Diagnosis of Porphyria after sternotomy for severe calcific coronary artery disease, a Case Report

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ABSTRACT

Acute intermittent porphyria (AIP) is an autosomal disorder marked by a deficiency of the enzyme, the hydroxymethylbilane synthase which is part of the heme biosynthesis. It is manifested clinically by multi-system involvement. Our patient does have chronic ischemic heart disease needed surgical revascularization; his sternotomy incision revealed the classical blackish discoloration of the bone marrow, which guided us for his work up and diagnosis.

Key words: acute intermittent porphyria (AIP), coronary artery bypass grafting (CABG), left internal mammary artery (LIMA).

Introduction

Porphyria, a hematological disease, which involves the heme metabolism, can present with multiple features. It has many clinical presentations which can mimic multiple diseases.

Here we present this case which was diagnosed with acute intermittent porphyria (AIP) during sternotomy for CABG.

We report a 40-year-old gentleman who, apart from smoking history, had no other risk factors for coronary artery disease. One more pertinent issue is that he had a chronic history of vague left loin pain which is intermittent and was treated as urinary gravels. This gentleman had recurrent attacks of angina chest pain, for which he was studied in the cardiology clinic and his work up included coronary angiogram which revealed three vessel coronary artery disease not amenable for percutaneous coronary intervention. After reviewing his coronary angiogram, there were multiple calcific lesions with variable distribution along his coronary territories (Figure 1). Decision was taken to operate on him and perform coronary artery bypass grafting. He was brought to the operating room for elective triple coronary bypasses for his diseased coronaries. Given his very young age, this raised the suspicion of a systemic disease.
After being prepped and draped in the usual sterile fashion, full primary median sternotomy was performed. Interestingly there was a dark black colored bone marrow spot at the distal lower part of the sternum (Figure 2), for which, an incisional biopsy was sent to the histopathology laboratory. During LIMA harvesting, multiple dark black spots covering multiple ribs were also noted.

Figure 1: Coronary Angiogram revealing multiple diffusely distributed calcific spots along the coronary territories.

Figure 2: Intra-operative view, demonstrating a blackish discolored spot in the lower aspect of the sternotomy incision.
When pericardium was opened and heart suspended in pericardial cradle, cardiopulmonary bypass was commenced at 2.4 L/M2 and patient temperature drifted to 34 c. and cardioplegic arrest done with ante grade and retrograde fashion. Coronary arteries were examined and showed diffuse calcification with multiple dark spots.

Surgery was uneventful and patient recovered fully and was discharged 10 days after multiple diagnostic tests were sent and confirmed his disease.

Discussion

Many groups of disorders that are due to accumulation of Porphyins can produce the disease of porphyria (1,2). It is inherited as autosomal pattern - which is most common - as well as autosomal recessive - rarely occurring Porphyria's affect many organs including CNS, skin, kidneys, liver and bone as well.

Vague presentations and lots of nonspecific signs and symptoms make the diagnosis difficult in solitary cases which have no family history of such a disease, as in this case.

Triggering factors that might precipitate the acute attacks of porphyria include alcohol, smoking, medications, fasting, stressful events, infections and others.

Other forms of porphyria can produce cutaneous manifestations which is not in the scope of this case.

Conclusion

Subtle changes of organ tissues can be the stepping stone for the workup of rare diseases. High index of suspicion and systemic examinations of all tissues apart from the planned interventions may make outcomes better. Our case can serve as a reminder to keep these rare diagnoses in mind when such a scenario may be faced.

References