The Cardiovascular Manifestations of Certain Rheumatological Disorders

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Abstract: Chiropractors and other practitioners of spinal manipulative therapy (SMT) are often called upon to treat the musculo-skeletal symptoms of patients suffering from a wide variety of rheumatological disorders. The SMT practitioner may be the primary contact, or alternatively these patients may seek help from the SMT practitioner after diagnosis from a medical practitioner. Either way it is essential that whoever treats these patients for their musculo-skeletal symptoms is fully cognisant of the cardiovascular manifestations which may accompany rheumatological disorders. This paper discusses ten rheumatological disorders and their possible cardiovascular manifestations.

Key Words: Cardiovascular disease, heart disease, arthritis, rheumatological disorders, connective tissue disease, spinal manipulative therapy, chiropractic.

Connective tissue diseases display certain common clinical features, including inflammation of the joints, serosal membranes, and blood vessels, as well as a high frequency of involvement of the internal organs rich in connective tissue. In addition they are all characterised anatomically by generalised alterations of, or in, the connective tissues.

The cardiovascular manifestations of the rheumatological conditions listed in Table 1 can be broadly divided into three pathological processes: inflammation of the connective tissues of the heart and its vessels, fibrosis of the connective tissues, and weakening of the connective tissues.

RHEUMATOID ARTHRITIS (RA)

As with the majority of these arthropathies the etiology of RA is unknown but there appears to be a genetic predisposition which results in a self-perpetuating immune response to a triggering agent, most probably a virus. The pathological consequences of this abnormal response are:-

a) A vast accumulation of plasma cells, lymphocytes, and macrophages within the connective tissues, which produce factors that destroy collagen and other structural components.

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b) The formation of nodules which contain a palisade of chronic inflammatory cells.

c) Vasculitis due to the circulating immune complexes which are deposited in vascular epithelium, generating local inflammation and infarction (1).

Pericarditis with associated friction rub is the most common clinical cardiac manifestation of rheumatoid arthritis and is found in between 10% and 30% of RA patients. The inflammatory response results in the accumulation of pericardial fluid, but generally not to the extent of tamponade. Nodules may also develop in the myocardial tissue resulting in functional impairment and in addition, coronary arteritis may result in myocardial infarction (1).

RHEUMATIC FEVER (RF)

Rheumatic Fever develops after a haemolytic streptococcal infection, creating a cardiac reaction which appears to be a cross-reactive immune response. RF affects the connective tissue of the heart by a fragmentation of the collagen fibres, fibrinoid degeneration, damage to cardiac muscle cells, and the formation of the pathognomonic Aschoff bodies (2). Apart from pericarditis in which both layers of the pericardium become thickened and covered with a fibrinous exudate, RF also results in myocarditis, and endocarditis. Myocarditis is characterised first by interstitial oedema, and then by myocytolysis, loss of striation, fatty degeneration, and vacuolation of the muscle fibres (3). Chronic endocardial inflammation

leads to ingrowth of small blood vessels into the valve cusps and scarring caused by new fibrous connective tissue. The valvular and chordal tissues gradually become fibrosed and eventually calcified and immobile. Permanent heart valve damage is the most important complication of RF, with the mitral being affected in nearly all patients, the aortic in 30-35%, and the tricuspid in 15% (2).

ANKYLOSING SPONDYLITIS (AS) AND REITER'S SYNDROME (RS)

These two disorders are classed as seronegative spondyloarthridities which primarily affect the tendon and ligament insertions, (enthesopathy) rather than the synovium as in RA. The pathogenesis of these diseases is not fully understood, as is their preference for the tendinous insertions, which pathologically results in an initial inflammatory reaction followed by fibrosis and ossification (3). AS in particular may produce aortitis which appears to start from perivascular lymphocyte infiltration, followed by necrosis, and then the replacement of normal muscle and elastic cells with dense scar formation (4). This is associated with an overlying thickening and atrophy of the intima, which can affect the entire length of the aorta. Destruction near the aortic valve may lead to stretching of the aortic ring and result in valvular incompetence and chronic aortic regurgitation. Additionally fibrosis within the heart tissue can result in heart block of any degree (5). RS can produce almost identical aortic changes as AS, as well as pericarditis and conduction defects (6,7).

SYSTEMIC LUPUS ERYTHEMATOSUS (SLE)

SLE is an inflammatory disease with multi-system involvement and is characterised by the development of distinct immunologic abnormalities, especially antinuclear antibodies (8,9). The most common cardiac manifestation is pericarditis, which has been found at autopsy in 80% of cases, in which there was either fluid accumulation or pericardial thickening. Pathologically the pericardium may reveal foci of inflammatory lesions, in which mononuclear cells predominate, or healed fibrous residua (8). Myocarditis is also often seen and is associated with tachycardia, ECG abnormalities, and unexplained cardiomegaly, with the latter often resulting in congestive heart failure, conduction abnormalities, and arrhythmias (8). As with the pericarditis of SLE the myocardium reveals an infiltration of mononuclear cells and fibrosis from healing, resulting in continued cardiac dysfunction(8).

