the benign FDG uptake in LHIS must not be mistaken for a malignant process such adenopathy or metastatic tumor. Fusion PET-CT will help localize radiotracer uptake to the atrial septum and differentiate it from surrounding structures such as the right hilum, pleura or mediastinum. In difficult cases, it may be necessary to correlate PET-CT findings with either MRI or MDCT in order to prevent inappropriate staging of the patient.

Importance

The condition of LHIS is a benign incidental finding and typically does not cause any symptoms. Since it may demonstrate increased FDG uptake on PET/CT, it must not be confused with a malignant process, leading to misdiagnosis, inappropriate follow-up imaging or inappropriate biopsy.

Fat infiltration in the atrial septum increases with age, correlates with increased epicardial fat in obese patients and is contiguous with epicardial fat deposits.[5] The LHIS process has been associated with atrial arrhythmias, thought to be the result of disruption of atrial conduction pathways.[2] In rare cases, LHIS results in right atrial and superior vena cava partial obstructive symptoms requiring surgical resection.[6,7]

Typical clinical case scenario

Lipomatous hypertrophy of the interatrial septum is typically encountered incidentally on echocardiogram, MDCT, MRI, or PET/CT. It most often has a benign course with estimated prevalence of 1% at autopsies, 2–8% on echo[1] and 2.2% on MDCT.[2] Once encountered, no further follow-up imaging is recommended. As discussed above, in rare cases LHIS may lead to hemodynamic obstruction or may cause atrial arrhythmias, but in the overwhelming majority of the cases it is completely benign causing no symptoms.

Differential diagnosis

The main differential consideration of LHIS is cardiac lipoma (Figure 2.4). Unlike lipoma, LHIS lacks a true fibrous capsule. In addition, primary cardiac lipomas are exceedingly rare.

Liposarcoma is a malignant fatty lesion that is usually large and will present with aggressive features such as invasion into surrounding cardiac or mediastinal structures and mass effect.

Other benign tumors such as myxoma, fibroma and rhabdomyoma do not contain fat. Teratomas may have fat features on CT or MRI but they will usually have a mix of tissue types including soft tissue, bone or teeth. They also lack the typical bilobed morphology characteristic of LHIS.

Melanoma metastasis will demonstrate high signal on T1-weighted imaging similar to LHIS, but unlike LHIS it will not lose signal on the fat-suppressed sequence and will avidly enhance on post-contrast sequences.

Teaching point

Lipomatous hypertrophy of the interatrial septum is typically an incidental finding at cardiac imaging characterized by fat and a characteristic dumbbell or bilobed shape. In the overwhelming majority of the cases it is asymptomatic and requires no further follow-up. Although it may demonstrate FDG uptake due to presence of brown fat, it must not be mistaken for a malignant or neoplastic process.

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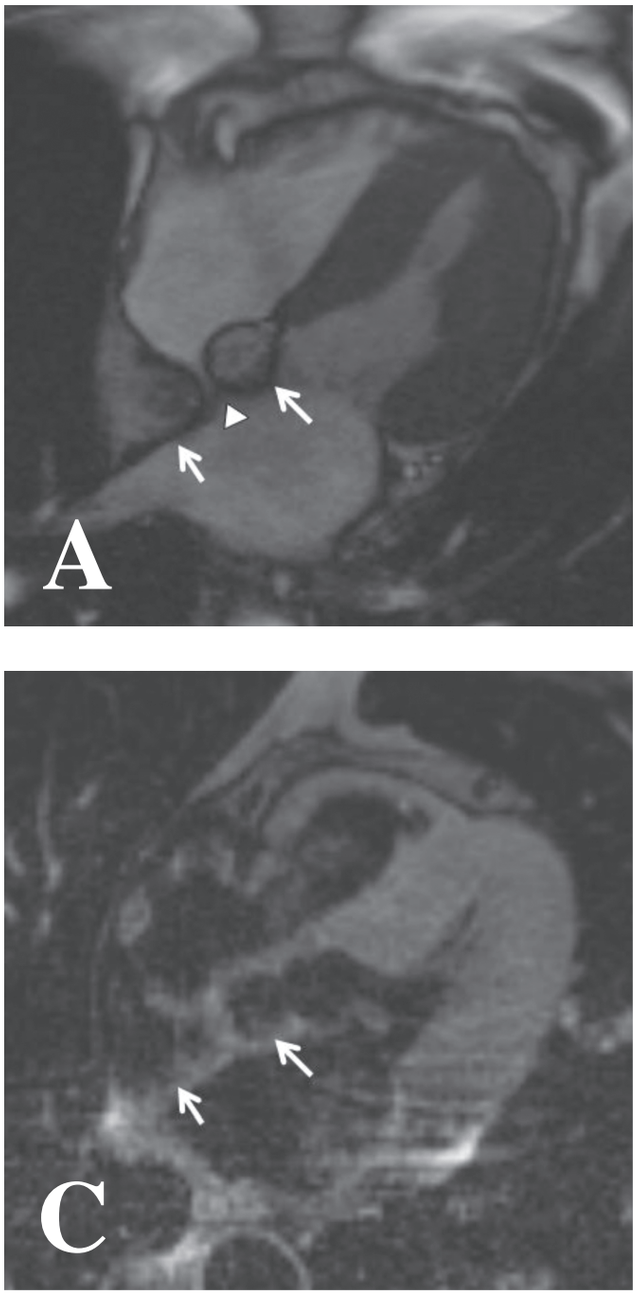

