

This article was commissioned by the Canadian Spondylitis Association in the interest of informing consumers about the Extra-articular Manifestations of Ankylosing Spondylitis. It was written by Dr. Dinny Wallis, MBChB. Dr. Wallis is a Rheumatology Specialty Trainee, Wessex Deanery, UK and was an AS Fellow, University of Toronto, 2012-2013. Dr. Wallis has received Fellowship funding from Janssen and an honorarium for writing this article from the Canadian Spondylitis Association. The honorarium was funded by an educational grant from Janssen.

Extra-articular manifestations (EAMs) of Ankylosing Spondylitis

INTRODUCTION

Ankylosing spondylitis (AS) is a chronic disease characterized by inflammation of the joints of the spine. It is one of a group of diseases called spondyloarthropathies which share clinical, genetic and radiological features. The term “extra-articular manifestations” refers to involvement of systems of the body other than joints, for example, the eyes, the gut and the skin. The presence of extra-articular manifestations may have important implications for treatment choices in AS.

Uveitis (Iritis)

Approximately 30-40% of people with AS will develop anterior uveitis (also called iritis). This is the term used to describe inflammation of the middle layer of the eye (the uveal tract).

Symptoms

Common symptoms of uveitis are sensitivity to light (photophobia), pain and / or redness of the eye and blurred vision. Usually one eye is affected at a time, but it is common for uveitis to recur in the other eye. Symptoms may develop over one to two days.

Treatment

Uveitis can be serious, leading to permanent damage to the eye and visual loss and should be managed as an emergency (ideally within 24 hours of onset) by an ophthalmologist. Uveitis usually responds to steroid drops along with dilating drops to

control pain from ciliary muscle spasm. If inflammation is persistent, steroids can be given by mouth or by injection into the eye. Immunosuppressant drugs may also be used.

Inflammatory bowel disease

Around 10% of people with AS will develop inflammatory bowel disease (IBD). The two most common forms of IBD are Crohn's disease and ulcerative colitis (UC). In both conditions the intestine or bowel becomes inflamed. Crohn's disease can affect any part of the gut from mouth to anus while ulcerative colitis affects the large intestine (colon) and rectum. Inflammatory bowel disease is not the same as irritable bowel syndrome (IBS).

Symptoms

IBD may cause abdominal pain, mouth ulcers, fatigue, diarrhoea (sometimes with blood) and weight loss.

Investigations

If a diagnosis of IBD is suspected, the patient will be referred to a gastroenterologist. Blood tests, stool tests, x-rays and endoscopy (examination of the inside of the gut) can all be used to diagnose and monitor IBD.

Treatment

There are a variety of treatment options for IBD. These include 5-ASA preparations (e.g. mesalazine), which dampen down inflammation in the bowel, glucocorticoids (steroids), immunosuppressants (e.g. azathioprine, 6-mercaptopurine, methotrexate) and biologic drugs (e.g. infliximab). Medication may be given by mouth, topically (e.g. enemas or suppositories) or by injection. Anti-inflammatories, which are the mainstay of management for AS, commonly cause gastrointestinal side effects and may exacerbate symptoms of IBD. Ideally they should be avoided in patients with IBD, but it may be decided to use them with care in selected patients with close monitoring. Biologics can

be effective in both AS and IBD. Rheumatologists and gastroenterologists will often work closely together to determine the best treatment approach.

Outlook

The prognosis for IBD is variable. It is a chronic (long term) condition. Some people may have intermittent flares with long periods of remission. Persistent disease or damage to the gut from inflammation may require bowel surgery.

Psoriasis

Around 10-20% of patients with AS have psoriasis. Psoriasis typically causes a red, raised rash (plaques) with silvery scale and is due to a high rate of turnover of skin cells where cells are formed and shed more quickly than normal.

Symptoms

Plaques typically occur on the knees and elbows, but psoriasis can also affect the scalp, body folds (armpit and groin), palms and soles (palmo-plantar pustulosis). Some plaques arise where the skin has been damaged e.g. by a cut or scratch. The nails are commonly affected with pitting, discoloration or thickening.

Treatment

A number of topical therapies (applied to the skin) can be helpful including emollients, steroids, tar preparations and vitamin D analogues. If psoriasis does not respond to topical therapies, drugs may be given in tablet or injectable form. Light therapy (phototherapy) is also used. Biologic drugs can be effective for both AS and psoriasis.

