Clinical Case Report

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Superior vena cava syndrome secondary to recurrent coronary artery fistula

A case report and literature review

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Abstract

Rationale: Coronary artery fistula (CAF) is characterized by an abnormal communication of a coronary artery with a cardiac chamber or a great vessel bypassing the capillary bed. Surgical closure of large or symptomatic CAF is the gold standard treatment. However the previously closed CAF still has the possibility to reopen. Superior vena cava syndrome (SVCS) is defined as a condition that occurs when the obstruction of the superior vena cava interrupts blood flow from the head, upper extremities, and thorax to the right atrium and can present a life-threatening situation. In this report, we described a case of SVCS, which was secondary to the compression by a huge aneurysm formed in a recurrent CAF, as a long-term complication associated with surgical treatment of CAF.

Patient concerns: A 47-year-old woman presented with chief complaint of progressive exertional dyspnea for one month.

Diagnoses: Superior vena cava syndrome and recurrent coronary artery fistula.

Interventions: Reoperation for ligation of the fistulous and excision of the aneurysm were performed.

Outcomes: The patient made an uneventful recovery and her postoperative course through 1-year follow-up was uneventful.

Lessons: First, SVCS is a rare but clinically important postoperative complication of surgical ligation of CAF. Second, surgical ligation of the fistula alone is unlikely to prevent the ectatic course. Therefore, long-term follow-up is mandatory for patients with CAF undergoing surgical closure.

Abbreviations: CAF = coronary artery fistula, LM = left main coronary artery, MSCT = multislice computed tomography , SVCS = superior vena cava syndrome, TTE = transthoracic echocardiogram.

Keywords: coronary aneurysm, coronary artery fistula, recurrent fistula, superior vena cava syndrome

1. Introduction

Coronary artery fistula (CAF) is a relatively rare entity, which is characterized by an abnormal communication of a coronary artery with a cardiac chamber or a great vessel bypassing the capillary bed. Closure of all large CAFs or symptomatic CAFs is recommended by the guideline to prevent the development of serious complications.[1] Traditionally, surgical closure of coronary fistulae, by external placation on the beating heart or by intracardiac closure using cardiopulmonary bypass, is the gold standard treatment. However, patients undergoing surgical treatment may be exposed to the risks of bleeding, post-pericardiotomy syndrome, myocardial infarction, recurrence, and stroke. So far, superior vena cava syndrome (SVCS) as a complication associated with surgical treatment has not been reported. Herein, we described a case of SVCS, which was secondary to extrinsic compression by a huge aneurysm formed in a recanalized CAF, as a long-term complication associated with surgical treatment of CAF.

2. Case report

The study was approved by the Ethical Review Committee of The Second Xiangya Hospital of Central South University and the informed consent was obtained from the patient. A 47-year-old woman was admitted with chief complaint of progressive exertional dyspnea for 1 month. The patient had a known history of a left main coronary artery to the right atrium fistula, which had been repaired by directly surgical ligation at the drainage site one year ago. Repeated transthoracic echocardiogram (TTE) performed at 3 days after procedure demonstrated no residual shunt. However, the patient re-experienced exertional dyspnea 1 year later. Physical examination was notable for signs of superior vena cava obstruction—marked jugular venous distention and facial edema. Chest radiography and resting electrocardiography revealed no abnormalities. The TTE showed a dilated left main coronary artery (LM) and a CAF originating from the LM with a retroaortic course and draining into the right atrium (Fig. 1). Selective coronary angiography showed no significant coronary artery stenosis but did not clearly demonstrate the CAF. To delineate the cardiac anatomy in more detail, ascending aortogram and cardiac multislice computed

Editor: N/A.

The authors have no conflicts of interest to disclose.

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Medicine (2017) 96:51(e9111)

