Imaging Forum in Cardiovascular Disease of the Korean Society of Cardiovascular Imaging

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The Korean Society of Cardiovascular Imaging (KOSCI) has held monthly imaging forums for the last 20 years. In this note from the KOSCI, we are presenting 4 interesting cases selected from the cases discussed in the KOSCI monthly imaging forums in 2016. The cases are examples of syphilitic aortitis, tuberculous aortitis, primary aortic intimal sarcoma, and solitary fibrous tumor of the pericardium. Through the monthly imaging forums, members can share their experiences with one another. The discussions involving rare diseases and their imaging findings enhance our ability to make proper diagnosis in clinical practice and provide ideas for new research topics. We want to share these opportunities with the readers of CVIA.

Key words Radiology · Cardiac imaging techniques · Computed tomography angiography · Magnetic resonance imaging.

SYPHILIC AORTITIS

A 37-year-old man visited our hospital after 2 months of dyspnea and stable angina. He had no risk factors of cardiovascular disease, and his initial chest X-ray and electrocardiogram were normal. Echocardiogram showed hypokinesia of the mid and basal segments of the anteroseptal myocardial wall. On coronary computed tomography (CT) angiography, diffuse circumferential arterial wall thickening was clearly visible around the aortic root and ascending aorta (Fig. 1A). The left main coronary artery ostium was nearly occluded (Fig. 1B). However, no significant stenotic lesions were observed in distal coronary arteries. Although the complete blood cell counts were normal and other autoimmune disease markers were all negative, the quantitative Venereal Disease Research Laboratory test was 1:8 reactive, and the fluorescent treponemal antibody absorption test was positive. The patient's medical history revealed that he had been treated for a painless penile ulcer (chancre) caused by primary syphilis 7 years previous. He was finally diagnosed with syphilitic aortitis. The patient underwent a per-

cutaneous transluminal coronary angioplasty (PTCA), and a 3×14-mm-sized drug eluting stent was deployed in the left main coronary artery ostium (Fig. 2). With penicillin treatment, the patient's symptoms and physical condition were improved.

Syphilis is a sexually transmitted disease caused by the spirochete *Treponema pallidum* [1]. Syphilitic aortitis is caused by tertiary syphilis and usually occurs 10–25 years after initial infection [2]. Cardiovascular syphilis is categorized with uncomplicated syphilitic aortitis, aneurysmal change of the aorta (40%), aortic insufficiency (29%), and coronary ostial stenosis (26%) [2].

The initial histological change manifests as perivascular plasma cells and lymphocytic infiltrates of the aortic adventitia, which result in endarteritis obliterans and adventitial narrowing and lead to ischemic medial necrosis and destruction of elastic fibers [3]. Syphilitic coronary ostial stenosis is caused by thickened intima and adventitia by fibrous tissues [3]. The severity of stenosis of the coronary artery ostium is associated with thickening of the aortic wall. Penicillin has been the first line of therapy for treatment of syphilis [1]. There is no standardized treatment for coronary ostial stenosis, and coronary artery bypass grafting has been regarded as an effective treatment in recent years [4]. However, as in our case and in the literature [5], PTCA with stenting is an additional treatment choice

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for syphilitic coronary artery ostial stenosis. Although less frequent, syphilitic aortitis should be considered in patients with coronary ostial lesions with aortitis.

TUBERCULOUS AORTITIS

A 50-year-old woman presented to the hospital with left shoulder

and back pain that had lasted for 1 year and had been aggravated 1 day prior. Contrast-enhanced CT of the thorax revealed a small outpouching contrast-filling sac at the thoracoabdominal aorta, suggesting saccular aneurysm with low attenuated hematoma (Fig. 3). There was no remarkable finding in the lung parenchyma except a tiny granuloma in the right upper lobe. The adjacent aortic wall demonstrated a slightly ir-

Fig. 1. Coronary CT angiography (A) of a 37-year-old man with syphilitic aortitis shows diffuse circumferential wall thickening at the ascending aorta (arrows), and a multi-planar reformatted left coronary artery image (B) shows an ostium (arrows) nearly occluded by thickened aortic wall. The distal left coronary artery is normal.

Fig. 2. Selective coronary angiography (right anterior oblique view 20°, caudal 20°) results of the left coronary artery (A) show occlusion at the left main ostium (arrow), and there is no residual stenosis after deploying a drug-eluting stent (3×14 mm) at the left main coronary artery ostium (B).

Fig. 3. A tuberculous pseudoaneurysm of the thoraco-abdominal thoracic aorta in a 50-year-old woman. Axial (A) and reformatted coronal contrast-enhanced CT images (B) at the mediastinal window setting show a small saccular pseudoaneurysm with a surrounding low attenuated hematoma in the descending thoracic aorta.

Fig. 4. A tuberculous pseudoaneurysm of the abdominal aorta in a 50-year-old woman. Axial (A) and reformatted coronal contrast-enhanced CT images (B) at the mediastinal window setting, performed 8 months after the images in Fig. 3, show a small contrast-filling out-pouching sac with an irregular peripheral enhancing mass-like lesion abutting the abdominal aorta, suggesting a pseudoaneurysm with associated concealed rupture.

