Computed Tomographic Findings in Unsuspected Aortic Dissection and Adult Polycystic Kidney Disease

H. Theodore Harcke, Wendy S. Warren, Christopher J. Gordon, Edward L. Mazuchowski

ABSTRACT

The coexistence of adult polycystic kidney disease and aortic dissection should be recognized by forensic pathologists and radiologists. Two cases with postmortem computed tomography prior to autopsy illustrate the appearance of polycystic kidneys and the hemorrhage patterns that provide clues to the presence and approximate location of the aortic dissections. Optimal imaging technique is discussed. *Acad Forensic Pathol.* 2016 6(4): 673-678

AUTHORS

H. Theodore Harcke MD, Office of the Armed Forces Medical Examiner

Roles: Project conception and/or design, data acquisition, analysis and/or interpretation, manuscript creation and/or revision, approved final version for publication, accountable for all aspects of the work, principal investigator of the current study.

Wendy S. Warren DO, Office of the Armed Forces Medical Examiner

Roles: Data acquisition, analysis and/or interpretation, manuscript creation and/or revision, approved final version for publication, accountable for all aspects of the work.

Christopher J. Gordon MD, Office of the Armed Forces Medical Examiner

Roles: Data acquisition, analysis and/or interpretation, manuscript creation and/or revision, approved final version for publication, accountable for all aspects of the work, principal investigator of the current study.

Edward L. Mazuchowski MD PhD, Office of the Armed Forces Medical Examiner

Roles: Data acquisition, analysis and/or interpretation, manuscript creation and/or revision, approved final version for publication, accountable for all aspects of the work, general supervision.

CORRESPONDENCE

H. Theodore Harcke MD, 1600 Rockland Road, Wilmington DE 19803, howard.harcke@gmail.com

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FINANCIAL DISCLOSURE

The authors have indicated that they do not have financial relationships to disclose that are relevant to this manuscript

KEYWORDS

Forensic pathology, Computed tomography, Aortic dissection, Polycystic kidney disease

INFORMATION

ACADEMIC FORENSIC PATHOLOGY: THE OFFICIAL PUBLICATION OF THE NATIONAL ASSOCIATION OF MEDICAL EXAMINERS

©2017 Academic Forensic Pathology International • (ISSN: 1925-3621) • <https://doi.org/10.23907/2016.063>

Submitted for consideration on 2 Aug 2016. Accepted for publication on 23 Aug 2016

INTRODUCTION

Postmortem computed tomography (CT) has become an adjunct to the forensic autopsy by providing a guide to the forensic pathologist performing the death investigation. Use of CT in deaths by natural causes is more difficult than those associated with ballistic injury and trauma because pathology involves differentiation of soft tissue, which is limited without use of contrast. Recognizable findings, such as organ morphology and hemorrhage pattern, however, can be used to prompt the search for particular pathology. Two cases of unsuspected adult polycystic kidney disease (APKD) illustrate this point.

METHODS

Prior to forensic autopsy, whole body digital radiographs and head-to-toe CT were obtained with the body in anatomic position. Radiography was performed using IMIX DDR (IMIX Americas Inc., Sterling, VA) and CT imaging was done on a General Electric Lightspeed Xtra (General Electric [GE] Medical Systems, Milwaukee, WI) scanner with extended table travel. The CT protocol obtains contiguous axial sections at 0.625 mm to produce isotropic voxels. Multiplanar reconstructions were viewed on a GE Advantage workstation (General Electric Medical Systems). The imaging studies were viewed by a forensic radiologist who reported findings to the medical examiner at the time of forensic autopsy. Both autopsies were performed by board-certified forensic pathologists.

CASE DESCRIPTIONS

Case 1: A 39-year-old male who was admitted to the hospital for treatment of deep vein thrombosis and pulmonary emboli became acutely unresponsive and died despite medical intervention. Postmortem CT showed irregular contours and cystic changes in enlarged kidneys and pulmonary congestion (**Image 1A**). The liver was homogeneous with no cysts seen. Diffuse retroperitoneal hemorrhage was noted, with margins of the abdominal aorta obscured (**Images 1B and C**). Autopsy revealed the presence of

lightly adherent thromboemboli in the distal right and left pulmonary arteries and major branches (**Image 2A**). The right (**Image 2B**) and left kidneys each weighed 220 g and contained numerous thin-walled cortical and medullary cysts measuring up to 3 cm. Diffuse retroperitoneal hemorrhage surrounded the kidneys, adrenal glands, ureters, abdominal aorta, inferior vena cava, and rectum. A 1 cm full-thickness defect was present on the posterior abdominal aorta approximately 11 cm proximal to the renal arteries – a Stanford B/DeBakey III dissection (**Image 2C**). Histologic sections from the defect showed transmural separation and dissecting hemorrhage through layers of the tunica media. There was no significant histopathologic abnormality associated with the defect. Elastin-stained slides revealed no significant disruption of the elastin layers. The liver was noted to grossly show slightly dilated vascular structures but no definitive cysts. Microscopic examination of the liver revealed mild steatosis and no fibrosis, cysts, or inflammatory activity.

