DOWNLOAD PDF INFLAMMATORY MYOPATHY

Chapter 1 : Inflammatory Myopathies Information Page | National Institute of Neurological Disorders and S

What are the inflammatory myopathies? The inflammatory myopathies are a group of diseases that involve chronic (long-standing) muscle inflammation, muscle weakness, and, in some cases, muscle pain. Myopathy is a general medical term used to describe a number of conditions affecting the muscles. All.

Publications Definition The inflammatory myopathies are a group of diseases, with no known cause, that involve chronic muscle inflammation accompanied by muscle weakness. The four main types of chronic, or long-term, inflammatory myopathies are polymyositis, dermatomyositis, inclusion body myositis, and necrotizing autoimmune myopathy. These rare disorders may affect both adults and children, although dermatomyositis is more common in children. Polymyositis and dermatomyositis are more common in women than in men. Inclusion body myositis is most common after age General symptoms of chronic inflammatory myopathy include slow but progressive muscle weakness that starts in the proximal musclesâ€”those muscles closest to the trunk of the body. Other symptoms include fatigue after walking or standing, tripping or falling, and difficulty swallowing or breathing. Some individuals may have slight muscle pain or muscles that are tender to the touch. Polymyositis affects skeletal muscles involved with making movement on both sides of the body. Dermatomyositis is characterized by a skin rash that precedes or accompanies progressive muscle weakness. IBM is characterized by progressive muscle weakness and wasting. Juvenile myositis has some similarities to adult dermatomyositis and polymyositis. Injections of adrenocorticotropic hormone gel may be another option for people who do not respond to or cannot tolerate other drug treatment options. Physical therapy is usually recommended to prevent muscle atrophy as well as to maintain muscle strength and range of motion. Some individuals may use a topical cream to treat skin problems associated with the disorder. IBM has no standard course of treatment. View Full Treatment Information Definition The inflammatory myopathies are a group of diseases, with no known cause, that involve chronic muscle inflammation accompanied by muscle weakness. Prognosis Most cases of dermatomyositis respond to therapy. Approximately one-third of individuals with juvenile-onset dermatomyositis recover from their illness, one-third have a relapsing-remitting course of disease, and the other third have a more chronic course of illness. The prognosis for polymyositis varies. IBM is generally resistant to all therapies and its rate of progression appears to be unaffected by currently available treatments. Necrotizing autoimmune myopathy generally responds well to long-term combination immunosuppressive therapies.

Chapter 2 : Diseases and Conditions Inflammatory Myopathies

Myopathy is the medical term for muscle disease. Some muscle diseases occur when the body's immune system attacks muscles. The result is misdirected inflammation, hence the name inflammatory myopathies.

Myositis, or general muscle inflammation, may be caused by: The inflammatory myopathies are rare and can affect both adults and children. Dermatomyositis is the most common chronic form in children. Polymyositis and dermatomyositis are more common in females while inclusion body myositis affects more men. Inclusion body myositis usually affects individuals over age General symptoms of chronic inflammatory myopathy include slow but progressive muscle weakness. Inflammation damages the muscle fibers, which causes weakness, and may affect the arteries and blood vessels that pass through muscle. Other symptoms include fatigue after walking or standing, frequent episodes of tripping or falling, and difficulty swallowing or breathing. Some individuals may have muscle pain or muscles that are tender to touch. Polymyositis affects skeletal muscles the type involved in body movement on both sides of the body. It is rarely seen in persons younger than age Generally, the onset occurs between age 30 and Signs and symptoms of polymyositis vary considerably from person to person, which can make it difficult to diagnose. Untreated progressive muscle weakness may lead to difficulty swallowing, speaking, rising from a sitting position, climbing stairs, lifting objects, or reaching overhead. Some people with polymyositis may also develop arthritis, shortness of breath, heart arrhythmias irregular heartbeats , or congestive heart failure when the heart is no longer able to pump out enough oxygen-rich blood. Dermatomyositis is characterized by a skin rash that precedes or accompanies progressive muscle weakness. The rash appears patchy, with purple or red discolorations, and characteristically develops on the eyelids and on muscles used to extend or straighten joints, including knuckles, elbows, knees, and toes. Red rashes may also occur on the face, neck, shoulders, upper chest, back, and other locations. There may be swelling in the affected areas. The rash sometimes occurs without obvious muscle involvement and often becomes more evident with sun exposure. Adults with dermatomyositis may experience weight loss or a low-grade fever, have inflamed lungs, and be sensitive to light. Adult dermatomyositis, unlike polymyositis, may accompany tumors of the breast, lung, female genitalia, or bowel. Children and adults with dermatomyositis may develop calcium deposits, which appear as hard bumps under the skin or in the muscle called calcinosis. Calcinosis most often occurs one to three years after disease onset but may occur many years later. These deposits are seen more often in childhood dermatomyositis than in dermatomyositis that begins in adulthood. In some cases of polymyositis and dermatomyositis, distal muscles, which are the muscles away from the center of the body, such as those in the forearms and around the ankles and wrists , may be affected as the disease progresses. Polymyositis and dermatomyositis may be associated with collagen-vascular or autoimmune diseases such as lupus. Inclusion body myositis IBM is the most common form of inflammatory myopathy in people age 50 years and older and is characterized by slow, progressive muscle weakness and wasting over the course of months or years. IBM affects both proximal and distal muscles, typically in the thighs and forearms, and is often occurs on both sides of the body, although muscle weakness may affect only one side of the body. Falling and tripping are usually the first noticeable symptoms. The disorder often begins with weakness in the wrists and fingers that causes difficulty with pinching, buttoning, and gripping objects. People may experience weakness in their wrist and finger muscles and atrophy thinning or loss of muscle bulk in their forearm muscles and quadriceps muscles in the thighs. Difficulty swallowing occurs in approximately half of IBM cases due to involvement of the throat muscles. Symptoms of the disease usually begin after the age of 50, although the disease can occur earlier. Unlike polymyositis and dermatomyositis, IBM occurs more frequently in men than in women. Necrotizing autoimmune myopathy NAM is a rare and relatively newly recognized subgroup of inflammatory myopathies. NAM can occur at any age but usually affects adults. Its symptoms are similar to polymyositis and dermatomyositis, with weakness in both the upper and lower body, difficulty rising from low chairs, climbing stairs, or lifting objects. However, the onset of these symptoms can be more severe and sudden, reaching their peak over a period of days or weeks. Other symptoms include fatigue, weight loss, and muscle pain. NAM

