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AL amyloïdosis, a rare and challenging complication of Waldenström macroglobulinemia.

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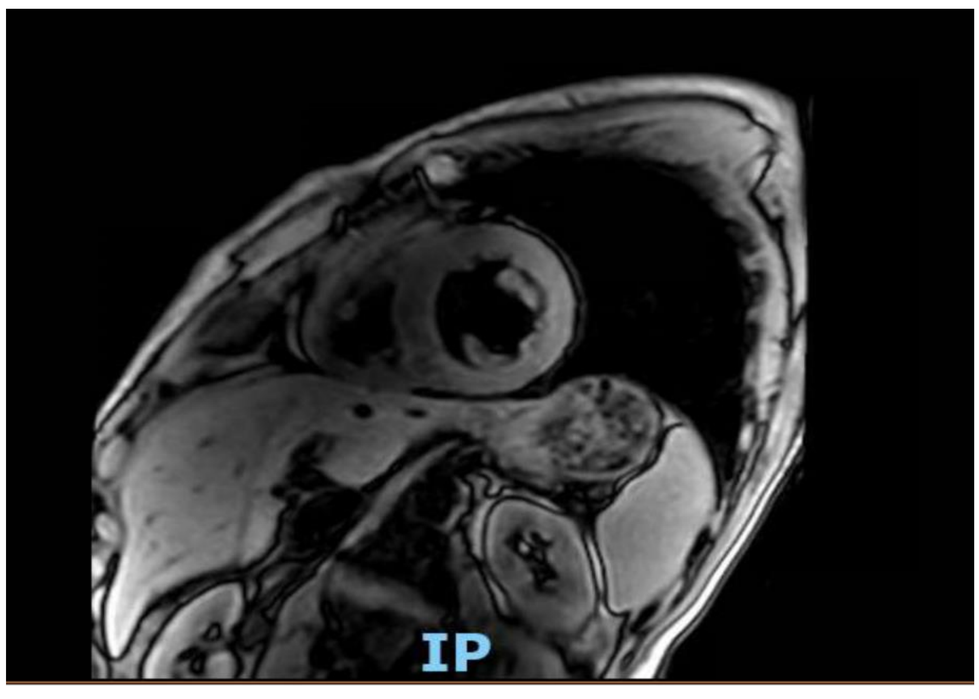
ABSTRACT

Waldenström Macroglobulinema (WM) is a rare B-cell disorder characterized by a bone marrow infiltration of lymphoplasmacytic cells, along with the identification of an IgM monoclonal gammopathy in the serum. Almost all cases are symptomatic at presentation, with symptoms related to either tumor burden or the IgM paraprotein. Light chain amyloïdosis is a rare complication of WM that occurs in 3% of cases and represents less than 5% of AL amyloïdoses. Here we report a case of cardiac AL-amyloïdosis that achieved a partial organ response after Daratumumab therapy.

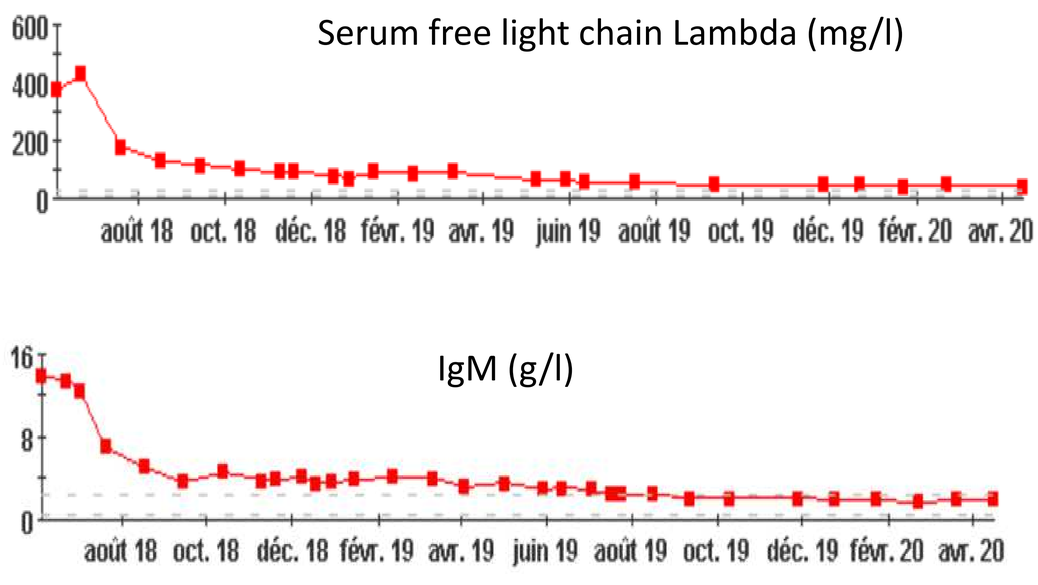
CASE REPORT

A 54 year-old man repeatedly complained of shortness of breath that was first attributed to coronaropathy that failed to improve after stenting. He had a 5-year history of IgM MGUS. Echocardiography identified a thickening of the septum that led to the diagnosis of AL lambda amyloidosis related to WM. Initial work-up pointed out a Mayo stage 3 cardiac involvement. He was successively treated with 5 cycles of Rituximab-bortezomib-dexamethasone suspended for a grade 2 peripheral polyneuropathy, followed by 5 cycles of Rituximab-bendamustine without any relevant hematological response or cardiac improvement. He finally achieved a complete hematological remission after six cycles of Daratumumab monotherapy, with a slightly progressive improvement of cardiac function. Treatment is ongoing with the hope to propose autologous transplantation.

Cardiac MRI



Paraprotein evolution



CONCLUSION

IgM-related AL amyloidosis is a distinct entity with a less favorable outcome compared to non-IgM amyloidosis; therefore, early diagnosis is critical for organ prognosis. However, in this case, the delay between the onset of symptoms and diagnosis was over one year. There is no consensus regarding the treament of this disease. Tackling the lymphoid clone remains the main goal in order to achieve an adequate response. This case highlights the potential role of immunotherapy with Daratumumab.

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