Enthesitis

Enthesitis is inflammation at the points where tendons and ligaments attach to bone (entheses). It can affect a number of areas, most commonly the back of the heel at the

Achilles tendon, under the heel (the plantar fascia), around the pelvic bones and in the chest wall.

Symptoms

Enthesitis causes pain and tenderness in a localised area.

Investigations

A diagnosis of enthesitis is usually based on the patient's symptoms and examination findings. Sometimes ultrasound or MRI may be helpful.

Treatment

Enthesitis may respond to exercises recommended by a physiotherapist, anti-inflammatory taken by mouth or applied to the skin in gel form, or steroid injection. In the case of enthesitis affecting the foot, supportive footwear and advice from a podiatrist may be helpful.

Osteoporosis

AS is known to be associated with osteoporosis. Osteoporosis literally means "porous bones". Thinning of the bones results in loss of bone strength and a higher risk of fractures. In AS, there is a particular risk of vertebral (spinal) fractures. When a vertebral body fractures it loses height and may be described as "wedged", "crushed", or "compressed."

Symptoms

Osteoporosis does not cause bone pain or other symptoms unless there is a fracture. Vertebral fractures may not cause any symptoms and do not usually cause problems with the spinal cord or other neurological complications. Losing height may indicate the

presence of a vertebral fracture, but height loss can also reflect structural changes relating to AS.

Investigations

The risk of osteoporosis can be assessed based on a number of factors including age, family history of osteoporosis, occurrence of previous fractures, alcohol intake, smoking history and other medical problems or medications. If a patient is thought to be at high risk of osteoporosis, a bone density scan will be performed. The presence of structural changes in the spine due to AS can make a bone density scan of the spine difficult to interpret but reliable measurements can be obtained from the hip.

Treatment

The risk of osteoporosis can be reduced through measures such as taking regular weight-bearing exercise, avoiding smoking and excess alcohol, eating a diet rich in calcium and maintaining a healthy weight (not being too thin). Osteoporosis can be treated with drugs to improve bone strength and reduce the risk of fractures. The most commonly used drugs are bisphosphonates (e.g. alendronate, risedronate) and can be given by mouth or intravenously. Calcium and vitamin D supplements will usually be prescribed as well.

Fatigue

Fatigue can be a major problem in AS. It is usually due to a combination of different factors including pain, loss of sleep, active inflammation and anaemia. Fatigue can make pain more difficult to manage.

Managing fatigue

Your doctor may wish to arrange blood tests to rule out other causes of fatigue such as thyroid disease or anaemia. Following a carefully planned exercise and rest routine, pacing activities, eating a balanced diet and getting into a good bedtime routine (sleep

hygiene) are important. Input from physiotherapy and/or occupational therapy can be useful.

Heart involvement

A minority of patients with AS will develop involvement of the aortic valve, which occurs as a result of inflammation of the aorta (the main blood vessel arising from the heart). Irregular heart rhythms (conduction disturbances) may also occur. These are rare problems. All inflammatory rheumatic diseases, including AS, appear to be associated with an increased risk of atherosclerotic heart disease.

Lung involvement

Inflammation and scarring of the joints where the ribs meet the spine or where the ribs meet the breastbone can lead to reduced expansion of the chest causing pain or restriction of deep breathing. This can be exacerbated by increasing curvature of the spine. Advice from a physiotherapist about deep breathing exercises may be helpful. In the past, AS was thought to be associated with scarring (fibrosis) of the upper part of the lungs, but this is not seen frequently.

Biologic therapy for extra-articular manifestations

Biologic drugs are synthetic molecules which block the action of various proteins thought to drive inflammation in AS. A number of different biologic drugs are available. They are given by injection and can be very effective in controlling active AS and enthesitis. They are generally reserved for use in patients who do not respond to traditional management with anti-inflammatories. There is some variability in the efficacy of different biologic drugs in treating extra-articular manifestations of AS but they are commonly used in treating severe psoriasis, IBD and uveitis. The effect of biologic drugs on osteoporosis is not completely understood but it is thought that with suppression of inflammation, bone density may improve. The choice of biologic drug in AS will be influenced by the presence of extra-articular manifestations.

SUMMARY

Extra-articular manifestations are common in AS. A multidisciplinary approach to management should be considered, including input from physiotherapy, occupational therapy and other medical specialties.

Further reading

Osteoporosis Canada www.osteoporosis.ca

Crohn's and Colitis Foundation of Canada www.cafc.ca

Canadian Dermatology Association www.dermatology.ca