occurs alone or after viral infections, in association with cancer, in people with connective-tissue disorders such as scleroderma, or, rarely, in people taking cholesterol lowering medications statins. Muscle weakness and pain may continue to worsen even after individuals stop taking the drugs. Childhood inflammatory myopathies have some similarities to adult dermatomyositis and polymyositis. They typically affect children ages 2 to 15 years. Symptoms include proximal muscle weakness and inflammation, edema an abnormal collection of fluids within body tissues that causes swelling , muscle pain, fatigue, skin rashes, abdominal pain, fever and contractures. Contractures result from shortening of muscles or tendons around joints, are caused by inflammation in the muscle tendons, and prevent the joints from moving freely. Children with inflammatory myopathies may have difficulty swallowing and breathing. The heart may also be affected. Between 20 to 40 percent of children with juvenile dermatomyositis develop calcinosis, which can cause significant muscle weakness and pain, joint contracture, skin ulcers, and decreased muscle bulk. Diagnosis is based on medical history, results of a physical examination that includes tests of muscle strength, and blood samples that show elevated levels of various muscle enzymes and autoantibodies. A biopsy sample of muscle tissue should be examined for signs of chronic inflammation, muscle fiber death, vascular deformities, or other changes specific to the diagnosis of a particular type of inflammatory myopathy. A skin biopsy can show changes in the skin associated with dermatomyositis. Chronic inflammatory myopathies cannot be cured in most adults but many of the symptoms can be treated.

Chapter 3 : Inflammatory Myopathy

Inflammatory myopathy (inflammatory muscle disease or myositis) is disease featuring weakness and inflammation of muscles and (in some types) muscle calendrierdelascience.com cause of much inflammatory myopathy is unknown (), and such cases are classified according to their symptoms and signs and electromyography, MRI and laboratory findings.

