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Cardiovascular disease in patients with autoinflammatory syndromes

A systematic review

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Introduction

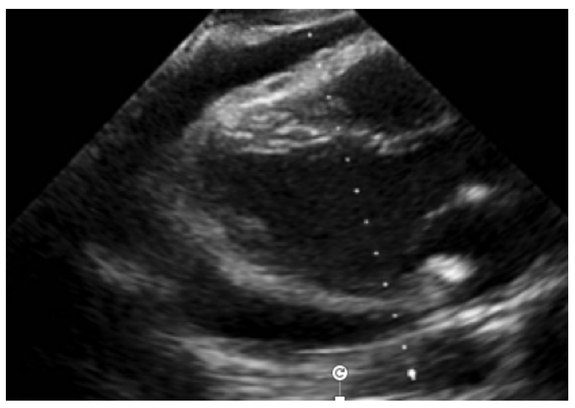
Autoinflammatory syndromes (AIS) are characterized by recurring events of inflammation, leading to a variety of organ manifestations and fever attacks. A subgroup of AIS are commonly referred to as hereditary periodic fever syndromes (HPFS).[1] There is substantial evidence that autoimmune diseases such as rheumatoid arthritis and systemic lupus erythematosus are strongly associated with cardiovascular morbidity and mortality. The link between AIS and cardiovascular disease (CVD) is not that clear, even if the concept of continuous inflammation as a risk factor for cardiovascular disease is widely accepted.[2,3] In this review we found several entities linked with CVD as shown in table 1.

Table 1

AIS with cardiovascular manifestations

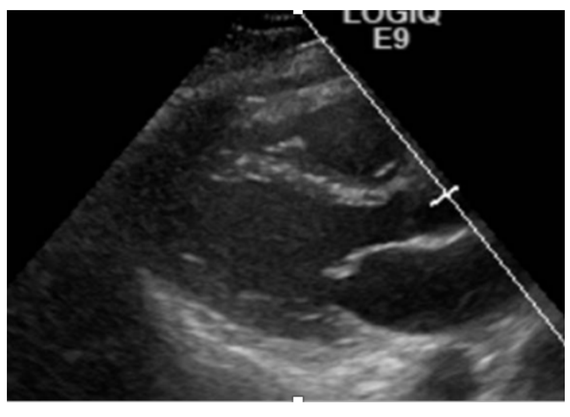
Behçets disease (BD)
Familial Mediterranean Fever (FMF)
Adult-onset Still´s disease (AOSD)
Systemic juvenile idiopathic arthritis (SJIA)
TNF receptor-associated periodic syndrome (TRAPS)
Synovitis, acne, pustulosis, hyperostosis, and osteitis (SAPHO)
Cryopyrin-associated periodic syndromes (CAPS)
Neonatal-Onset Multisystem Autoinflammatory Disease (NOMID)
Pyogenic arthritis, pyoderma gangraenosum, and acne (PAPA)

Figure 1



62 year old female patient with pericardial effusion diagnosed with IRAP

Figure 2



Same patient after 6 months of therapy with the Interleukin-1 antagonist Anakinra

Methods

For this systematic review, literature search was performed using Ovid´s Medline. Our search displayed 5966 results from which we retained 121 articles and reviews. In addition, we screened references of retrieved articles for further relevant papers and included them where appropriate.

Results

Based on the rarity of some entities, lack of data however led to exclusion of some rare AIS. Especially for behcet´s disease (BD), adult-onset still´s disease (AOSD) and familial mediterranean fever (FMF) there is an association with a number of cardiovascular abnormalities (table 2). BD is the AIS, which is most strongly associated with manifestation in the arterial and venous system. AOSD is strongly associated with cardiac inflammation. Serositis is common in patients suffering FMF.

Table 2

cardiac and vascular manifestations in BD, FMF and AOSD

	cardiac manifestations	*vascular manifestations*
BD	Aortic valve regurgitation Congestive heart failure Coronary artery aneurysms Coronary artery disease Diastolic and systolic dysfunction Intracardiac thrombi Myocardial infarction Myocarditis	Aneurysms Budd-Chiari syndrome Deep vein thrombosis Dural sinus thrombosis Inferior vena cava thrombosis Pulmonary artery aneurysm Pulmonary artery thrombosis Superior vena cava thrombosis Thrombophlebitis
FMF	Amyloidosis Cardiac tamponade ECG-abnormalities IRAP (figure 1 and 2) Myocardial infarction Pericardial effusion Rheumatic carditis	Atherosclerosis Henoch-Schönlein Purpura Polyarteritis Nodosa
AOSD	Aortic regurgitation Cardiac tamponade Endocarditis Myocarditis Pericardial effusion Pericarditis	Portal vein thrombosis

Conclusion

In contrast to autoimmune diseases, the link between AIS and cardiovascular disease is not that clear, even if inflammation is a feature of both. Due to the rarity of many AIS, little is known about related cardiovascular manifestations for a number of those diseases. However, cardiac and vascular manifestations of AIS may also be characteristic for single AIS and may even be a clue for the correct diagnosis. To summarize, cardiac and vascular manifestations are relatively rare, but clinicians should pay attention to these potential life-threatening complications.

references:

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