

## Gastrointestinal and liver complications of Sjögren's Syndrome

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At least one third of patients with Sjögren's Syndrome will have abdominal symptoms at some point in the course of their condition (Davidson et al, 1999). This article discusses the potential effects of Sjögren's on the gastrointestinal (GI) system and liver.

### Dysphagia and oral candida

There are many potential causes of swallowing difficulties (dysphagia) in Sjögren's Syndrome. The oral dryness can make chewing and swallowing difficult and these problems are exacerbated if dental disease has led to loss of some or all of the natural teeth. The saliva not only reduces in volume but changes in quality and many patients need to drink water to help swallowing. Several studies have shown changes in the muscular contractions within the oesophagus (food gullet) which may hamper swallowing and one study found evidence of an oesophageal web in 10% of patients with Sjögren's. These webs consist of an elastic membrane of oesophageal mucosa -the tissue normally lining the gullet. Oral candida (thrush) is a common problem and may affect up to 75% of patients with Sjögren's at some point in their illness & can contribute to oral symptoms. The management of candida has been comprehensively discussed in a recent newsletter article (John Hamburger & Stephen Porter, Summer 2006, Vol 20, issue 2).

### Gastritis

Dyspepsia (heartburn and indigestion) is a common symptom in the normal population. There is a known association in the general population between the presence of infection with bacteria known as helicobacter pylori and the presence of indigestion and ulcers. Helicobacter pylori infection rates are similar in patients with Sjögren's Syndrome and the normal population (57% v 62%) but whereas eradication of the bacterial infection cured the symptoms in the normal patients it did not have a similar effect in the Sjögren's patients (Sorrentino et al.).

### Irritable bowel syndrome

Irritable Bowel syndrome is a poorly understood condition characterised by the presence of bloating, intermittent diarrhoea and abdominal colic, which may be relieved by a bowel action. It is thought to affect up to 20% of the general population at some time and is slightly commoner in women. There is no diagnostic test -the diagnosis is often made on the presence of classical symptoms and absence of other causes. In many patients the symptoms are chronic. There are no studies looking at the prevalence of irritable bowel syndrome in patients with Sjögren's Syndrome but anecdotal reports suggest it is common. Dietary change including increasing the fibre content of the diet may help.

### Coeliac disease

Coeliac disease is an intolerance to gluten (a wheat protein) and leads to malabsorption. Common symptoms include weight loss

and anaemia and simple screening blood tests are usually helpful in confirming a diagnosis. Evidence of coeliac disease was found in 4.5% of patients with SS in one Hungarian study (Szodary et al, 2004). This compares with a prevalence of 4.5 -5.5 per 1000 in the normal European population. In another study antibodies to tissue transglutaminase (TIG), an antibody strongly associated with coeliac disease, were present in 12% of patients with SS compared to 4% of normal controls. On further investigation over 70% of the anti-TIG positive patients were found to have biopsy evidence of coeliac disease (Luft et al 2003). Overall therefore coeliac disease is ten times commoner in patients with Sjögren's Syndrome than in the normal population. Treatment is with a gluten free diet.

### Pancreatitis

The pancreas is a gland producing insulin and a variety of enzymes important in the digestion of fats & proteins within the bowel. Subtle inflammation of the pancreas is common in patients with Sjögren's Syndrome but only rarely causes clinical concern. Studies have shown mild elevation of pancreatic enzymes in about 35% of patients with Sjögren's (Ostuni et al 1996) but more serious problems occur in no more than 1% of patients.

### Autonomic nervous system dysfunction

The autonomic nervous system controls bodily functions not under conscious control e.g. heart rate, gastric emptying etc. Autonomic nervous system dysfunction is seen in a proportion of patients with Sjögren's Syndrome and can be a factor in bowel disease e.g. gastric emptying is slowed in approximately 70% of symptomatic patients with Sjögren's Syndrome (Kovacs et al, 2003).

### Liver

Mild elevation of liver enzymes are seen in up to a quarter of patients with Sjögren's Syndrome but most of these patients are asymptomatic and more serious disease is rare. In one large study of 300 patients with primary Sjögren's Syndrome some signs of liver involvement were found in 7% of patients but the majority of these were asymptomatic (Skopouli et al, 1994). The commonest associated liver condition is primary biliary cirrhosis (see below). Some patients have mild liver inflammation visible on biopsy which is felt to be a manifestation of the Sjögren's itself but other forms of liver disease are rare and probably associated by chance alone

### Primary biliary cirrhosis

Primary Biliary cirrhosis (PBC) is a slowly progressive, inflammatory condition of the liver. There are many similarities between Sjögren's and Primary biliary cirrhosis - both affect primarily women and are thought to be autoimmune. They both cause a similar pattern of inflammation on biopsy. In studies Sjögren's Syndrome is found in about 17% of patients with

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PBC (Parikh et al, 2001) and conversely PBC has been found in about 6% of patients with Sjogren's. It is probable that there is a true 'overlap' syndrome where patients develop both conditions alongside each other. Treatment is usually with ursodeoxycholic acid (UDCA) which can slow the progress of the liver condition and improve the long-term outlook.

### **Sclerosing cholangitis**

Sclerosing Cholangitis is a relatively rare liver condition usually found in association with ulcerative colitis and affecting predominantly men but there have been a few case reports of it occurring in patients with Sjogren's - these are probably chance occurrences and there is no indication that the conditions are linked. Diagnosis and treatment usually requires a specialist referral.

### **Hepatitis C**

Hepatitis C can cause a chronic viral infection and consequent liver damage. It is not common in the UK and can be passed from mother to child, acquired from contaminated blood or via sexual intercourse with a carrier. All blood for transfusion in the UK is now screened for Hepatitis C. Some patients with Hepatitis C develop a Sjogren's like illness with associated liver involvement. Treatment is usually in a specialist centre.

### **Lymphoma**

Lymphoma, usually of the MALT type, occurs in about 1% of patients with Sjogren's Syndrome and very occasionally involves the GI tract. The presentation is usually with chronic diarrhoea, malabsorption and weight loss. Fortunately in most cases lymphoma responds to treatment with chemotherapy.

### **Summary**

In conclusion gastrointestinal involvement is common in patients with Sjogren's Syndrome but in the majority of cases is mild.

### **References**

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