Reviewer's report

Title: Paroxystic atrial fibrillation as the first symptom of light chain deposition disease: a case report

Version: 5 Date: 16 October 2007

Reviewer: Elsayed Soliman

I am familiar with the literature and believe that this case meets one of the 7 criteria for evaluation in the journal: Unexpected or unusual presentations of a disease

Has the case been reported coherently?: Yes

Is the case report authentic?: Yes

Is this case worth reporting?: Yes

Is the case report persuasive?: Yes

Does the case report have explanatory value?: Yes

Does the case report have diagnostic value?: Yes

Will the case report make a difference to clinical practice?: Yes

Comments to authors:

General

The authors present a case of paroxysmal atrial fibrillation (AF) as an unusual first manifestation of Light Chain Deposition Disease (LCDD). The authors have effectively presented the case in an interesting way supported by a nice electron microscopy photograph. However their conclusion that “the onset of acute arrhythmias (such as AF) associated with an increase in serum creatinine might conceal a relatively rare disease such as LCDD” should be revised. AF and slight elevation of creatinine (1.4 mg/dl, in this case) are common in all patients with cardiac problems even in the early stages. Hence, no specific diagnosis or even direction towards a specific diagnosis could be suspected based merely on the presence of AF and mild renal impairment in such patients, nevertheless a rare one like LCDD. Add on, although it is unusual for LCDD to be manifested “early” by cardiovascular manifestations, it is usual to see arrhythmias such as AF occurring once the heart gets involved by diseases such as restrictive cardiomyopathy (as in the reported case). Therefore, I may recommend that the direction of reporting this medical case could be changed to address cardiovascular manifestations (signs and symptoms of restrictive cardiomyopathy...
in this case) as an unusual “first” manifestation of the LCDD. The fact that cardiovascular manifestations such as restrictive cardiomyopathy occur in advanced stages of LCDD not as an early manifestation makes representing this case an interesting thing. Using restrictive cardiomyopathy (not AF which is mostly secondary to restrictive cardiomyopathy) as an entry point for differential diagnosis makes more sense because although rare, LCDD is on the list of differential diagnosis of restrictive cardiomyopathy especially in comparison with amyloidosis. This would emphasize on considering all causes of restrictive cardiomyopathy when making treatment decisions.

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Revisions necessary for publication

(1) It was not clear to me if the authors think that AF is a direct manifestation of LCDD (because of deposition of light chain fragments in the heart conductive system, for example) or secondary to restrictive cardiomyopathy. If it is the later, restrictive cardiomyopathy should be then the cardiovascular manifestation to be addressed, as mentioned above. If it is the earlier, it would be necessary then to mention why they think that AF is not secondary to restrictive cardiomyopathy which is actually the most reasonable explanation for such an AF.

(2) The conclusion should be cautiously stated to avoid implying that presence of AF and mild impairment of renal function should be considered as a clue for possible rare disease such as LCDD. Just mentioning what it is unusual in this case would be enough as a conclusion

(3) As liver involvement (specially cholestatic jaundice) is one of the common late extra-renal manifestations of LCDD, it would be interesting to know the reason for which the patient underwent cholecystectomy. Liver involvement could be another unusual early manifestation of LCDD in this patient.

(4) In the title (and in the text), using the word “Paroxysmal” would be preferred rather than “paroxystic” on referring to the intermittent AF.

What next?: Accept after minor revisions

Quality of written English: Needs some language corrections before being published