Fig. 5. Lower extremity CT angiography of a 74-year-old man with aortic intimal sarcoma shows an irregular, eccentric, large intraluminal mass in the descending aorta on an axial image.

regular appearance. We diagnosed the patient with ruptured saccular aneurysm at the thoraco-abdominal aorta and performed resection of the aortic aneurysm and replacement with a rifampin-soaked Hemashield straight graft.

Eight months later, the patient revisited the hospital due to fever and abdominal pain. At the time of admission, her vital signs were stable except for fever (38.3°C). Laboratory tests revealed high C-reactive protein (11.9 mg/dL) and normal white blood cell count (7,700/mm³). On the CT evaluation, a small outpouching pseudoaneurysm and an about 6.5-cm-sized irregular peripheral enhancing mass-like lesion were found at the abdominal aorta (Fig. 4). Based on diagnosis of mycotic pseudoaneurysm associated with concealed rupture, we performed a second operation of abdominal aortic replacement with rifampin-soaked Hemashield straight graft. In the operation field, abscess pocket was associated with abdominal aortic perforation. Pathologic examination of the resected specimen showed

Fig. 6. MR angiography of the aorta shows high signal intensity with internal intermediate signal intensity on T2WI (A), peripheral enhancement after gadolinium contrast injection (B) of the mass, and the shape of the mass is irregular in dynamic enhancement image and 3D reconstruction images (C and D).

chronic granulomatous inflammation and multinucleated giant cells, and polymerase chain reaction of mycobacterial tuberculosis was positive. Anti-tuberculous medications were started with an HREZ regimen (isoniazid, ethambutol, rifampicin, pyrazinamide).

Tuberculous aortitis is expected to increase in incidence throughout world with the rise in *M. tuberculosis* infection due to increased co-infection with human immunodeficiency virus and multiple drug-resistant tuberculosis [6]. Tuberculous infection into the aortic wall can occur by direct extension from contagious lesions such as infected lymph nodes, empyema, and pericarditis or by hematogenous or lymphangitic spread from a primary lesion [7]. Caseation necrosis involving the en-

tire thickness of the aortic wall results in perforation, either with massive hemorrhage or formation of perivascular hematoma. The latter can become encapsulated and retain communication with the lumen, known as pseudoaneurysm. Tuberculous aortitis occurs in less than 1% of patients with latent tuberculosis, but mortality rates are as high as 60%. The high mortality associated with this disease is related to the perforation of the pseudoaneurysm into the adjacent organs, causing fatal extravasation [6,8]. Tuberculous aortic aneurysm can develop despite anti-tuberculous medication, probably due to poor drug penetration into caseous necrotic tissue. Therefore, combination medical and surgical approaches are required for successful management [8,9].

Fig. 7. Positron emission tomography–CT shows fludeoxyglucose uptake and a photon defect area within the mass.

PRIMARY AORTIC INTIMAL SARCOMA INITIALLY PRESENTING AS A LOW EXTREMITY THROMBOEMBOLISM

A 74-year-old male presented to our emergency department with a chief complaint of right leg numbness, which started suddenly that morning. Initially, the symptom involved both legs with tingling sensation, but the symptom in the left leg only lasted for a few minutes. His right foot was cold and pale, and physical examination showed a pulseless right dorsalis pedis artery.

He underwent lower extremity artery CT, which showed abrupt cut off of the right distal anterior tibial and dorsalis pedis arteries without distal opacification of contrast material. Both distal posterior tibial arteries also showed abrupt cut off with distal obstruction. An about 1.1×4.6-cm-sized, eccentric, intraluminal filling defect was incidentally noted at the left lateral

Fig. 8. A 32-year-old woman presented with abnormal findings on chest X-ray. (A) More prominent bulging contour of the left atrium appendage on annual chest X-ray. (B) The same contour two years prior. This pericardial mass was diagnosed as solitary fibrous tumor.

aspect of the mid-thoracic aorta and was thought to be a thrombus (Fig. 5).

Additional MR angiography was performed; on T2WI, high signal intensity with internal intermediate SI of the mass was seen. T2WI also showed peripheral enhancement after gadolinium contrast injection, suggesting tumor rather than thrombus (Fig. 6). In dynamic enhancement image and 3D reconstruction images, the mass was seen as an ulcer-like filling defect, which favored tumor (Fig. 6D). Positron emission tomography-CT showed fludeoxyglucose (FDG) uptake and a photon defect area within the mass (SUV max 4.6) (Fig. 7).

Emergency thrombectomy of the right posterior tibial artery was performed, and pathological study showed thrombus with some atypical cells, which led to suspicion of malignancy. Resection of the aortic mass and graft interposition of the thoracic aorta were performed. Histological examination revealed infiltrating atypical cells between the intima and media of the aortic wall. Immunohistochemical studies were positive for

CD 10 and vimentin and negative for CD 34, suggesting a poorly differentiated malignant tumor, consistent with aortic intimal sarcoma.