Figure 2.1 Typical imaging appearance of lipomatous hypertrophy of the interatrial septum (LHIS) on MRI. **A.** Horizontal long-axis bright blood image obtained in a 67-year-old male with a question of right atrial mass on echocardiography demonstrates a characteristic dumbbell-shaped mass (arrows) of the atrial septum with sparing of the fossa ovalis (arrowhead). Atrial thickness was 22 mm. **B.** Axial T1-weighted black blood image shows high-signal intensity in the mass (arrows) **C.** Fat-saturated post-contrast T1-weighted image demonstrates signal drop out in the atrial mass (arrows) indicating macroscopic fat.

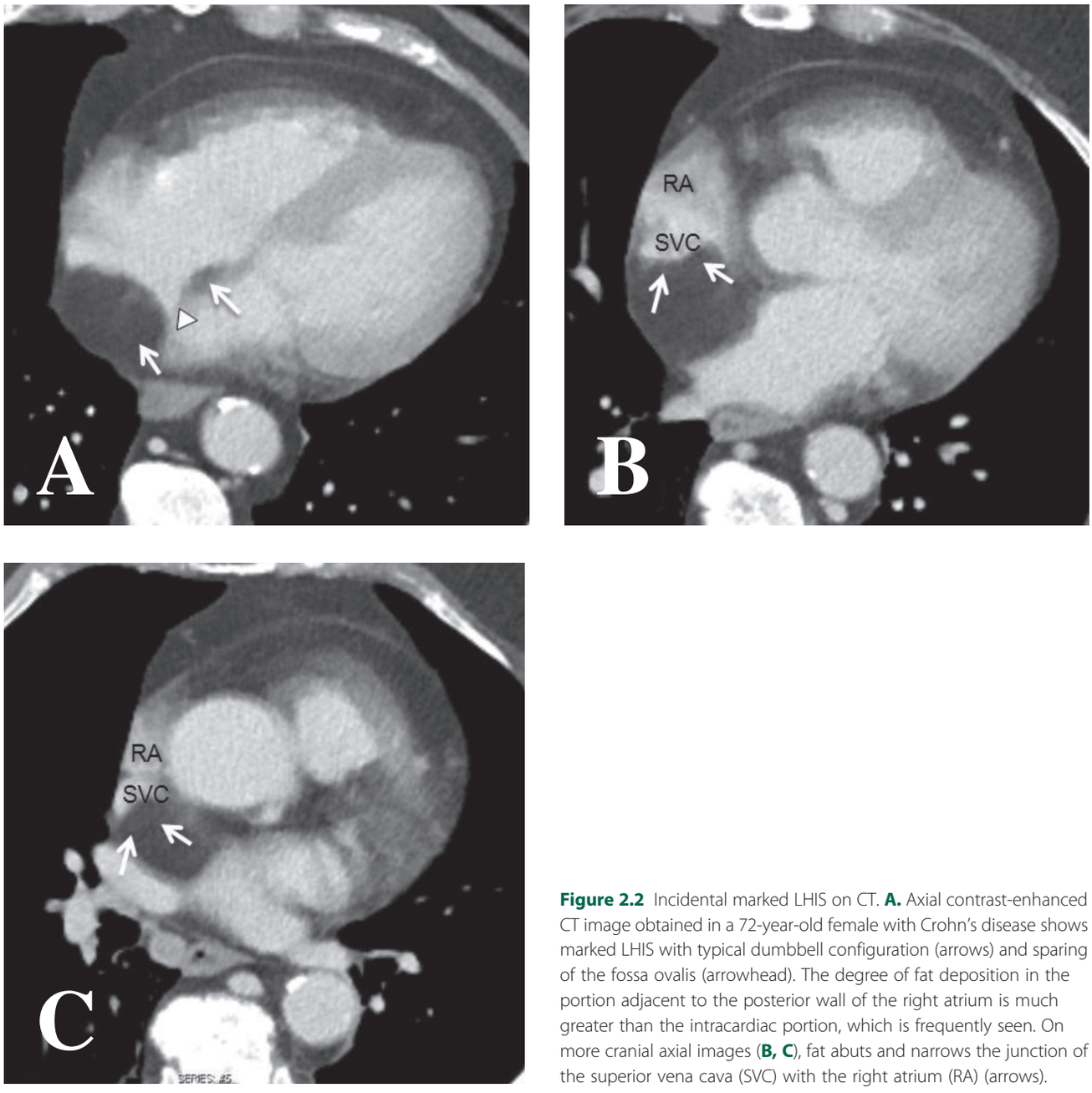

Figure 2.2 Incidental marked LHIS on CT. **A.** Axial contrast-enhanced CT image obtained in a 72-year-old female with Crohn's disease shows marked LHIS with typical dumbbell configuration (arrows) and sparing of the fossa ovalis (arrowhead). The