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Aneurysmal Dilatation of Medium Caliber Arteries in Turner Syndrome

Pritam R. Polkampally, M.D., Jatin R. Matta, PA-C, MPAS, Dorothea McAreavey, M.D.* , Vladimir Bakalov, M.D.†, Carolyn A Bondy, M.D.†, and Ahmed M. Gharib, M.D.

National Institute of Diabetes and Digestive and Kidney Diseases, National Institutes of Health

* Clinical Center, National Institutes of Health

† National Institute of Child Health and Human Development, National Institutes of Health

Turner syndrome is the most common chromosomal abnormality in female subjects, affecting 1 in 2000 live births. 1· 2 The condition is characterized by many clinical abnormalities including short stature, web neck, lymphedema, and variable cardiovascular findings such as bicuspid aortic valve sometimes with aortic stenosis, coarctation of the aorta and increased risk of aortic aneurysm and dissection. 3 Importantly also, all-cause mortality in Turner syndrome is three-fold higher than in the general population at all ages1· 2. We report aneurysmal dilation of medium caliber arteries involving the celiac axis and coronary vessels in two women with Turner syndrome participating in a clinical research protocol approved by the National Institute of Child Health and Human Development Institutional Review Board.

A 53-year old woman with Turner syndrome was noted to have a suspicious lesion on a hepatic ultrasound performed in a research protocol and follow-up with computed tomography (CT) was recommended. Abdominal CT showed a dilated segment in the celiac artery that measured 11 mm in diameter compared to 6 mm in diameter proximally (Figure 1A). The subject had hyperlipidemia, but was normotensive with normal hemoglobin A1C, and had normal coronary arteries by CT angiogram. She had received hormone replacement therapy between ages 10 and 50 years. Her father had had an abdominal aortic aneurysm at age 69 years.

A 44 year old woman with Turner syndrome underwent coronary CT angiography as part of the research protocol. She had minimal calcification involving the circumflex coronary artery and non-calcified plaque in the left anterior descending (LAD) artery without significant stenosis. The mid LAD revealed a small ectatic or aneurysmal segment measuring approximately 4 mm in diameter compared to 2 mm diameter vessel proximally (Figures 1B, 1C). The subject had elevated body mass index, but had normal hemoglobin A1C and no other cardiovascular risk factors. She had received hormone replacement therapy since age 12 years. Neither subject had coarctation or dilation of the aorta and both had tricuspid aortic valve leaflets.

These images illustrate the presence of aneurysms in medium caliber vessels in Turner syndrome. There are insufficient data to determine the prevalence of aneurysms in this syndrome or to compare with age and gender -matched controls. The aneurysms may reflect the predisposition to atherosclerosis described in Turner syndrome 1· 2 even though neither

subject had hypertension, insulin resistance, estrogen deficiency or significant atherosclerosis on imaging. Other less frequent causes of aneurysms in medium size vessels such as fibromuscular dysplasia, collagen vascular diseases, infection, and cystic medial necrosis 4 were not present in the subjects.

Celiac aneurysms are extremely rare; the described aneurysm is small and thus, the risk of rupture is very low. The subjects are under periodic surveillance because the aneurysms could be relevant if cardiovascular or abdominal symptoms develop. Early recognition and intervention are crucial particularly because rupture of a celiac aneurysm is associated with significant clinical morbidity and mortality⁵.

The findings provide a reminder that Turner syndrome is associated with a generalized vasculopathy as well as congenital cardiac and other defects.

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Figure 1. Panel A demonstrates computed tomography angiogram (CTA) three dimensional shaded surface display of abdominal aorta showing aneurysmal dilatation of the celiac artery (black arrow). Panels B and C demonstrate CTA of the coronary vessels with a small aneurysm in the mid left anterior descending coronary artery seen on three dimensional shaded surface display (black arrow on B) and multiplanar reconstruction stretch views (red arrow on C). Figure 1.