Received: 24 October 2017 / Received in final form: 14 November 2017 / Accepted: 15 November 2017

http://dx.doi.org/10.1097/MD.0000000000009111

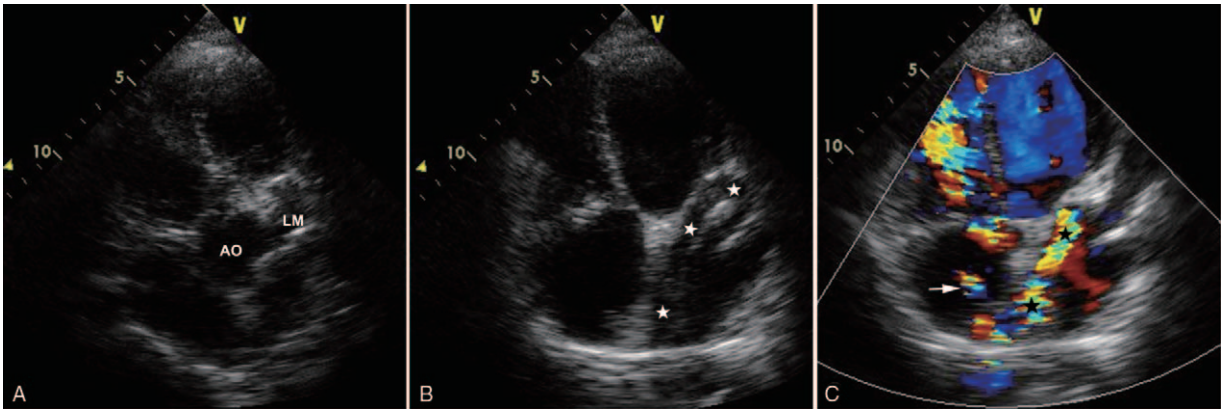

Figure 1. Transthoracic echocardiogram revealed the ostium of the left main (LM) coronary artery was significantly dilated (A) and the fistula originated from the LM, coursing around the aorta (B, stars) and draining into the right atrium (C, arrow). LM = left main.

tomography (MSCT) were performed (Fig. 2). These detailed images provided confirmation and also demonstrated a huge, measuring $4.5 \times 3.9 \times 3.6$ cm aneurysm located in the distal portion of the CAF. Furthermore, the superior vena cava was significantly collapsed due to extrinsic compression by the aneurysm, which revealed the cause for SVCS. Reoperation for ligation of the fistulous and excision of the aneurysm were performed. Intra-operatively, the recurrence and aneurysmal dilation of the previously ligated CAF was verified (Fig. 3A). Histologic examination of the removed aneurysm revealed no signs of atherosclerosis and inflammatory reaction (Fig. 3B). The patient made an uneventful recovery and was discharged home 2 weeks later. The postoperative MSCT showed the fistula was successfully closed but the LM remained grossly dilated. Thus, she received antiplatelet therapy with aspirin for 1 year and warfarin for 6 months to reduce the risk of thrombosis or late

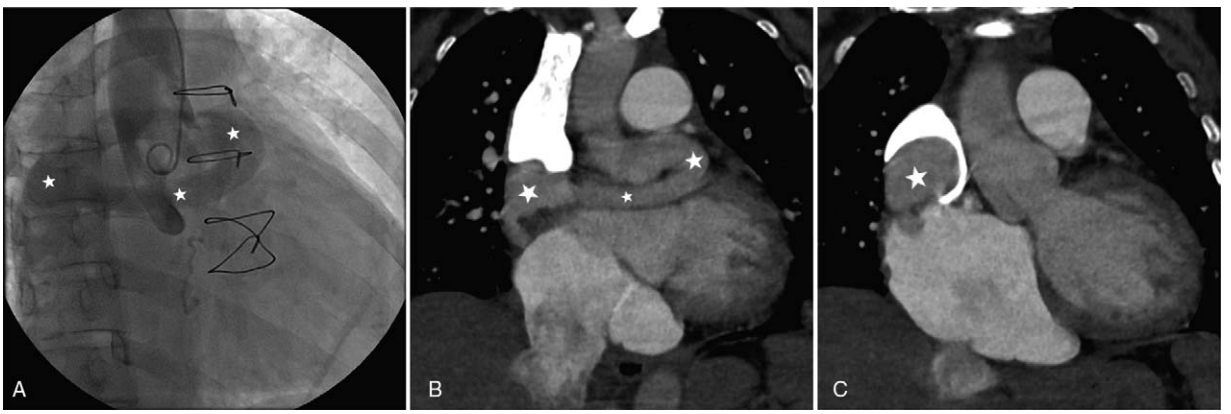

Figure 2. Ascending aortogram confirmed the presence of recurrent coronary fistula (A, stars) and Cardiac multislice computed tomography highlighted the grossly aneurysmal dilation of the fistula compressing the superior vena cava (B and C, stars).