degree of fat deposition in the portion adjacent to the posterior wall of the right atrium is much greater than the intracardiac portion, which is frequently seen. On more cranial axial images (**B, C**), fat abuts and narrows the junction of the superior vena cava (SVC) with the right atrium (RA) (arrows).

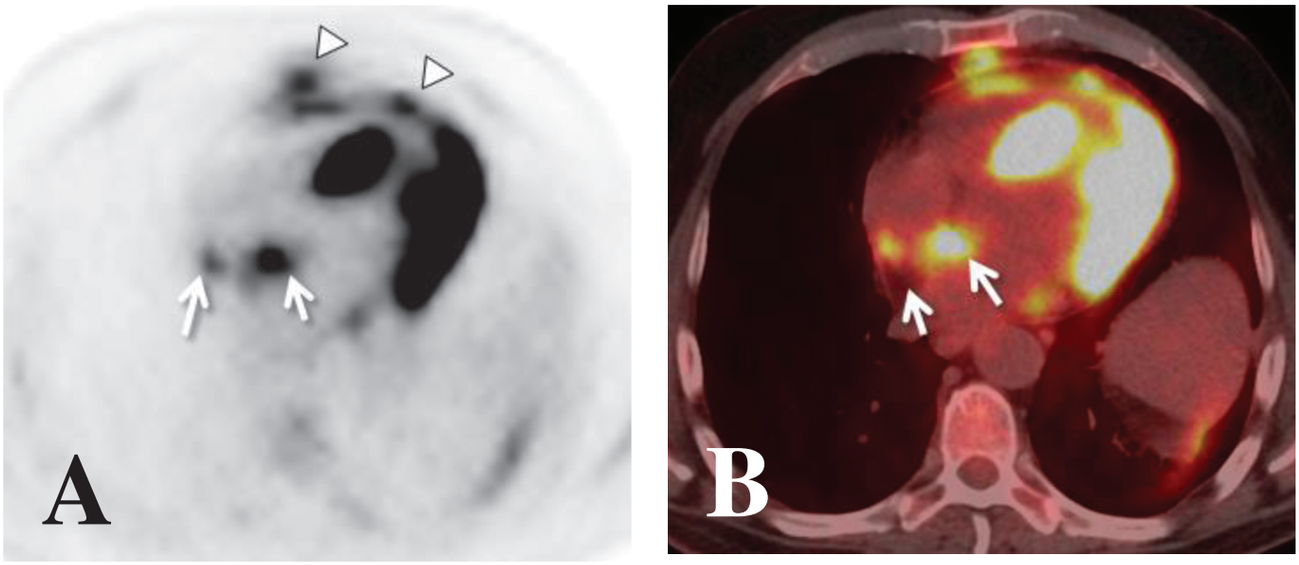

Figure 2.3 Atrial septal hypermetabolism due to brown fat. **A.** Axial image from FDG-PET examination performed in a 59-year-old male with lymphoma involving the pericardium demonstrates increased radiotracer uptake in the heart posteriorly (arrows) and along the pericardium anteriorly (arrowheads). **B.** On fused PET-CT images, the foci of increased uptake correlate with fat in the interatrial septum (arrows). These represent regions of hypermetabolic brown fat, not uncommonly seen in the interatrial septum, and should not be mistaken for malignancy.

