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Medi Quest BRS Hospital

A monthly News letter from BRS Hospital

PANNICULITIS – A BIG RED HERRING

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A few weeks ago, a middle aged lady had come to the emergency department with complaints of severe abdominal pain that was intermittent in nature with a colicky character. The only other associated symptom was a low grade fever. There were no aggravating or relieving factors to the pain and there was no vomting, diarrhea or burning micturition. On clinical examination, there was a diffuse abdominal tenderness but no focal tenderness.

The patient was a known case of Sjogren's disease who was on treatment but was poorly compliant to medication. In order to further evaluate the patient and identify the etiology, the patient was admitted to the ICU and started on pain killers.

Initial labs revealed a mild leucocytosis and elevated inflammatory markers (ESR, CRP). To rule out structural and surgical causes of abdominal pain, an abdominal CT with contrast was done. The CT showed mesenteric panniculitis of the abdominal fat pads. Patient was restarted on corticosteroids and responded very well. Pain reduced dramatically over night and fevers subsided. Over the course of her stay, the inflammatory markers also normalised.

The purpose of this article it understand what Sjögren's syndrome is, what its clinical features are and where mesenteric panniculitis fits into the spectrum of the clinical features of Sjögren's syndrome.

What is Sjögren's syndrome? What are it's clinical features?

Sjögren's syndrome was first described in patients with chronic arthritis. Swedish physician Henrik Sjogren noticed that there many women with chronic arthritis, who had associated dryness of the eyes and mouth. Because of this dryness this syndrome also came to be known as Sicca syndrome.

Today we know that Sjögren's syndrome is an autoinflammatory disorder which commonly affects the exocrine glands of the body. It is characterised by inflammation in the initial stages followed by fibrosis in the later stages of the illness. Sicca syndrome commonly involves the lacrimal and salivary glands and fibrosis of these glands can lead to hypofunctionality which presents as dryness of the eyes and mouth.

The most common presenting feature in patients with Sjögren's syndrome is dry eyes and dry mouth. Patients often present with complaints of burning of eyes, foreign body sensation or a gritty sensation in the eyes. They may also

present with difficulty in swallowing, increased thirst, and recurrent salivary stones.

The dryness is not limited to the eyes and the mouth, however. Essentially any mucocutaneous membrane can be prey to the dryness. Sjogren's can affect the skin, the tracheobronchial tree, and the vaginal mucosa and predispose all these areas to infection.

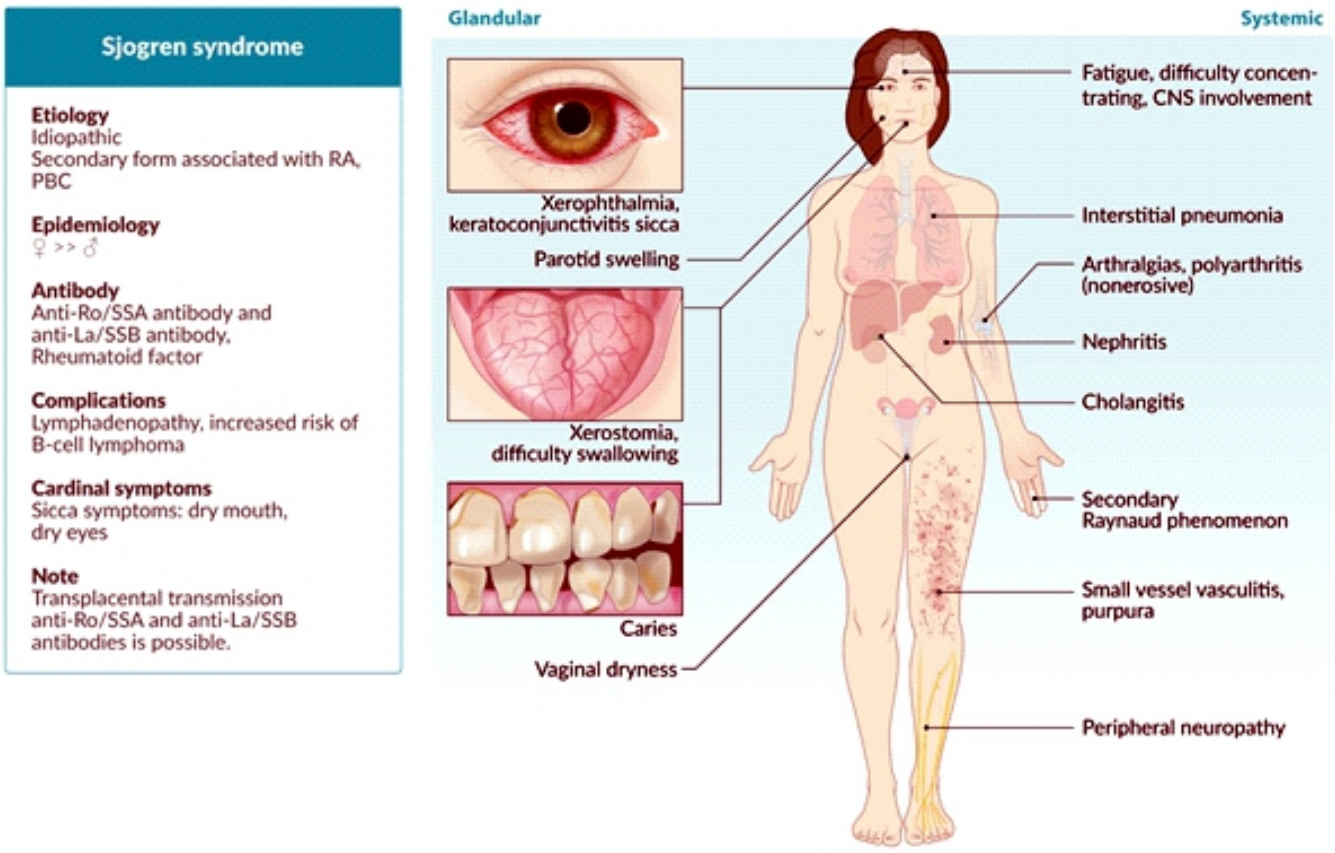
In addition to causing damage to the glands, about half of the patients with Sjogren's also present with extra glandular features. There is well documented involvement of the joints, kidneys, lungs, nervous system and the gastrointestinal tract. In rare cases, Sjogren's can also lead to development of hematological malignancies. Involvement of these systems can greatly increase the morbidity and mortality in Sjögren's syndrome.