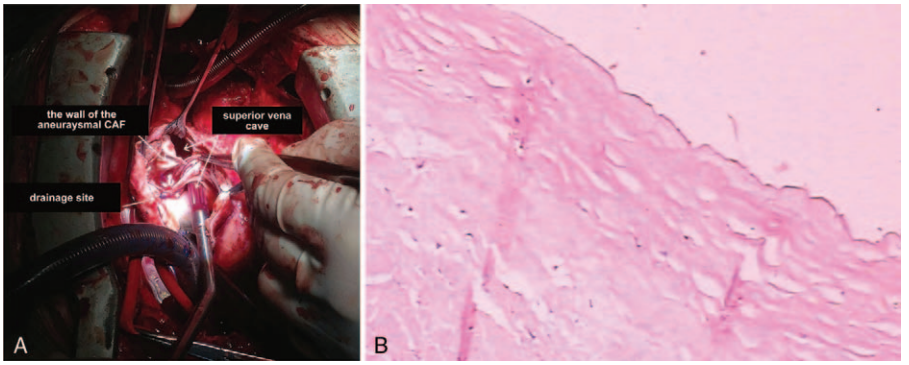

Figure 3. (A) Intraoperative view. (B) Histology of the removed aneurysm (hematoxylin-eosin, original magnification ×100).

myocardial ischemia due to retrograde thrombus propagation secondary to the persistent LM dilation. Her postoperative course through 1-year follow-up was uneventful.

3. Discussion

The recurrence of CAF is an uncommon but well-described complication of surgical treatment. The echocardiogram is the most commonly used modality to monitor the status of the fistula after surgery. The gold standard for identifying the recurrence remains coronary angiography. However, in our case, due to the large lumen of the LM and grossly aneurysmal dilation of the recurrent CAF, the contrast agent injected through a small catheter was hard to fill up the entire fistula tract. In this situation, angiography using a pigtail catheter is helpful for delineate the anatomical features of this disorder precisely. More importantly, the MSCT can provide a superior visualization of the adjacent anatomical structures. Actually, the images obtained using MSCT in our patient were more informative than those acquired by conventional coronary angiography.

Due to the limited cases number and different postoperative workup in previous studies, the reported incidence of CAF recurrence after surgical closure varied from 0% to 19%.[2,3] Fortunately, most patients with recurrent coronary fistula were asymptomatic and without hemodynamic disturbance, thus additional interventions in these cases were not compulsory. However, not all recurrent CAFs have a benign nature. Kostis et al[4] described a successfully ligated fistula reopened and led to re-experience the preoperative symptoms. In the present case, we described the grossly aneurysmal dilation of a recurrent CAF as an unusual cause of superior vena cava obstruction. Previously, very few reports of SVCS associated with coronary vessels were presented in literature. We found 2 case reports that described the association of such syndrome with aneurysms of saphenous vein grafts[5] and with giant coronary artery aneurysm.[6] To the best of our knowledge, our case was the first time to report the combination between recurrent CAFs and SVCS. As the distal portion of the recurrent CAF was located in immediate proximity to the superior vena cava, the aneurysmal dilation of recurrent CAF in the distal segment caused the compression of the superior vena cava which led to SVCS. In addition, the aneurysmal formation of previously ligated CAF suggested that the ectatic course of CAF seems unlikely to be influenced by a simple ligation. Araya et al[7] reported a patient who died 6 months after operation for a CAF because of the recurrent aneurysmal fistula that ruptured into the mediastinum, also suggesting that surgical ligation of the fistula alone also does not prevent subsequent rupture.

The mechanism of the recurrence is still unclear. Kostis et al[4] pointed out that the pulsatile aortic pressure transmitted to the probable small residual lumen between the infolded walls of the vessel at the level of the ligature resulted in reestablishment of the fistula, recommending that division and oversewing should be done whenever feasible to prevent CAF from recurrence. However, one patient in Said's report,[8] who underwent excision of the fistula, developed recurrent CAF suggesting that even if the CAF was divided, it still has the possibility to reconnect.

In conclusion, CAF recanalization with giant aneurysm formation after surgical closure causing SVCS, as in the present case, are extremely rare. The present case highlights 2 clinical issues of CAF treatment: firstly, superior vena cava obstruction is a rare but clinically important postoperative complication of surgical ligation of CAF. Secondly, surgical ligation of the fistula alone is unlikely to prevent the ectatic course. Therefore, long-term follow-up is mandatory for patients with CAF undergoing surgical closure.

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