Virtua Cardiology Performs Nuclear Scintigraphy for Cardiac Amyloidosis

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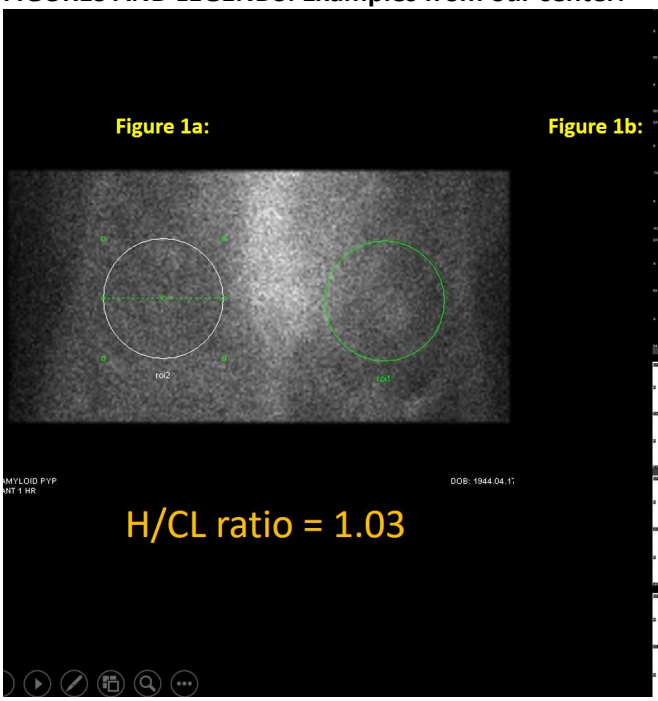
Cardiac amyloidosis (CAm) is an infiltrative restrictive cardiomyopathy resulting from myocardial deposition of misfolded amyloid protein causing a diverse spectrum of systemic diseases. Most cardiac cases result from two protein precursors: amyloid immunoglobulin light-chain (AL, wherein protein is monoclonal immunoglobulin light-chain) and amyloid transthyretin (ATTR, wherein proteins transthyretin serum transport protein). Different types of CAm display heterogeneity in clinical course, prognosis, and treatment approach.

At Virtua Cardiology we are now performing standardized protocol for cardiac amyloid nuclear scanning. Radionuclide imaging using bone-avid radiotracer **Technetium-99m pyrophosphate** (99mTc-PYP) has emerged as an alternative to biopsy for diagnosing ATTR amyloidosis. **In the presence of a Grade 2-3 positive 99mTc-PYP, cardiac scan without evidence for monoclonal proteins in blood and urine, diagnosis of ATTR cardiac amyloidosis can be made without a biopsy (specificity and PPV >98%).**

Diagnosis of CAm remains challenging due to clinical overlap with more common diseases causing LVH (i.e., HTN, CKD, hypertrophic cardiomyopathy, aortic stenosis). ATTR appears quite common compared to AL with prevalence 10 to 16% of older patients with CHF and aortic stenosis. The most common mutation in ATTR (Val122Ile) has been demonstrated in 3 to 4% of African-Americans, suggesting nearly 2 million carriers in U.S. with risk of CAm.

Treatment options are rapidly expanding. ATTR amyloidosis now have several trials of monoclonal-antibodies, novel therapeutics that suppress TTR expression (FDA approved) and are successful in reducing all-cause mortality. It is evident early diagnosis will be essential to afford the most effective treatment option.

FIGURES AND LEGENDS: Examples from our center:



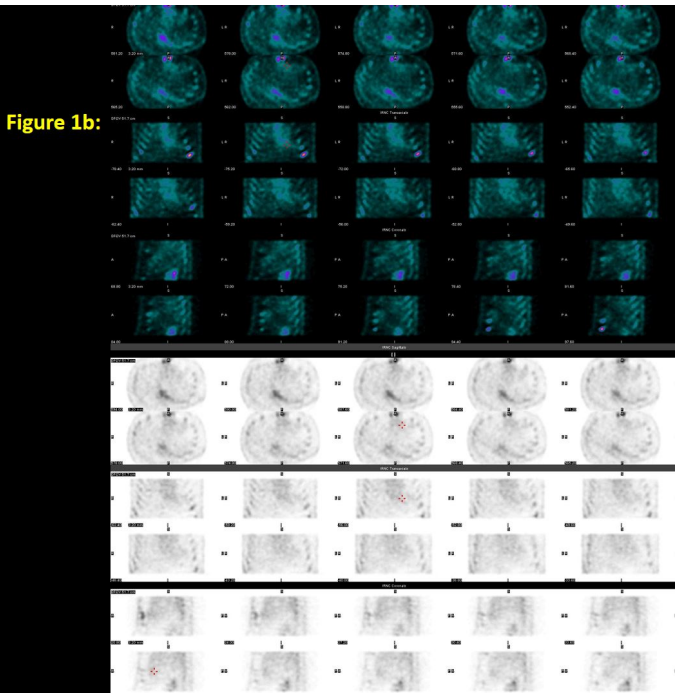


Figure 1: Amyloid scan negative for ATTR Cardiac Amyloidosis (Normal) in a patient with Grade 0 uptake:

1a: Frontal planar image showing normal sternal and rib uptake but no 99mTc-PYP uptake in myocardium. Semiquantitative tracer count ratio- Heart/Contralateral ratio < 1.5. Region of interest (circle) should be positioned to minimize overlap with sternal or focal rib uptake, and maximize coverage of the heart without including adjacent lung.

1b: SPECT image distribution (color and black/white) in same patient showing no 99mTc-PYP uptake in myocardium.

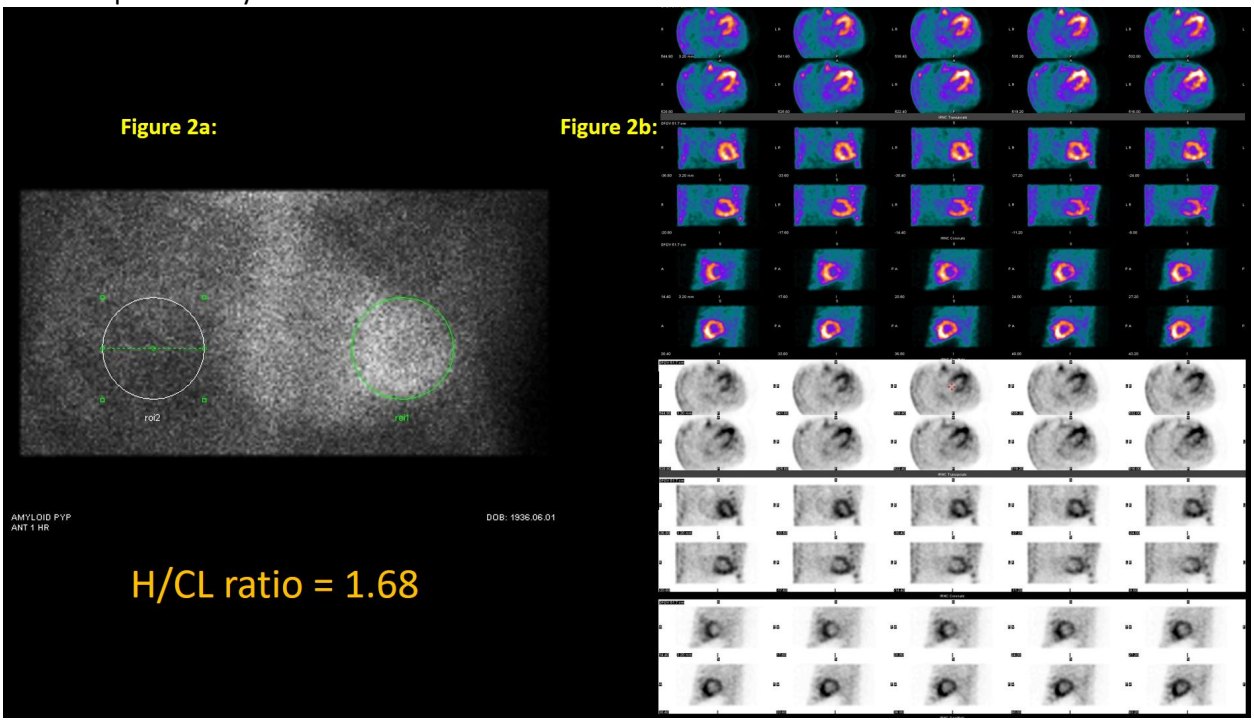


Figure 2: Amyloid scan positive for ATTR Cardiac Amyloidosis (abnormal) in a patient with Grade 3 uptake:

2a: Frontal planar image showing normal sternal and rib uptake but no 99mTc-PYP uptake in myocardium. Semiquantitative tracer count ratio- Heart/Contralateral ratio > 1.5.

2b: SPECT image distribution (color and black/white) in same patient showing 99mTc-PYP uptake in all segments of myocardium.