Cardiovascular Revascularization Medicine 16 (2015) 116–123

Contents lists available at ScienceDirect

Cardiovascular Revascularization Medicine

Case Review

Large coronary intramural hematomas: a case series and focused literature review

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ARTICLE INFO

Article history:
Received 31 July 2014
Received in revised form 22 October 2014
Accepted 23 October 2014

Keywords:
Vessel wall pathology
Intravascular ultrasound
Diagnostic
Treatment strategy

ABSTRACT

Isolated spontaneous coronary intramural hematoma is a unique subset of spontaneous coronary artery dissection that is characterized by a hemorrhage limited to the medial–adventitial layers, causing subsequent hematoma formation without visible intimal flaps. It is an infrequent and serious coronary vessel wall pathology, with poorly understood underlying pathogenic mechanisms. Affected individuals may present with a broad spectrum of symptoms ranging from acute coronary syndromes (ACS) to cardiogenic shock or even sudden cardiac death. The disease entity causes challenges in terms of both diagnostics and treatment strategy. Coronary intramural hematomas can also occur iatrogenically, as a complication to percutaneous coronary intervention (PCI). Coronary angiography (CAG) has limited diagnostic value in the absence of intimal dissections, and lesions are often angiographically ambiguous. Intravascular ultrasound (IVUS) is an important diagnostic tool in establishing the correct diagnosis, as it provides a complete vessel wall assessment, and enables morphometric information regarding the magnitude and severity of the underlying hematoma. Due to the rarity of this clinical scenario, no randomized, controlled trials exist to guide treatment, and no consensus regarding management is available. Currently, treatment strategies are based on a case-by-case clinical assessment, and experiences described in previous, limited retrospective studies and case reports.

1. Introduction

Spontaneous coronary intramural hematoma is a unique and rare subset of spontaneous coronary artery dissection (SCAD), where a hemorrhage within the vessel wall is thought to be the underlying cause, leading to separation of the mural layers. The hematoma formation is limited to the medial–adventitial layers, and no flaps are visible when assessed with tomographic techniques [1,2]. Previously, rupture of the vasa vasorum without communication with the lumen has been proposed as the underlying pathophysiological mechanism [3–6]. This serious condition usually presents as acute coronary syndrome (ACS), but symptoms may range from chest pain to cardiogenic shock or sudden death [1,2], depending on hematoma location, number of affected vessels, lumen compromise, and restriction of coronary blood flow. It is more frequent in young women, particularly in the peri- and postpartum period [7] and with oral contraceptive use, and in young and middle-aged patients without obvious cardiovascular risk factors [8,9].

Coronary intramural hematoma can also occur iatrogenically, as a complication to percutaneous coronary intervention (PCI) [10–12]. In this patho-physiological scenario, the hematoma begins as a trauma triggered dissection to the media at an atherosclerotic reference segment site, and propagates along the medial plane into a more normal artery segment, but does not re-enter the lumen [11,13,14]. The incidence is increasing due to the growing use of interventional diagnostics and treatment of coronary artery disease.

Intravascular ultrasound (IVUS) is an important diagnostic tool, which may provide important morphometric information regarding the magnitude and severity of intramural hematomas. Also, when the decision to treat has been made, IVUS provides precious information regarding vessel size and hematoma length that helps guide PCI treatment and stent selection. Management strategies may range from conservative medical treatment to percutaneous or surgical interventions depending upon the anatomy and extent of the hematoma, compromise of the vessel lumen and the clinical circumstances [10,15], and the prognosis varies widely as the numbers of reported cases are limited [6].

We present three cases of coronary intramural hematomas that originated from different causes, and the important role of IVUS in the diagnostic process and subsequent treatment is highlighted. Based on previous, limited case reports and retrospective analyses, a focused literature review assessing the existing management options and prognosis, is provided in the discussion section.

1.1. Case 1

A 76-year old female with a history of hypertension, smoking and a family history of ischemic heart disease was admitted due to gradual increasing dyspnea over one week. Symptom onset was preceded by a few days with gastroenteritis causing pronounced diarrhea and vomiting.

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http://dx.doi.org/10.1016/j.carrev.2014.10.009
1553-8389/

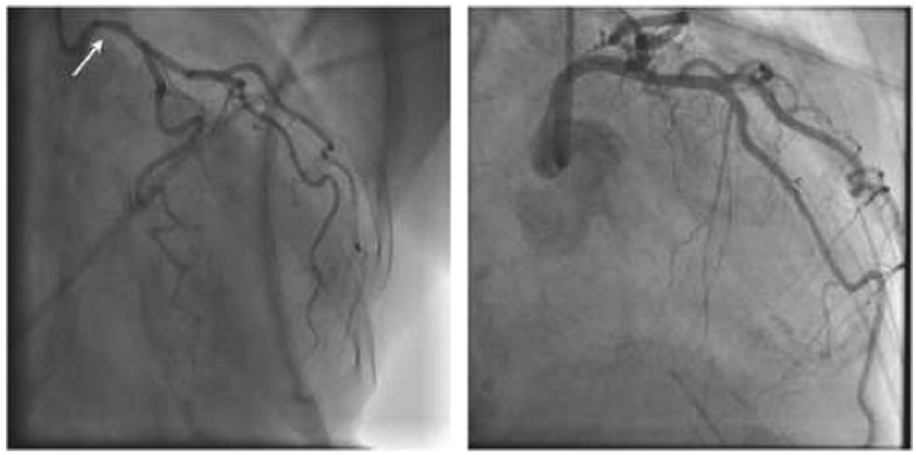

Fig. 1. Coronary angiography revealing a small caliber left main stem (*white arrow*) (*left*), and 1 month later with normal caliber vessels (*right*).