Loss of coordination and balance [1] Lack of fine and gross motor control [1] Cause[edit] Acquired noninflammatory myopathy can be caused by a variety of factors including metabolic abnormalities, drugs, nutritional deficiency, trauma, and upstream abnormalities resulting in decreased function. Two of the most common causes of ANIM are hyperthyroidism and excessive steroid use, while many drugs used to treat rheumatism are known to be inducing agents. Most cases of ANIM can be linked to drugs or dietary abnormalities. Drug induced myopathy[edit] It is not uncommon for drugs to damage muscle fibers. Particular families of drugs are known to induce myopathies on the molecular level, thus altering organelle function such as the mitochondria. Use of multiple drugs from these families in conjunction with one another can increase the risk of developing a myopathy. Statins[edit] Prescribed statins for dyslipidemia are associated with muscle toxicity. Symptoms of this muscle toxicity include combinations of cramping, weakness, aching or tenderness; and are often experienced in the quadriceps, pectoral, biceps, low back, or abdominal region. Symptoms tend to worsen with muscle exercise, and often continue after a patient is removed from statin therapy. Statins induce myopathy by inhibiting protein synthesis within the muscle. Often, the damage is found within the mitochondria. Corticosteroids[edit] Corticosteroids often cause muscle weakness to some degree in patients. Symptoms are usually weakness of the proximal muscles, neck flexor, and in extreme cases, respiratory muscle weakness can also occur. These side effects are more common in women than in men, for reasons that are unknown. A muscle biopsy shows a vacuolar myopathy without significant cell death or inflammation. Muscle enzymes are increased, commonly lactate dehydrogenase LDH. This drug is known to have toxic effects on myofibrils, resulting in muscle pain and tenderness. Biopsies demonstrate more molecular damage and dysfunction within the mitochondria, higher lipid storage, and vibrant red myofibrils. These symptoms are commonly found in the proximal muscles of the body. Chemical imbalances brought on by abnormal diets may either affect the muscle directly or induce abnormal functionality in upstream pathways. Hyperthyroidism is one of the most common ways to acquire ANIM. The muscles exhibit a pathology similar to an overdose of epinephrine commonly known as adrenaline. Patients with hyperthyroidism show weakness of shoulder girdle muscles in particular with this condition often being asymptomatic. More serious weakness of core and limb muscles may present. Vitamin D induced ANIM is most commonly associated with sleep deprivation as it induces tonsillar and adenotonsillar hypertrophy, as well as weakens the airway muscles. Vitamin D induced ANM can also be associated with daytime impairment through this pathway. This is due to muscular contusions and partial or complete loss of function for affected muscle groups. Basic exams will test for where the muscle weakness is and how weak it is. This is performed by testing for proximal and distal muscle strength, as well as testing for any signs of neurogenic symptoms such as impaired sensation, deep tendon reflexes, and atrophy. Measurement of serum levels of muscle enzymes [1].

Chapter 4 : Inflammatory Myopathies | ARUPConsult

Inflammatory myopathies are acquired, treatable autoimmune diseases, characterized clinically by weakness and soreness of muscles and elevated CK and pathologically by myonecrosis and mononuclear inflammatory infiltrates.

The pathology of dermatomyositis includes inflammation, vasculitis, and perifascicular atrophy. The inflammatory cells are predominantly B-cells with smaller numbers of CD4-positive T-cells and are found around blood vessels, in the septa between muscle fascicles, and in fibroadipose tissue around muscle. The key pathological change of dermatomyositis is a vasculitis, which involves endomysial and perimysial capillaries and arterioles. This vasculitis begins with endothelial swelling and is followed by endothelial necrosis and capillary loss. Tubuloreticular cytoplasmic inclusions TRIs are often seen in endothelial cells. TRIs also occur in lupus and other collagen vascular diseases but are absent in polymyositis and inclusion body myositis. The vasculitis is thought to be caused by circulating anti-endothelial antibodies. Interaction of these antibodies with vascular antigens activates complement, leading to formation of the membranolytic attack complex MAC , which destroys endothelial cells. Perifascicular atrophy Perifascicular atrophy Dermatomyositis. A distinctive feature of dermatomyositis is atrophy and degeneration of myofibers at the periphery of fascicles perifascicular atrophy-PFA , which occurs even in absence of inflammation. It has been proposed that PFA is caused by ischemia from loss of the endomysial capillary bed. This affects more severely the distal portion of the vascular field, which is the periphery of each fascicle. In support of this hypothesis, ischemic infarction of muscle is seen in some cases of dermatomyositis. However, PFA is not seen in diabetic and other angiopathies. Recent evidence indicates that the CD4-positive cells in dermatomyositis are plasmacytoid dendritic cells. These cells secrete type 1 interferons, which induce genes and molecular cascades that cause muscle injury. A key interferon-induced pathology is deficiency of titin, an intramuscular protein that is a scaffold for contractile filaments. It is now thought that diffusion of interferons in the perifascicular areas causes the PFA. Dermatomyositis and polymyositis and less frequently inclusion body myositis are associated with scleroderma, mixed connective tissue disease, and cancer. The association with cancer is stronger with dermatomyositis. Some of these extra-muscular manifestations are associated with circulating antibodies to anti-Jo-1 an anti tRNA synthetase autoantibodies. Mixed CTD Mixed connective tissue disease. Inflammaton in muscle tissue and in the connective tissue septa between muscle fascicles. The muscle biopsy in systemic lupus erythematosus and other connective tissue diseases CTD shows most often interstitial perivascular mononuclear infiltrates without vascular injury or myonecrosis. In some patients, however, manifestations of CTD overlap with inflammatory myopathy. Patients with mixed CTD have muscle pathology that closely resembles dermatomyositis. The inflammation of polymyositis, dermatomyositis, and collagen vascular disease subsides rapidly with corticosteroids, so the biopsy should be done before treatment is started. Inclusion body myositis IBM. Vacuoles rimmed with basophilic granules. Vacuole filled with granules. Modified Gomori trichrome stain. Myelinoid bodies and abnormal filamentous inclusions Sporadic inclusion-body myositis s-IBM is the most common muscle disease in old people. It has an insidious onset and causes slowly progressive proximal and distal weakness with mild CK elevation. The pathology of s-IBM is highly characteristic and combines inflammation and myofiber degeneration. The inflammation is similar to polymyositis with cytotoxic CD8-positive lymphocytes invading and destroying myofibers. The degenerative changes consist of accumulation of myelinoid bodies and amyloid deposition. Affected myofibers have vacuoles or cracks, which contain basophilic granules. These are best seen in cryostat sections stained with modified Gomori trichrome. The granules consist of myelinoid membranous bodies. In or near the vacuoles there are small chunks of amyloid that can be detected with Congo Red stains. The pathogenesis of IBM is not known but probably involves abnormal protein processing associated with ageing of myofibers, and the deposition of toxic protein polymers that damage myofibers and trigger inflammation. S-IBM is refractory to immunosuppressive therapy that is used in other inflammatory myopathies. There are hereditary myopathies with similar myofiber changes but without inflammation. The pathogenesis of s-IBM is not known but probably involves abnormal protein processing associated with ageing of myofibers, and the deposition of