Case 2: A 62-year-old male was found unresponsive in his home after feeling ill and receiving treatment for gastrointestinal complaints. The postmortem chest radiograph showed cardiac enlargement and mediastinal widening (**Image 3A**). Postmortem CT showed massive kidneys composed of multiple cysts of varying size and density (**Image 3B**). Areas of the liver suggested cysts. The heart was enlarged and this was noted on the CT to relate to a large hemopericardium (**Image 3C**). The ascending aorta was widened and had irregular margins (**Image 3D**). At autopsy, the kidneys showed numerous thin-walled cysts (**Image 4A**). A 950 mL bloody pericardial effusion was noted. At the aortic valve annulus, there was a longitudinal intimal tear joining a transverse intimal tear with accompanying false lumen – a Stanford A/DeBakey II dissection (**Image 4B**). Histologic sections demonstrated a split in the media and layered deposition of blood elements, degenerative changes in the media, and mixed inflammation. The liver contained multiple scattered cysts measuring up to 2.5 cm (**Image 4C**).

DISCUSSION

Adults with APKD are known to be at risk for vascular abnormality. The most widely recognized is the intracranial or Berry aneurysm (1). Aortic dissection is also reported and Silverio et al. have reported on 27 cases (two from their clinical experience and 25 from review of the literature) (2). The Stanford A type dissection was seen most often in their series (62% of cases) and our Case 2 matches this pattern with the dissection in the ascending portion of the aorta. In Case 1, the dissection was in the abdominal portion of the descending aorta at the level of the diaphragm and corresponded with the Stanford B type, which was seen in 28% of cases in the Silverio et al. series (2).

The use of postmortem imaging to assist the forensic pathologist is becoming more common; its value

lies in identifying relevant findings that facilitate the autopsy, and in some cases, obviating the need for an invasive procedure. While death determination with postmortem CT is recognized to be more difficult in cases of death by natural causes, there are findings that can point the radiologist and pathologist to the answer (3). In both of our cases, recognition of polycystic kidneys was key. The contour and size of the kidneys together with low-density internal cystic areas were visible on both axial images and multiplanar reconstructions. In the Silverio series, ten of 27 cases manifested liver cysts. The liver appearance was checked and, in Case 2, cystic areas were suggested; none were evident in Case 1. Autopsy confirmed the CT findings concerning presence and absence of liver cysts, but it was noted that liver cyst number and size was better visualized at autopsy. This is in part technical. Kidney and liver visualization in both of our cases was subop-

Image 1: Postmortem computed tomography (CT) findings in Case 1. **A)** Coronal CT shows irregular contours and cystic changes in enlarged kidneys. Coronal **(B)** and axial **(C)** show diffuse retroperitoneal hemorrhage. Margins of the abdominal aorta are obscured.

timal because the scans were performed with the arms at the sides of the body in anatomic position. This introduces beam hardening artifacts. Repeat scanning of the chest and upper abdomen with the arms overhead has been proposed for cases where best possible soft tissue visualization is required (3).

After recognition of the cystic kidneys, the suspicion of aortic dissection was based upon the bleeding pattern. In Case 1, there was diffuse retroperitoneal hemorrhage with obscuring of the proximal abdominal aorta suggesting origin at this level. In Case 2, the enlarged heart showed a pericardial effusion with two components of differing density consisting of an outer ring of lower density surrounding a hyperdense inner ring. Shiotani et al. described this CT finding in 18 of 30 patients with pericardial effusion due to aortic dissection. They termed this the “*hyper-dense armored*

heart” (4). Additional indications of ascending aortic dissection were widening and marginal irregularity. In both of these cases, the autopsy confirmed the location of the dissection. Without an angiographic study, the dissection itself is not defined; however, the presence and probable general location of dissection was possible. The success of postmortem angiography in identifying aortic dissection has been reported along with a cautionary case report where dissection was caused by the procedure (5, 6).

There are multiple conditions in which aortic dissection can occur and the suspected mechanism seems to relate to hypertension and/or tissue abnormality. The association with APKD invokes both of these as possibilities. There are genetic defects in APKD that affect the functional synergy between arterial smooth muscle cells and adjacent elastic tissue. It has been

Image 2: Autopsy findings in Case 1. **A)** Pulmonary emboli, **B)** Cystic right kidney, **C)** 1 cm full-thickness defect on the posterior abdominal aorta approximately 11 cm proximal to the renal arteries.

proposed that this disruption will lead to arterial wall lesions, which account for the vascular complications in APKD (7).

CONCLUSION

Those performing death investigations should recognize the association of aortic dissection and APKD

and be able to use the findings on postmortem CT to aid in the performance of the autopsy. In the future, with added experience, consideration can be given to the use of CT to supplant an autopsy. Presently, the need to specifically identify dissection location requires at least a partial autopsy.

Image 3: Radiographic findings in Case 2. **A)** The postmortem chest radiograph showed cardiac enlargement and mediastinal widening. **B)** Axial computed tomography (CT) showed massive kidneys composed of multiple cysts of varying size and density. **C)** Axial and **D)** coronal CT show that the cardiac enlargement is due to a large hemopericardium with two components: a hyper-dense inner ring, and an outer ring of lower density. The ascending aorta is widened with irregular margins.

Image 4: Autopsy findings in Case 2. **A)** External view of the kidneys showing numerous thin-walled cysts creating a cobbled surface. **B)** At the aortic valve annulus, a longitudinal intimal tear joins a transverse intimal tear with accompanying false lumen. **C)** The liver contained multiple scattered cysts measuring up to 2.5 cm.

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