SCLERODERMA

Scleroderma is a chronic connective tissue disease dominated by cutaneous manifestations, which initially begin with a prefibrotic stage with marked oedema, followed by the classical fibrotic stage, with thickening, atrophy and tethering, deformity, and loss of normal skin elasticity (9). The etiology and pathogenesis of scleroderma is unknown. Myocardial fibrosis has been noted in up to 80% of patients and is associated with intermittent microvascular ischemia of the myocardium. Clinically the patient may present with ventricular gallops, sinus tachycardia, signs of congestive heart failure, and pericarditis, due to thickening and fluid accumulation (10).

POLYARTERITIS NODOSA (PAN)

PAN is characterised by inflammation and necrosis of blood vessel walls and closely resembles the vascular pathology of AS and RS (11). Following the inflammation of the artery wall there is fibrous proliferation in the intima, which narrows and often occludes the lumen, therefore causing ischaemic change to the organ being supplied (12). The normal architecture of the vessel wall, including the elastic laminae is disrupted and may result in thrombosis and aneurysmal dilation. The heart has been shown to be affected in 70% of cases at autopsy, usually involving diffuse arteritis of the myocardium. Myocardial infarction, congestive heart failure may result from coronary insufficiency or hypertension, or both (13,14).

POLYCHONDRITIS

The essential features of Polychondritis are inflammation with progressive loss of the structural integrity of some cartilaginous tissues. The effect of these changes on the cardiovascular system can be quite serious as they may result in aortic regurgitation, due not to valvulitis of the aortic valve, but rather to progressive dilation of the aortic ring and frequently the descending aorta (15).
Destruction of the valve cusps may occur also, and aneurysms of the ascending thoracic and abdominal aorta may develop (16).

JUVENILE RHEUMATOID ARTHRITIS (JRA)

Like adult rheumatoid arthritis JRA is an inflammatory arthritis and although the two disorders are of the same family they differ in several important ways, predominantly, in the mode of onset and the disease course (17). Pericarditis is especially common in JRA but is often subclinical , and effusion does not usually progress to tamponade or result in constrictive pericarditis. Myocarditis is less common in JRA but when present may produce cardiac enlargement and or failure. Nodular formation is also less common but has been found in the valve leaflets and pericardium of JRA patients (18).

MIXED CONNECTIVE TISSUE DISEASE (MCTD)

MCTD embraces those patients who clinically and serologically do not fit into any one diagnostic group. MCTD predominantly shows an overlap between SLE, Scleroderma, and myositis and may manifest itself cardiovascularly in the same fashion as those disorders (19).

It must also be remembered that these rheumatological disorders may indirectly affect the cardiovascular system. As the majority of these diseases are systemic they may also impair the function of other organs like the kidney and lungs leading for example to hypertension and thus added stress on the cardiovascular system.

CONCLUSION

Although some of the aforementioned disorders would not be commonly seen in chiropractic general practice, it is not unusual for patients to present with the likes of ankylosing spondylitis or rheumatoid arthritis, and often with these conditions previously undiagnosed. As primary contact practitioners it is essential that we recognise and are aware of the sometimes life threatening cardiovascular manifestations of these rheumatological disorders.

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Table 1.

Rheumatological disorders which may present with cardiovascular manifestations.

MANIFESTATION	RHEUMATIC FEVER	S L E	REITER'S SYNDROME	POLY-CHONDRITIS	POLY-ARTERITIS	MIXED CONNECTIVE TISSUE DISEASE	RHEUMATOID ARTHRITIS	JUVENILE RHEUMATOID ARTHRITIS	ANKYLOSING SPONDYLITIS	SCLERODERMA
PANCARDITIS	COMMON 50%	RARE								
PERICARDITIS	COMMON	COMMON 80%	INFREQUENT	INFREQUENT	RARE	FREQUENT	OFTEN 10%	COMMON 7-45%	RARE	INFREQUENT
MYOCARDITIS	COMMON	OFTEN 8-25%		INFREQUENT	COMMON 70%	FREQUENT	RARE	OCCASIONAL	RARE	FREQUENT
ENDOCARDITIS	COMMON	UNCOMMON	RARE	INFREQUENT	RARE		RARE	RARE		
CARDIOMEGALY	COMMON	OFTEN				COMMON	RARE	OCCASIONAL	OFTEN	
VALVULAR DEFORMITY	COMMON	UNCOMMON		COMMON						
CONGESTIVE HEART FAILURE	COMMON	OFTEN			INFREQUENT	FREQUENT	RARE			OFTEN
CORONARY ARTERITIS					COMMON					
CORONARY ARTERY DISEASE		OCCASIONAL 2-8%					OCCASIONAL			
MYOCARDIAL INFARCTION		OCCASIONAL		INFREQUENT	COMMON					
ANEURYSMS				COMMON 30%	COMMON					
AORTIC REGURGITATION			INFREQUENT						OFTEN 10%	
CLINICAL SIGNS										
CONDUCTION ABNORMALITIES	COMMON	COMMON 34-70%	INFREQUENT	INFREQUENT	COMMON	COMMON			OFTEN	COMMON 50%
TACHYCARDIA		OFTEN				COMMON				OCCASIONAL
ARRHYTHMIA										COMMON
FRICTION RUBS	COMMON	OFTEN 8-48%	INFREQUENT				OFTEN	OCCASIONAL		OCCASIONAL
MURMURS	OFTEN	COMMON 16-44%				INFREQUENT		OCCASIONAL		
PERICARDIAL PAIN	COMMON	OFTEN 8-48%								