Acute thromboembolism is commonly caused by atherosclerosis, but the possibility of tumor in the proximal artery should be considered [10]. Aortic intimal sarcoma is an aggressive tumor with extremely low incidence [10-12]. It is chiefly accompanied by embolic phenomena [11]. MR angiography of the aorta is the most sensitive imaging modality for detection of such a tumor. Furthermore, MRI can differentiate tumor from atheromatous plaque by enhancement and reveal the extent of the tumor without an invasive procedure [10,11].

PRIMARY CARDIAC SOLITARY FIBROUS TUMOR

A 32-year-old woman with no previous medical history was admitted for abnormal findings on chest X-ray. The bulging

Fig. 9. A 32-year-old woman with cardiac solitary fibrous tumor on cardiac MRI. SFT shows (A) T1 iso, (B) T2 high SI, (C) homogenous intense early, and (D) delayed enhancement (E) On cardiac CT, the left main coronary artery is displaced due to the mass effect. (F) The left atrial appendage is not visible, and it seems that some of the mass is protruding into the left atrium.

contour of the left atrium appendage gradually became prominent on the chest X-ray performed for annual screening (Fig. 8). The patient had no symptoms such as dyspnea, chest pain, or palpitation. Her blood and chemistry tests were normal.

Chest CT showed a well-defined enhancing mass in the left side pericardial space, accompanied by an internal low attenuation foci and pericardial effusion. On the echocardiogram, myocardial motion with a pericardial mass was considered to be smooth and not infiltrated. The mass showed FDG uptake SUVmax of 1.8 and mild hypermetabolism on PET-CT.

On the cardiac MRI, a T1 iso and T2 high signal intensity mass showed homogenous intense enhancement at early (10 seconds) and delayed phase (5 minutes) after contrast injection (Fig. 9A-D). In the cine image, the mass was abutting the proximal ascending aorta and left ventricle epicardium but showed a sliding sign.

On cardiac CT, the proximal portion of the left main coronary artery was displaced due to the mass effect, but there was no direct invasion. The left atrial appendage was not visible, and it seemed that some of the mass was protruding into the left atrium (Fig. 9E and F).

Coronary angiography confirmed a feeding artery from the proximal LAD branch to the mass. In this procedure, a radial artery approach was used, and the patient complained of repeated pain due to spasm. At the time of sheath removal, the patient showed bradycardia and decreased consciousness. After fluid and atropine administration, there was a brief recovery of consciousness and improved blood pressure, but sudden collapse with seizure-like movement was subsequently observed. After cardiopulmonary resuscitation, extra-corporeal membrane oxygenation (ECMO) insertion was performed.

One week later, ECMO was weaned, mass excision was performed, and cardiopulmonary bypass was conducted. The mass was not attached to the pericardium, but was attached to the left lateral wall of the pulmonary artery and the posterior wall of the aorta. The mass was not attached to the left atrium appendage, left atrium, or left ventricle. The feeding artery to the mass, seen in preoperative coronary angiography, was not found. The cardiac tumor did not have a stalk, and the origin of the tumor was not clear.

The mass was 12.0×7.3×5.5 cm and was surrounded by a smooth capsule. The lobulating mass was rubbery and uniformly pale yellow, and no bleeding or necrosis was observed. The mass was diagnosed as a solitary fibrous tumor (SFT) with STAT6 and CD34 positivity. Based on mitosis count <1/10HPF, CD34 positivity, and Ki-67 5% positivity, the mass was diagnosed as benign. The patient remains symptom-free 8 months after the operation.

DISCUSSION

Primary cardiac tumors are rare. Cardiac SFT is extremely rare, with only 20 cases having been published to date. As far as we know, this case is the first in South Korea.

SFTs originate not from the mesothelial layer but rather from the submesothelial, noncommitted, or mesenchymal layer. Mesenchyme is a pluripotent tissue with a variety of potential differentiation pathways. A mesenchymal, rather than mesothelial, origin for SFTs is supported by the finding of SFTs in numerous extrapleural sites [13].

Chest radiography, echocardiography, CT, and magnetic resonance imaging (MRI) are each useful in the diagnosis of cardiac mass [14]. Chest X-ray or echocardiography can be used to initially detect a cardiac mass. A large cardiac mass shows abnormalities such as cardiomegaly and pericardial effusion on chest X-ray. Echocardiography can show mass position, size, shape, attachment to the chamber, and mobility. CT can more precisely determine size and location, and it can show extracardiac extension and tumor relationships with other structure such as great vessels or coronary arteries. In addition, calcification or cystic change of a mass can be seen on CT. MRI can show tissue properties, myocardial infiltration, and pericardial involvement as well as mobility of the mass through the cine image [15].

One of the weaknesses of this case is that the origin of the tumor is unclear. Recently, SFTs associated with hormonal function have been reported [16,17]. For this case, because there was syncope after coronary angiography, an additional hormone study could have provided additional useful information.

In conclusion, we report an extremely rare primary cardiac tumor, solitary fibrous tumor. Our case is meaningful in that it utilized various image modalities available for preoperative evaluation of cardiac tumors. In this case review, we were able to compare the advantages of these image modalities.

Conflicts of Interest

The authors declare that they have no conflict of interest.

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