British Sjögren's Syndrome Association

Originally published in the Summer 2014, Volume 29, Issue 2 of Sjögren's Today magazine

Respiratory involvement in Sjögren's Syndrome

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Although dry eyes and mouth are the classical symptoms of Sjögren's Syndrome approximately 70% of patients will have 'extra-glandular' symptoms at some point in the course of their condition. Involvement of other organs is commonest in those patients with the characteristic anti-Ro/La antibodies. Raynauds (poor circulation in the hands and feet) and thyroid disease are the commonest 'extra-glandular' manifestations but lung involvement is seen in a proportion of people.

Significant lung disease is rare but many patients complain of a chronic cough. Cough is a nonspecific reaction to irritation anywhere from the pharynx to the lungs and is a common symptom in the normal population, being reported by 10-20% of adults. The cough reflex may be triggered by mechanical or inflammatory changes or irritants in the airways. Chronic cough tends to be inhibited during sleep. Common causes of chronic cough include smoking, postnasal drip, asthma, and gastro-oesophageal reflux. Certain drugs (in particular angiotensin-converting enzyme inhibitors) can also precipitate cough. It is always worth considering and excluding these causes first but in most patients with Sjögren's Syndrome the cough is related to drying of the mucous membranes. A combination of reduced quantity and increased viscosity (stickiness) of the mucus secretions which normally line the airways is probably to blame for the persistent dry cough experienced by many patients with Sjögren's Syndrome. In terms of treatment simple remedies include ensuring you remain as hydrated as possible and using a humidifier to reduce dryness in the air. There is some evidence that treatment with oral pilocarpine (a drug which non-specifically stimulates secretions by stimulating muscarinic receptos on the surface of secretroy cells) can help the chronic cough.

More serious lung disease is unusual. One study found Chest X-Ray changes in 14% of patients with Sjögren's Syndrome. These most commonly consisted of fine reticular changes (seen as fine lines on the X-Ray) affecting the lower lobes of the lung and accompanied by fine crackles in the lung bases on clinical examination. Subtle changes on High resolution CT (HRCT) scanning (a very sensitive type of X-Ray of the chest) were seen in up to 30% (most commonly parenchymal linear opacities and bronchiolar abnormalities) but only 26% of patients with changes

on scanning had symptoms. The majority of these symptoms were mild with very little correlation between symptoms, clinical signs and radiological findings. Other studies confirm that pulmonary involvement is often sub-clinical with abnormal findings on very detailed testing in up to 75% but again poor correlation between the finding of subtle abnormality on intensive testing and actual symptoms in day to day life.

Very occasionally patients present acutely with lymphocytic interstitial pneumonitis (LIP). This is a condition sometimes seen in patients with Sjögren's, Lupus or other connective tissue diseases. The patient usually presents with what is initially felt to be a chest infection, with shortness of breath, a dry cough and sometimes a high temperature. However they don't respond as expected to treatment with antibiotics. On examination they will have crackles in the lung bases and on X-Ray non-specific shadowing is often noted. Characteristic changes of bibasilar pulmonary infiltrates may be seen on HRCT scanning. Bronchoscopy (where the lungs are examined

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directly with a fibre-optic tube) is sometime carried out to confirm the diagnosis and confirms that the infiltrates consist of dense accumulations of inflammatory white cells (lymphocytes and plasma cells). LIP usually responds well to treatment with steroids and usually settles promptly. As with most systemic complications in Sjögren's LIP is commoner in those patients with Ro and/or La autoantibodies.

A very rare cause of shortness of breath is right sided heart failure due to pulmonary artery hypertension. This is where there is back pressure on the heart from lung disease and it is known as Pulmonary Artery Hypertension or PAH for short. It is sometimes seen in other rheumatic diseases such as scleroderma and very occasionally in lupus but is relatively rare in Sjögren's Syndrome. There are a few case reports in the medical literature and there have been a couple of studies which suggest that mild (and usually asymptomatic) disease may occur in up to 15% but that more severe disease is very rare. One study looked at 107 patients with Sjögren's Syndrome (104 females, 3 males, average aged 56 years and performed echocardiograms (detailed heart scans) in all. They found evidence of PAH in sixteen (15%) of this group of patients. It was mild in 13 and moderate in 3. They found that PAH was more likely to occur in patients with Ro/La antibodies and low complement levels. PAH may be treated by cardiologists with various drugs to try and relieve the pressure on the heart but it may also be worth treating the underlying disease (in this case Sjögren's) which has led to the development of the problem in the first place.

In conclusion therefore a chronic dry cough is the most common lung manifestation of Sjögren's Syndrome. Other lung disease is rare.