toxic protein polymers that damage myofibers and trigger inflammation. PM and DM are treated with corticosteroids. Azathioprine , methotrexate, and cyclosporin are used in severe cases. IBM is refractory to corticosteroids and immunosuppressive agents. Weakness affects most severely muscles that are innervated by brain stem nuclei, such as extraocular and facial muscles, and causes drooping of the eyelids, diplopia, and inability to chew. Death is due to respiratory compromise. Pathologically, the muscle is either normal or shows myofiber atrophy and aggregates of lymphocytes in the endomysium. In severe cases, there may be myonecrosis. Electron microscopy shows abnormal motor end plates. Axon terminals are normal and the number of synaptic vesicles is adequate, but there is loss of post-synaptic membrane such that the post-synaptic region is simplified, showing a few wide folds without branching. The primary synaptic cleft is widened. Ten percent of patients with myasthenia gravis, especially older males, have thymomas, and most other patients have follicular hyperplasia of the thymus. Myasthenia gravis is caused by antibodies to the acetylcholine receptor AChR protein. These antibodies also cause degradation of AChR and lysis of post-synaptic membranes. Improvement of strength following administration of edrophonium Tensilon test or neostigmine are diagnostic. Treatment consists of anticholinesterase drugs, corticosteroids and thymectomy. Further Reading Mammen AL. Clinical presentation, autoantibodies, and pathogenesis. Ann NY Acad Sci ; Pathogenic considerations in sporadic inclusion-body myositis, a degenerative muscle disease associated with aging and abnormalities of myoproteostasis. J Neuropathol Exp Neurol.

Chapter 5 : Inflammatory Myopathies Fact Sheet | National Institute of Neurological Disorders and Stroke

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Chapter 6 : Acquired non-inflammatory myopathy - Wikipedia

Inflammatory myopathies are muscle diseases characterized by muscle weakness. The three major inflammatory myopathies are polymyositis, dermatomyositis, and inclusion body myositis. Each type has different findings: One of the major effects of the inflammatory myopathies is difficulty in swallowing.

Chapter 7 : Inflammatory myopathies

Inflammatory myopathies: Association with neoplasms Frequency may depend on ethnic background Risk of malignant disease Temporal Highest: Near time of myositis diagnosis (1 to 3 years).

Chapter 8 : Inflammatory myopathy - Wikipedia

Idiopathic inflammatory myopathy refers to a group of conditions that affect the skeletal muscles (muscles used for movement). Although the condition can be diagnosed at any age, idiopathic inflammatory myopathy most commonly occurs in adults between ages 40 and 60 years or in children between ages 5 and 15 years.

Chapter 9 : Rheum2Learn Inflammatory Myopathies

Edit concept Create issue ticket Myositis Inflammatory Myopathy. Myositis is a term used to describe inflammation of the muscles. These conditions represent the largest group of acquired and potentially treatable causes of skeletal muscle weakness.