The electrocardiogram (ECG) showed a left bundle branch block (LBBB), and Troponin-I was moderately elevated (5.2 ng/ml). An echocardiogram showed global hypokinesia of the left ventricle with an ejection fraction (EF) of 15%–20%. She was diagnosed as being a "late presenter" with ST segment elevation myocardial infarction (STEMI), and treatment with aspirin, ticagrelor and fondaparinux was started. Also treatment for heart failure due to a poor left ventricular function was initiated.

A subacute coronary angiography (CAG) revealed angiographically normal coronary arteries, apart from a small caliber left main stem (LM), with a diameter less than that of the proximal left anterior descending (LAD) and the left circumflex (CX) (Fig. 1, *left*). There was Thrombolysis In Myocardial Infarction (TIMI) flow grade 3 in all coronary segments. Subsequently, we performed an IVUS of the LM and LAD, which identified the cause of the luminal narrowing to be a large intramural hematoma starting at the proximal LM and progressing to the mid LAD with a length of approximately 40 mm (Fig. 2, *row 1 + 2*). Minimum lumen area (MLA) was 4.8 mm^2 at the LM and 4.1 mm^2 at the LAD. There was no evidence of dissection, neither was there any evidence of atherosclerosis. The patient

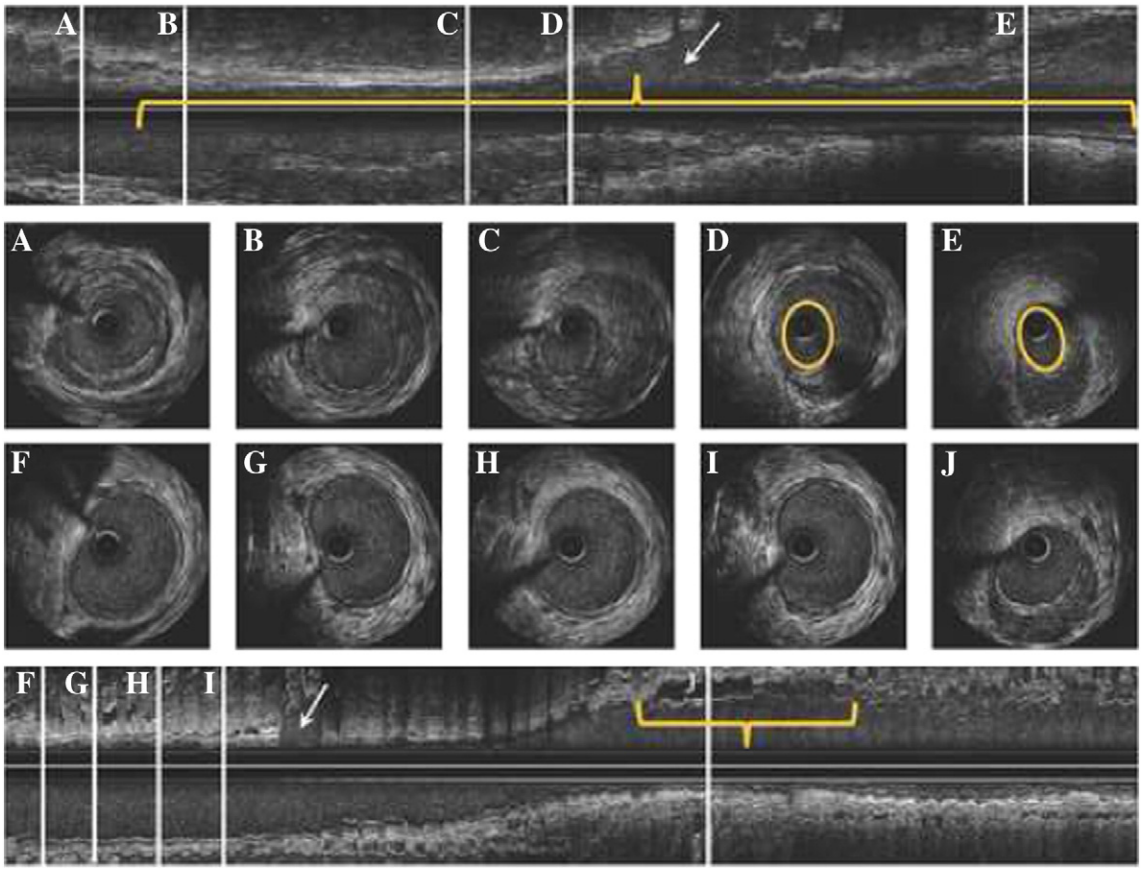

Fig. 2. First and second row: Diagnostic IVUS revealing a large intramural hematoma, which is primarily localized at the LM, and extends into the proximal LAD (length approximately 40 mm (*yellow bracket*)) with cross-sectional IVUS areas (CSAs) of the hematoma extending from the LM (*left*) to the proximal LAD (*right*). Minimal lumen area (MLA) at the LM (4.8 mm^2) is indicated by a *yellow luminal contour* in CSA (D), while MLA at the LAD (4.1 mm^2) is indicated by a *yellow luminal contour* in CSA (E). Third and fourth row: Repeat IVUS images of the spontaneously near- resolved hematoma at 1-month follow-up from the LM (*left*) to the proximal LAD (*right*). CSA (I) corresponds to CSA (D) in the second row, and CSA (J) corresponds to CSA (E) in the second row. A *yellow bracket* in the longitudinal image indicates the residual hematoma at the proximal LAD. *White arrows* (longitudinal images) indicate the ostium of the left circumflex.