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Case report

Sudden death due to coronary artery dissection associated with fibromuscular dysplasia revealed by postmortem selective computed tomography coronary angiography: A case report

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ABSTRACT

We present an autopsy case of sudden death due to coronary artery dissection associated with fibromuscular dysplasia (FMD) in a young female patient. Postmortem selective coronary artery computed tomography (CT) angiography revealed dissections of the left anterior descending and left circumflex arteries. These findings were confirmed by subsequent autopsy. Histopathological examination revealed coronary artery FMD, which is considered a risk factor for dissection.

To the best of our knowledge, this is the first postmortem radiology–pathology correlation of coronary artery dissection associated with FMD.

1. Introduction

Coronary artery dissection has been reported as a rare cause of acute coronary syndrome and sudden cardiac death. The predisposing conditions for coronary artery dissection have been subdivided into atherosclerotic and nonatherosclerotic categories. Nonatherosclerotic entities have included peripartum state, connective tissue disorders, systemic inflammatory conditions, coronary artery spasm, shear-stress-related factors, and fibromuscular dysplasia (FMD) [1].

Coronary artery dissection associated with FMD has been considered very uncommon [2–4], though recently published data based on radiological analysis have suggested the incidence of coronary artery dissection associated with FMD is more frequent than previously thought [1,5,6].

We report an autopsy case of sudden death due to coronary artery dissection associated with FMD. In this case, postmortem selective computed tomography (CT) coronary angiography was performed during autopsy. As far as we know, this is the first report of postmortem radiological evaluation of this entity.

2. Case report

2.1. Clinical summary

A 28-year-old female individual worked at a bookstore as usual without emotional stress or physical exertion, but reported abdominal and chest pain beginning in the morning. At 16:33, she suddenly ran into the office and fell on the floor. At 16:47, when the rescue crew arrived, she was still conscious, but at 17:04,

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on arrival at the hospital, cardiac arrest occurred. Without any response to resuscitation, she was pronounced dead at 20:20.

The patient had no remarkable past history, but had a noteworthy family history. Her mother had died suddenly at age 30 years. Her grandmother had also died young. Both causes of death were not clear, and without detailed postmortem examinations. The patient's father strongly desired to know the cause of death out of concern for his surviving daughter.

2.2. Radiological examinations

Prior to autopsy (3.5 days after her death, with the body held at 4 °C in the interim), a postmortem whole-body CT scan was performed and interpreted by a forensic pathologist with four years of experience with postmortem imaging. The scan was performed using a 16-row multidetector CT scanner (Eclos®, Hitachi Medical Corporation, Tokyo, Japan) with the following protocol and parameters: collimation 1.25 mm, reconstruction interval 1.25 mm, tube voltage 120 kV, tube current 200 mA, and scan speed of one rotation per second. Contrast medium was not administered. Both soft-tissue and bone kernels were used for reconstruction. Image interpretation was done by both the forensic pathologist and a board-certified radiologist. This scan showed no findings suggesting her cause of death. The scan found no calcifications in any of the visible arterial walls, including the coronary arteries, and no detectable aortic ectasia. There was no abnormality in her bony structure such as scoliosis or pectus excavatum. There was no evidence of dural ectasia.

During autopsy, postmortem selective CT coronary angiography was performed by the forensic pathologist, following the protocol described in our previous report [7]. After extraction of the heart, the tissue surrounding the coronary arteries was carefully removed and two 6-Fr coronary angiography catheters were inserted into the right and left coronary ostia, respectively, and secured with a ligature at the side of the aorta. These two catheters were connected to an infusion bag pressurized to 100–150 mm Hg filled with high-viscosity contrast medium composed of 15 mL iohexol (Omnipaque 300®, Nycomed Inc., Princeton NJ, USA) in 500 mL polyethylene glycol (PEG). Intra-bag pressure could be regulated manually, and if the bag clamp was open, contrast media flew passively into the cannulated vessels. The heart was positioned in the CT gantry and a non-contrast CT scan was performed with the bag clamped, followed by a contrast-enhanced scan approximately 3 min after allowing the contents of the bag to flow into the arteries. Both scans were performed using 0.63 mm collimation with 120 kVp and 200 mA s. Image reconstruction was performed with slice thickness of 0.625 mm.