Figure 2.4 Cardiac lipoma. Axial contrast-enhanced CT image of the chest obtained in a 67-year-old male with abdominal pain demonstrates a well-encapsulated, ovoid-shaped fat density mass adjacent to the left atrioventricular groove compatible with cardiac lipoma (arrows).

CASE 3

Cardiac pseudotumor due to caseous mitral annular calcification

Stefan L. Zimmerman

Imaging description

At cardiac MRI (CMR), mitral annular calcification (MAC) is low in signal intensity on bright blood, T1- and T2-weighted images and can appear as a smooth, multilobulated mass or masses in the region of the atrioventricular groove (Figure 3.1). Caseous, also known as liquefactive, MAC is a rare variant that is typically a larger, rounded mass with central liquefactive necrosis, composed of calcium, inflammatory cells, and cholesterol.[1] Given that calcification is not well depicted by MRI, MAC can be mistaken for a cardiac tumor. Caseous MAC can occasionally be large – up to several centimeters in size – and may displace mitral valve leaflets, resulting in valvular dysfunction such as regurgitation or stenosis. On post-contrast images, there is occasionally a thin rim of enhancement due to fibrous tissue surrounding the calcification (Figure 3.2).[2,3] If CT is available, MAC is easy to identify on non-contrast images due to presence of high attenuation calcification (Figure 3.1).[3]

Importance

MAC at CMR can be mistaken for a cardiac tumor or other type of cardiac mass, leading to inappropriate additional testing as well as patient anxiety. Although benign, the presence of MAC is a marker of increased cardiovascular risk. MAC was associated with a 50% greater likelihood of cardiovascular events at follow-up in the Framingham Heart Study and predicted both increased all-cause and cardiovascular death.[4]

Typical clinical scenario

MAC is a common disorder. In the Multi-Ethnic Study of Atherosclerosis, MAC was detected by CT in 9% of a cohort of 6814 subjects from age 45–8.[5] Caseous MAC is more rare, affecting $<$ 1% of subjects with MAC.[1] MAC is often discovered incidentally at echocardiography, and usually requires no further imaging. However, if the diagnosis is uncertain, patients may be referred to advanced imaging with MRI or CT.

Differential diagnosis

MAC should be differentiated from a true cardiac tumor. Location in the mitral annulus, low T1 and T2 signal intensity, and lack of enhancement are distinguishing characteristics. Myxomas are the most common primary cardiac tumor and can be located near valves; however, unlike MAC, will have increased T2 signal and heterogenous enhancement.[6] Cardiac fibroma can have a low signal on T1 and T2-weighted images due to fibrous content; however, it typically demonstrates either homogenous or heterogenous enhancement and is intramyocardial in location.[6]

Teaching point

Mitral annular calcification may be mass-like in appearance at CMR and can be mistaken for a cardiac tumor. The characteristic location, low signal, and lack of enhancement should suggest the correct diagnosis. For difficult cases, non-contrast CT demonstrating calcification can be used for confirmation.