In addition to primary Sjogren's, features of Sjogren's can also be seen along with features of SLE, RA and other autoimmune disorders. This is referred to as secondary Sjogren's or Sjogren's overlap syndromes.

What is Panniculitis?

Panniculitis is a heterogenous group of disorders which are characterized by inflammation of the subcutaneous fat. Because fat is not considered to be the first culprit in any pathology it can often go unnoticed or only found incidentally when evaluating for other possible causes or pain or fever.

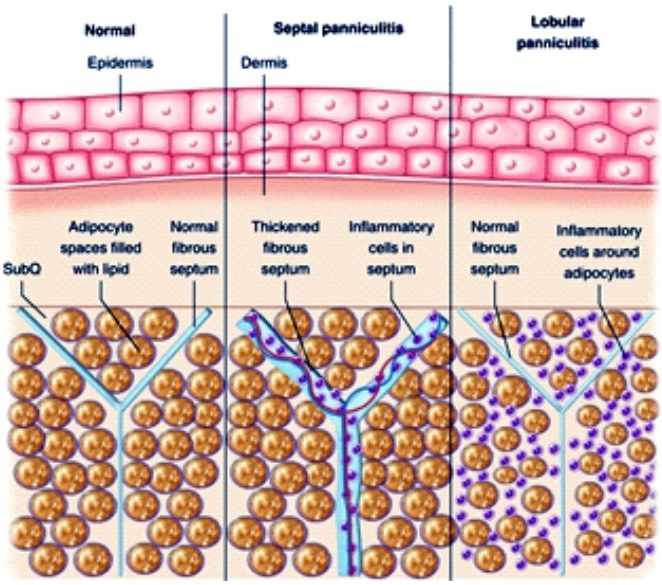
Broadly panniculitis can be classified as being septal or lobar based on the biopsy findings. This morphological classification is made based on the location of the inflammatory infiltrates. If the infiltrates are congregated around the septa then it is considered to be septal panniculitis. However, the infiltrates congregate within the substance of the fat between the adipocytes then it is considered to be lobular panniculitis.



Bear in mind that in many cases, the panniculitis may evolve from the septal morphology to a lobar morphology. A septa-lobular or mixed morphology can also be present complicating the diagnosis.

The other important histologic feature to look for in patients with panniculitis is the presence of vasculitis. Septal panniculitis with or without vasculitis and lobar panniculitis with or without vasculitis can have an important distinctions in differential diagnoses.

Mesenteric panniculitis is a subset of panniculitis. In which the fat pockets around the intestines housed in the mesenteric folds are inflamed. It can lead to a spectrum of clinical features ranging from non specific abdominal pain to intestinal obstruction.



Diagnosis of Sjögren's syndrome

Usually the diagnosis of Sjögren's syndrome is made on clinical features since most of the tests available for the same are coroboratory and not confirmatory.

A lacrimal gland or salivary gland biopsy is usually indicated. These biopsies show aggregates of lymphocytes which are CD4 predominant. The most common biopsy done is a minor salivary gland biopsy which is taken by making a small incision on the inner lip.

Looking at the tear film by assessing the tear film breakup time or Schirmer's test can help quantify ocular dryness and also help assess response to treatment.

Presence of anti Ro (Anti SS-A) or anti La (Anti SS-B) antibodies can help clinch the diagnosis especially in cases of overlap syndromes. They can also warn about possibilities of complications like pulmonary involvement.

Mesenteric panniculitis in patients with Sjögren's syndrome is a very rare occurrence and it is important to keep it in mind as a possibility since it can resemble a surgical abdomen. Usually a contrast CT of the abdomen can help identify the inflammation.

Treatment of Sjogren's and Panniculitis.

Most of the treatment of Sjogren's revolves around immunosuppression and symptomatic relief. Immunosuppression is started with anti malarials (hydroxychloroquine) and glucocorticoids. Long term treatment may require medications like methotrexate, azathioprine and cyclophosphamide.

Dry eye is treated with artificial tears. In severe cases punctual cauterisation is done. Cyclosporine eye drops can be used to increase tear production.

Non pharmacological treatment like chewing gum, and drinking water are used in initial stages of dry mouth. Cevimuline and pilocarpine can be used to increase salivary production.

Extraglandular involvement necessitate the use of corticosteroids in a tapering dose.

Mesenteric panniculitis is usually self limiting in patients with Sjogren's. The pain can subside with use of NSAIDs alone. But in recalcitrant conditions, it will respond well to corticosteroids therapy.

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