Postmortem selective CT coronary angiography data were viewed and reconstructed at a workstation (Vincent, Fujifilm, Tokyo, Japan). As shown in Fig. 1A–G, curved multiplanar reconstructed images of the opacified left anterior descending coronary artery (LAD) show two opacified lumens separated by an irregular cord-like filling defect extending from the proximal to middle portion, characteristic of coronary artery dissection with a visible intimal flap. The images also show partial occlusion of the first major septal branch by the flap. The results of left circumflex artery (LCX) angiography are shown in Fig. 1H and I. Dissection is also suggested in image I in the proximal portion of the LCX. The right coronary artery (RCA) (Fig. 1J and K) appears normal. There were no gross signs of FMD, such as aneurysmal changes or string-of-beads appearance, in the angiographic images.

2.3. External examination and conventional autopsy

An autopsy was performed just after postmortem CT, according to the national forensic autopsy standards, which include opening all three cavities of the body (head, chest and abdomen) [8]. She was a normally-nourished Asian woman weighing 50.5 kg and measuring 158 cm in height. The skin did not show any abnormalities. The heart was normally formed, weighing 225 g. Grossly, other organs including the brain, lungs, thyroid, liver, pancreas, spleen, both adrenal glands, both kidneys, gastrointestinal tract, uterus, and ovaries had no abnormality. At the same time as the autopsy, postmortem selective CT coronary angiography was performed as described above.

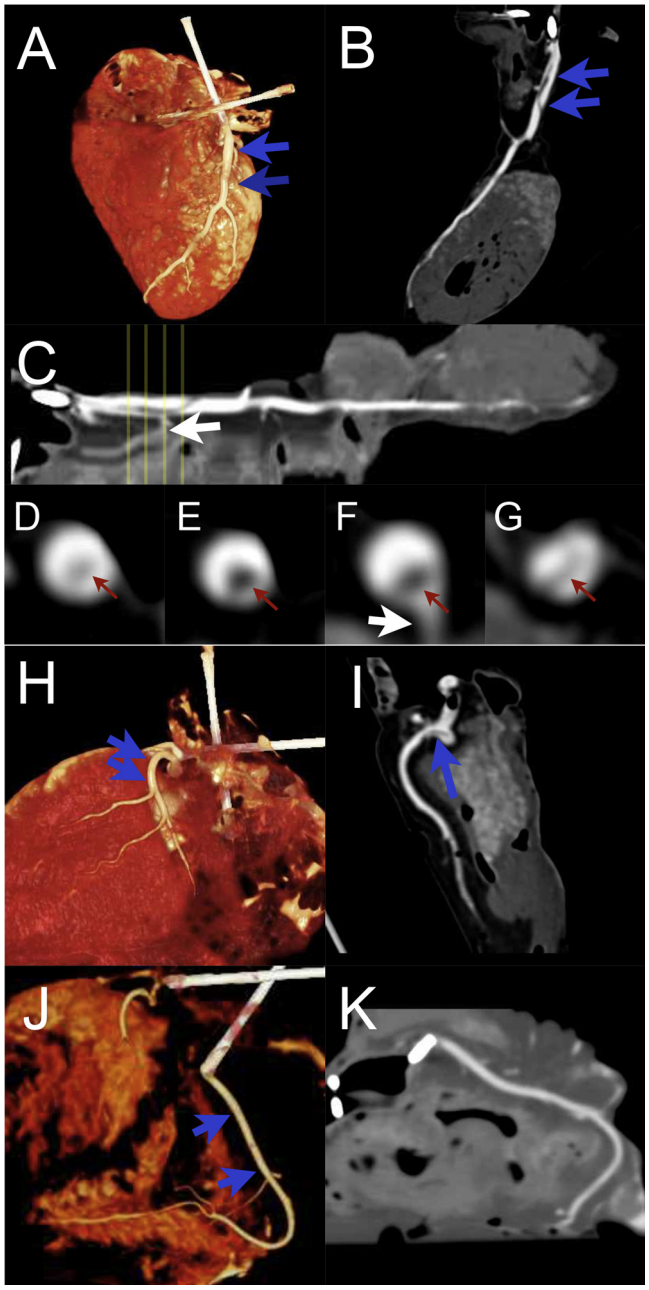

Fig. 1. Results of postmortem selective CT coronary angiography. Three-dimensional images showing the LAD (A), LCX (H), and RCA (J) without definite conformational changes (arrows). Angiographic curved planar reconstructed (CPR) image of LAD (B), stretched CPR image (C), and its axially sectional image (D–G) show an elongated filling defect consistent with an intimal flap (arrows). Yellow lines on image (C) anterior to posterior correspond to (D)–(G), respectively. White arrows on image (C) and (F) indicate the first major septal branch. Angiographic CPR image of the LCX also shows a similar structure in its proximal portion (I). Angiographic CPR image of the RCA shows no abnormality (K). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)