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◇ CASE REPORT ◇

心臓超音波検査にて心外膜のアミロイド沈着を疑い剖検により同定された全身性ALアミロイドーシスの1例

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抄　録

症例は60代男性．20XX年Y－8ヵ月より下腿浮腫と労作時呼吸困難を自覚した．20XX年Y－4ヵ月に症状増悪のため前医を受診し，心不全の診断で内服加療が開始された．その後も症状は改善せず，20XX年Y月に当院循環器内科を紹介受診した．外来で精査予定となっていたが，呼吸困難，腹痛，下痢を訴え当院へ救急搬送．消化器症状の原因断定は困難であったが，徐脈と低血圧は持続することから，一時的ペースメーカーを挿入し入院となった．入院第4病日に洞不全症候群に対して恒久的ペースメーカーを挿入．入院第6病日の心臓超音波検査で，左室駆出率47%，左室拡張障害，両心肥大，心外膜の輝度亢進と肥厚を認め，長軸方向ストレインでのapical sparingより心アミロイドーシスを疑った．上下部消化管内視鏡検査を行ったところ，胃および大腸にびらんを認め，生検でアミロイド沈着があり，免疫染色でAL（κ型）の診断となった．全身性ALアミロイドーシスに伴う心不全・消化管症状と診断され，治療が開始された．しかし，心不全は改善せず，入院第48病日に心停止となった．剖検が施行され，アミロイドは心臓，消化管，肺，腎臓等，複数の臓器に沈着を認め，心筋組織にはびまん性，心外膜には脂肪組織に入り込むように不均一に沈着していた．心筋組織へのアミロイド沈着が一般的だが，心外膜に沈着を認める症例は稀であり，心臓超音波検査でその可能性が示唆されていたことから，文献的考察を含めて報告する．

A case of systemic AL amyloidosis identified by autopsy with suspected epicardial amyloid deposits on echocardiography

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Abstract

A man in his 60s complained of lower leg edema and exertional dyspnea on Y-8 months, 20XX. He visited a former doctor on Y-4 months, 20XX because of worsening symptoms. He was diagnosed with heart failure and treated with medications. He was referred to our Department of Cardiology on Y month, 20XX because his symptoms persisted. He was scheduled to undergo examinations on an outpatient basis, but he complained of dyspnea, abdominal pain, and diarrhea and underwent emergency hospitalization. Although it was difficult to determine the cause of gastrointestinal symptoms in the emergency department, bradycardia and hypotension persisted. He was admitted and underwent temporary pacemaker implantation. A permanent pacemaker was implanted for sick sinus syndrome on the 4 th hospital day. On the 6th hospital day, echocardiography showed a left ventricular ejection fraction of 47% , diastolic dysfunction, bi-ventricular hypertrophy, epicardial brightness and thickening, as well as apical sparing on longitudinal strain. Cardiac amyloidosis was suspected based on these findings. Upper and lower gastrointestinal endoscopy revealed erosion in the stomach and large intestine. Biopsy showed amyloid deposits, and immunostaining revealed the ALκ type. Based on these findings, systemic AL amyloidosis was diagnosed. Although medications were started, the symptoms of heart failure persisted, and cardiac arrest and death occurred on the 48 th hospital day. An autopsy was performed to evaluate the pathology of the underlying diseases, and amyloid was found to be deposited in multiple organs including the heart, gastrointestinal tract, lungs, and kidneys. It was diffusely deposited in myocardial tissue and unevenly deposited in the epicardium in a way that penetrated adipose tissue. Amyloid deposits in myocardial tissue are common, but cases with amyloid deposits found in the epicardium are rare, and given that the possibility of this was suggested by echocardiography before death, we report this case with a review of the literature.

Keywords

systemic AL amyloidosis，amyloid，epicardium

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