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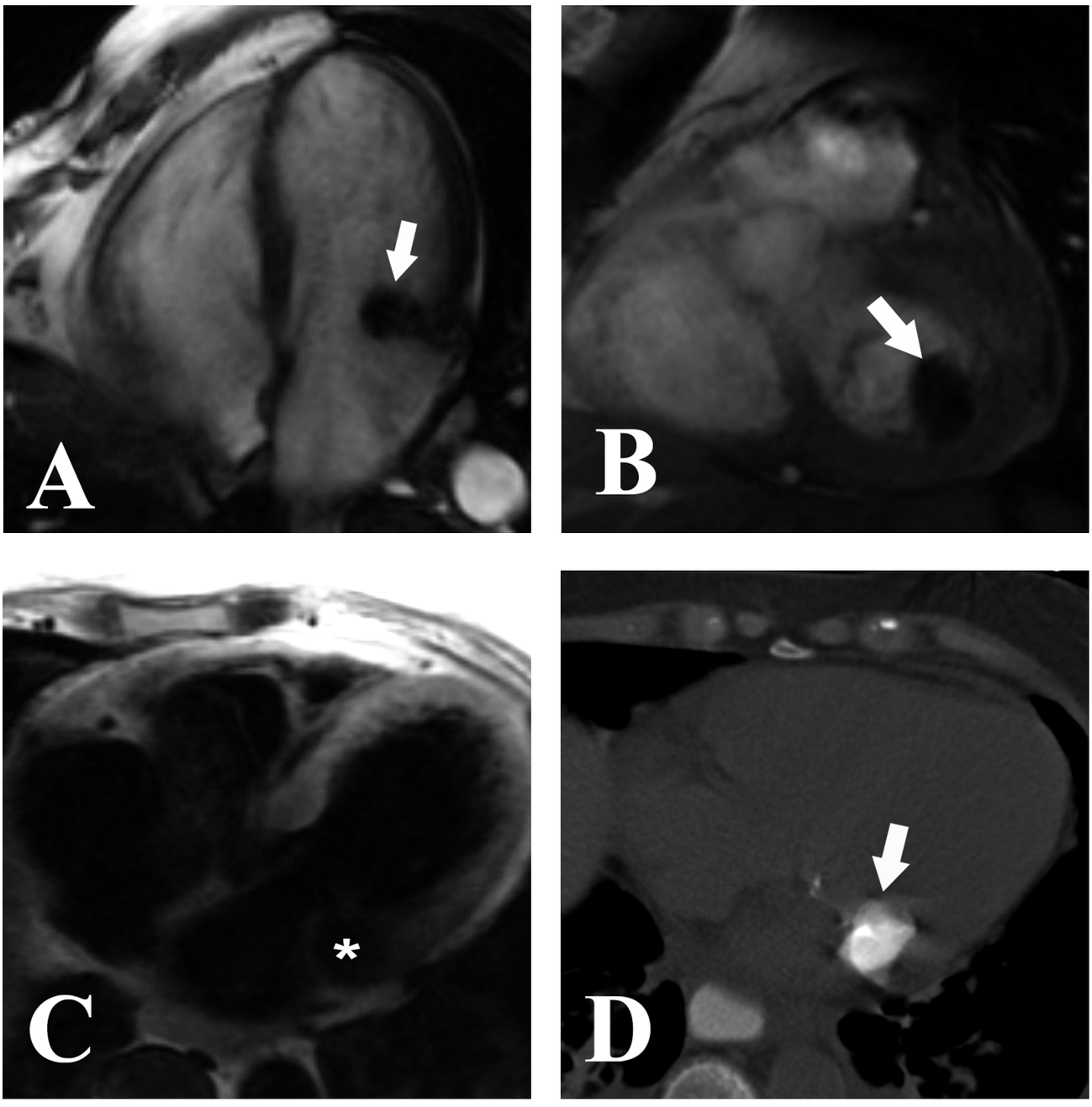

Figure 3.1 **A**. Four-chamber steady-state free precession bright blood image in an 81-year-old female referred to cardiac MRI for a mass found at echocardiography demonstrates a smooth, low signal mass in the region of the mitral annulus (arrow). **B**. Short-axis steady-state free precession bright blood image again demonstrates the low signal, round cardiac mass in the region of the mitral annulus (arrow). **C**. Axial T1-weighted dark blood image obtained demonstrates low signal within the mitral annular calcification (asterisk). **D**. Non-contrast CT examination performed in the same patient demonstrates calcification corresponding to the location of the mass on MRI, diagnostic of mitral annular calcification (arrow).

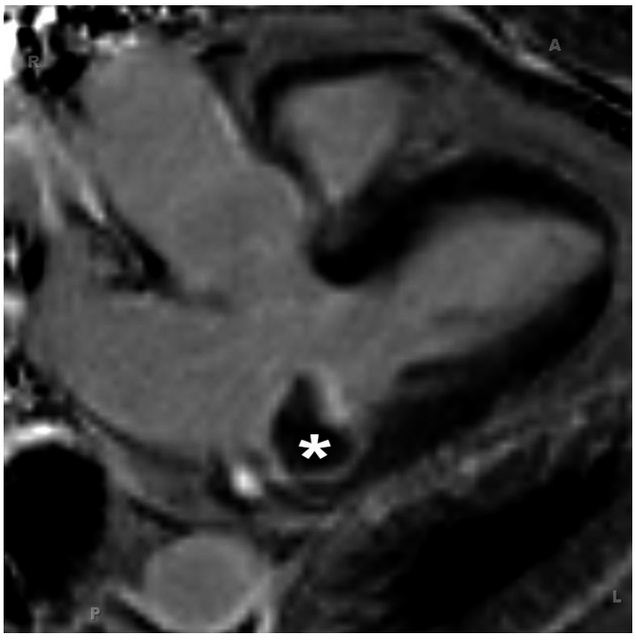

Figure 3.2 Three-chamber delayed enhancement image from an MRI performed in a 61-year-old male with mitral annular calcification demonstrates a low signal intensity mass (asterisk) in the mitral annulus with a characteristic thin rim of delayed enhancement related to fibrous tissue.