RELAPSING POLYCHONDRITIS

A Fleeting Form of Systemic Arthritis

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Relapsing polychondritis is a systemic rheumatic disease that destroys cartilage. It can occur alone or in patients with other autoimmune disorders.

Fleeting Arthritis with Ear Involvement

Relapsing polychondritis (RPC) is a rare rheumatic autoimmune disorder that causes inflammatory lesions in cartilage and connective tissue, primarily cartilage in the ear, eyes, and trachea. Tissue inflammation in these collagen-rich tissues is known as chondritis, whereas the term polychondritis signifies that multiple locations in the body are eventually affected. Relapsing refers to the episodic nature of this disorder. Periods of remission are interrupted by acute episodes of active disease. Symptoms also vary in severity, usually becoming progressive over time.

Systemic Involvement

RPC is classified as a systemic disorder because various tissues located in different parts of the body may be affected. In most cases, RPC affects and has the potential to destroy cartilaginous tissues in the ears, nose, larynx, joints, pulmonary (lungs) bronchi, ribs and the trachea. Other organs and tissues may also be affected, including the eyes, heart, kidney, blood vessels, and central nervous system. RPC usually occurs alone although it may occur in patients with generalized vasculitis (inflammation of blood vessels) or in patients with other autoimmune disorders.

Who is Affected?

Relapsing polychondritis was first described in 1923. It primarily affects patients between the ages of 30-50 although patients of any age, including newborns, may be affected. Males and females are affected equally. The incidence of RPC is rare although it may be under-reported, RPC is difficult to diagnose since many symptoms are seen in several different conditions.

The autoimmune mechanism in RPC is not clearly defined although patients with RPC typically have autoantibodies to native collagen proteins (anti-collagen II). In addition, individuals with the immune system marker HLA-DR4 are at increased risk for developing RPC. RPC has also been known following trauma to the ear, including piercings.

Signs and Symptoms
Patients with RPC typically present with symptoms of gradual hearing loss, visible ear changes causing the ear to appear floppy although the earlobe is spared, hoarseness, sore throat, cough, nasal congestion, nose swelling (saddle ear), or shortness of breath. Patients who show signs of pulmonary involvement and shortness of breath often tend to have more severe or critical disease courses. Overall, ear involvement (auricular chondritis), which is characterized by redness and painful swelling affecting one or both ears and hearing loss, is the primary complaint in 55 percent of patients; ocular or eye symptoms (such as scleritis, sicca syndrome, painful exophthalmos (proptosis or eye bulging), opthalmoplegia, keratoconjunctivitis, conjunctivitis, uveitis and chondritis) occurs in 32 percent of patients; painful nasal chondritis in 30 percent of patients; and an intermittent form of arthritis, which occurs in conjunction with sensory symptoms and eventually affects about 70 percent of patients.

Arthritis in RPC tends to affect the small joints in the ribs, hands, feet, chest and spine, and it often resembles rheumatoid arthritis or ankylosing spondylitis although it is not erosive. Patients with cardiac involvement may show signs of mitral valve regurgitation. Most patients develop dermatological changes, most prominently mouth sores or skin ulcerations. Less frequently patients may develop nervous system symptoms, such as headache, seizures, or cognitive changes.

Vasculitis may also occur as part of the disease process or as a consequence of another co-existing autoimmune disorder. RPC has been found to occur in patients with myasthenia gravis, diabetes mellitus, autoimmune thyroid diseases, primary biliary cirrhosis, vitiligo, mixed connective tissue diseases, and other autoimmune disorders.

**Diagnosis**

Studies of joint fluid and tissue in patients with RPC show changes similar to those seen in rheumatoid arthritis. Tissue samples taken from affected sites taken during biopsies show depleted amounts of proteoglycans in cartilage and significant inflammation. Inflammation is indicated by an infiltration of plasma cells and white blood cells. As the disease progresses the tissue becomes granular with little regeneration of cartilage. This granulation leads to narrowing of the respiratory passages and the development of nodules in the ears. When the aorta is involved, the heart loses its normal elasticity, and lymphocytic white blood cells cluster in the outer media of the aorta. These changes, which can lead to aortic aneurysm, are similar to those seen in Marfan's syndrome, idiopathic medial necrosis, ankylosing spondylitis, and Rieter's syndrome. Nearly 25 percent of patients develop kidney lesions. A urinalysis is useful for detecting protein, blood and microalbumin. Tests for collagen II autoantibodies are usually positive.

Pulmonary function tests in patients with laryngeal or tracheal involvement show diminished function in both inspiration and expiration. The erythrocyte sedimentation rate (ESR or sed rate) is typically elevated, and patients may have mild anemia. Imaging tests may show the presence of calcifications, tracheal thickening, or nodules in cartilage.
Current diagnostic criteria for RPC include cartilaginous tissue inflammation in two of three sites (external ear, nasal cartilage, or larynx/trachea), and two other features, including eye inflammation, ear (audiovestibular) damage, or arthritis in the absence of RF. Biopsy is no longer considered necessary for diagnosis although it can be helpful in distinguishing RPC from other rheumatic conditions. A muscle biopsy and a blood test for creatinine phosphokinase (CPK, CK) are useful in differentiating RPC from polymyositis. In polymyositis, the CPK is markedly elevated and muscle changes are noted.

**Treatment**

Corticosteroids are the primary treatment for RPC. Non-steroidal anti-inflammatory medications are also used to help manage pain.

Untreated, relapsing polychondritis has a high mortality rate. Mortality is usually due to respiratory problems such as airway obstruction, pneumonia, aortic valve disease, or aortic aneurysm.

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