DISEASE EVOLUTION OF ATTR AMYLOIDOSIS OBSERVED ON A BONE-SCAN.

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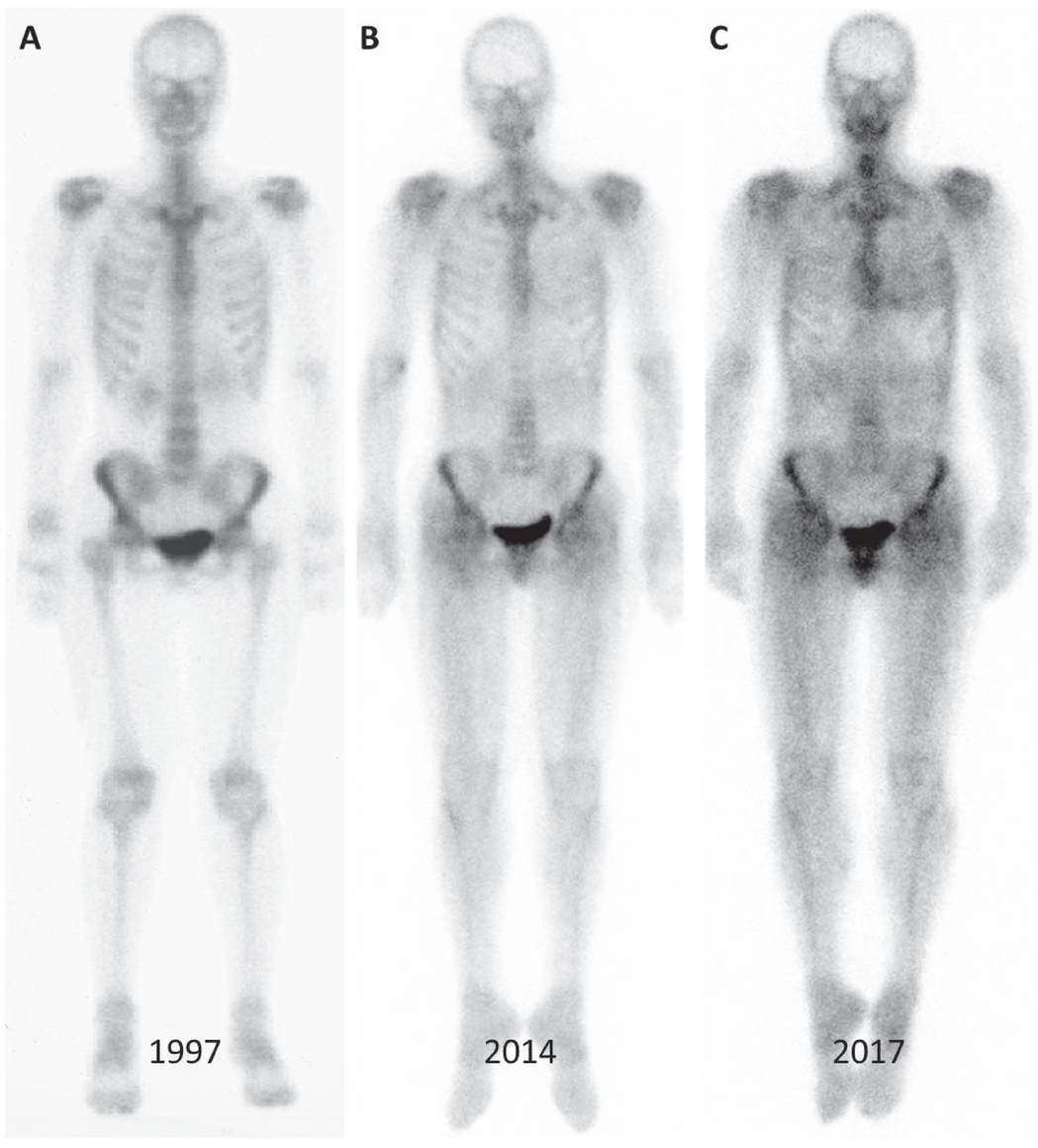
Case

A 30-year-old female was referred to our hospital in 1997 for arthralgias in the presence of a family history (mother and uncle) of hereditary ATTR Val71Ala amyloidosis. During work-up a bone scintigraph was performed that did not yield specific information. Non-specific arthralgias was the diagnosis at that time.



Initial assessment for systemic amyloidosis proved negative. Seven years later (2004) she was seen again because of concerns regarding the family history of ATTR amyloidosis. She then turned out to be carrier of the TTR Val-71Ala mutation and again no symptoms or signs of ATTR amyloidosis were found. In 2008 and 2009 vitreous opacities were removed from both eyes that contained amyloid. In 2014 amyloid (1+) was detected in subcutaneous abdominal fat tissue. Cardiac or neurologic symptoms were still lacking in 2017.

Results

Three sequential ^{99m}Tc-diphosphonate bone scans were performed during 20 years. Seventeen years after initial assessment no clinical or biochemical abnormalities were detected whereas minute amyloid was found in fat tissue. The bone-scan, however, showed increased tracer uptake within the myocardium and pronounced in soft-tissue. Another 3 years later progression of uptake was seen on the latest bone-scan.

Figure 1. Three consecutive ^{99m}Tc-HDP bone-scans.
A. Normal tracer bio distribution with a slight increase in uptake in the larger joints, no signs of cardiac involvement. B. Abnormal tracer bio distribution with increased uptake within the soft tissue and strongly increased uptake surrounding the shoulder and hip joints. Furthermore, increased cardiac tracer uptake (grade 1). C. Progressive disturbance of the tracer bio distribution with strongly increased soft tissue uptake, accompanied by (relatively) decreased bone uptake. Also, cardiac uptake appears to be more pronounced (grade 2).

Conclusion

- Further studies of the relevance of bone-scans in the follow-up of ATTR patients and carriers are needed.
- This relevance may even be present before the onset of clinical signs of systemic involvement.

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