CASE REPORTS

LIMB ISCHAEMIA AND MULTISYSTEM DYSFUNCTION AS A CONSEQUENCE OF
CHOLESTEROL EMBOLIZATION SYNDROME

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ABSTRACT

Following a Coronary artery bypass graft and insertion of an intra-aortic balloon pump, the 63 year-old male patient developed signs of progressive multi-organ dysfunction and ischaemic changes in both of his lower limbs. The diagnosis of cholesterol embolization syndrome was made. This condition has been noted to be a complication of invasive procedures and endovascular therapies, and is often overlooked and difficult to treat. While the patient’s renal function and pancreatitis showed improvement, the lower limb ischaemia was progressing. Eventually, the patient underwent a bilateral below knee amputation.

Keywords: Cholesterol emboli, Cholesterol crystal embolization, Digital ischaemia, Intra-aortic balloon pump, Plaque rupture.

INTRODUCTION

Cholesterol embolization syndrome (CES) is a rare and often overlooked consequence of advanced atherosclerotic disease. The underlying pathology is that plaque disruption occurs, leading to the contents of the plaque embolizing throughout the vascular system1. Plaque disruption can occur spontaneously but is said to be iatrogenic in >70% of cases2. The treatment of CES is largely symptomatic3. Interventions include surgical resection and endovascular approach3. Here we present a case with typical features of CES.

CASE SUMMARY

A 63-year-old male patient, with diabetes mellitus and smoking history, underwent coronary artery bypass grafting. In an attempt to improve his faltering ejection fraction, an intra-aortic balloon pump (IABP) insertion was done with great difficulty. On the first post-operative day, the patient began to develop dusky and cold extremities in the lower limbs bilaterally. He had bilateral ankle edema and his distal pulses were not palpable despite audible arterial signals on hand-held doppler. However, a detailed duplex scan of the lower limbs indicated there was nearly normal blood flow. His total leukocyte count was high (14.8 x 10³), as was creatinine phosphokinase level (1531 U/L). On the Second post-operative day, his platelet count decreased to 57,000 and signs of renal impairment surfaced.

In the days to follow, the limb ischemia and renal impairment worsened. There was a persistent leukocytosis with thrombocytopenia of 22,000. On the 10th post-op day, the patient developed abdominal pain and distension. Investigations revealed amylase level of 1600 U/L; suggestive of acute pancreatitis. The C-reactive protein level was persistently elevated. In the light of the patient’s history, the prior insertion of an IABP4, characteristic findings, and the diagnosis of exclusion was found to be CES.

With the diagnosis of CES and thrombocytopenia, anticoagulation was not started. While the patient’s renal function began to improve and abdominal symptoms regressed, he failed to show improvement in the lower limbs with regards to the ischemia and eventually underwent a bilateral below knee amputation, however the limbs were not sent for histopathology as it would not have changed the outcome and further treatment.

DISCUSSION

CES was first described in an autopsy in 1862 by Fenger and Fanum5. CES consists of advanced atherosclerotic plaques in large caliber vessels rupturing, rel-
easing cholesterol crystals into the circulation, resulting in gradual ischemic changes and multiorgan dysfunction by building up in smaller vasculature. Mortality ranges between 5-16%. Male gender, age over 60 years, heavy smoking, with diabetes mellitus, hyperlipidemia, abdominal aortic aneurysms, cardiovascular surgery, endovascular therapy and anticoagulation have all been stated as risk factors. More than 70% of the cases are iatrogenic. CES as a complication of an intra-aortic balloon pump occurs in 0.1% as reported by a review article.

The clinical manifestations of CES most commonly occur in the skin in the form of livedo reticularis, skin discoloration, gangrenous digits. Pancreatitis has also been reported as a rare complication of CES which was seen in our patient. The definitive diagnosis for CES is made by biopsy. C-reactive protein is raised and 80% of cases show elevated eosinophil counts. Treatment is symptomatic. Statins and corticosteroids have been have resulted in clinical improvement in some cases. The use of anticoagulants in patients with CES is a controversial topic. Some suggest anticoagulants should be avoided, as they have been assumed to promote plaque disruption. A study showed 7% of confirmed CES cases with no previous endovascular or operative history could be attributed to anticoagulant therapy.

This condition can be prevented through risk factor modification in the form of control of tight control of co-morbid conditions and limitation of intravascular procedures and techniques. Preventing CES with specific emphasis to the IABP, can be done by using smaller sized catheters with a sheathless technique.

CONCLUSION

Cholesterol embolization syndrome (CES) is a complication of invasive procedures involving large caliber vessels. For this reason, if a patient begins to develop ischemic extremities and show signs of progressive renal shutdown after an invasive procedure, it is very important to consider CES. Although the complication rate for CES following intra-aortic balloon pump is only 0.1%, these are only the cases that were confirmed to be CES, the actual figure may be much higher and need to be recognized. This topic is becoming increasingly significant with the advances and increased use of endovascular treatment and interventional radiology.

CONFLICT OF INTEREST

This study has no conflict of interest to be declared by any author.

REFERENCES