CORRESPONDENCE

The Diagnosis and Treatment of Sjögren's Syndrome

by Dr. med. Ana-Luisa Stefanski, Dr. med. Christian Tomiak, Prof. Dr. med. Uwe Pleyer, Prof. Dr. med. Dr. med. dent. Thomas Dietrich, Prof. Dr. med. Gerd Rüdiger Burmester, and Prof. Dr. med. Thomas Dörner in issue 20/2017

Filamentary Keratitis as Indication

In their review on Sjögren's syndrome (1), the authors Stefanski and co-workers highlighted the interdisciplinary care required for this serious chronic inflammatory autoimmune disease. In addition to subjective patient symptoms of eye irritations, including foreign body sensation, burning, stinging, and photophobia, objective signs can be very important for diagnosis; these include not only Schirmer's test (as mentioned by the authors) but also superficial punctate keratitis and pronounced conditions of filamentary keratitis. For tearing deficiencies, as occur in sicca syndrome, not only a reduction of the aqueous phase of the tear film but also a disturbed lipid phase (meibomian gland dysfunction) and a reduced mucin phase (goblet cell dysfunction) should be taken into account (2). Despite intensive therapy with artificial tear drops, sicca syndrome can be so pronounced that it can lead to incapacity for work.

DOI: 10.3238/arztebl.2017.0660a

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Further Symptoms of Sjögren's Syndrome

The article by Stefanski et al. provides a good overview of the diagnosis and therapy of Sjögren's syndrome (1).

In the case of acute skin vasculitis, which is often associated with microbleeds that are detectable by capillary microscopy, fondaparinux should be used at a dose of 2.5 mg once a day until the vasculitis has subsided (2).

Skin vasculitis is also often associated with circulating immune complexes, which sustain vasculitis and can lead to loss of protein through the kidneys (3).

In my experience, patients with this condition can be successfully treated using a dose of approximately 420 mg bromelain three times a day to eliminate circulating immune complexes, and a dose of 18 to 54 mg rutin (taken as either tea or tablets) to seal blood vessels

(especially in kidneys). In addition, capillary microscopy examination often reveals acrocyanosis or Raynaud's phenomenon, which can be treated with an alpha-receptor blocker or calcium channel blockers, respectively (4).

Further, the inflammation often leads to iron deficiency, which should likewise be treated. Chronic fatigue syndrome can also be associated with anemia due to low erythrocyte count. In this case, anemia can be treated with erythropoietin, usually in combination with iron.

DOI: 10.3238/arztebl.2017.0660b

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In Reply:

We would like to thank the authors of the correspondences for their constructive contributions.

We fully agree with Prof. Schmidt that Schirmer's test is not a suitable examination for determining corneal damage. Rather, examinations carried out by ophthalmologists are necessary for this purpose, such as the local application of vital dyes (Lissamine green or fluorescein), as recommended in the new classification criteria cited. This also allows assessment of cornea and conjunctiva damage to be determined, which in turn plays a role in the classification criteria. We are especially grateful that additional causes of dry eyes' challenges with intact aqueous phase were mentioned, due for instance to a disturbed lipid or mucin phase. For reasons of space, this was only shown for differential diagnoses purposes in *Table 1* (1) of our article.

The working capacity of persons with Sjögren's is reduced as compared to the normal population. In addition to sicca symptoms, fatigue, age, and the burden of fibromyalgia appear to be the most important predictors of this (2).

We thank Prof. Kieseewetter for pointing out that Raynaud's phenomenon is frequent (although unspecific), occurring in 10% to 37% of persons with primary Sjögren's syndrome (pSS). Fortunately, the

symptoms are relatively mild; the available treatment options, including avoidance of cold and calcium channel blockers, were recently presented very extensively (3). Capillary microscopic changes are also described in pSS patients with and without Raynaud's, as described for example in the publication by Corominas H. et al. cited by Kiesenwetter. However, no specific pattern for pSS has been detected. Immune complex glomerulonephritis with pronounced proteinuria can also occur in pSS and is most frequently associated with cryoglobulinemic vasculitis. However, glomerulonephritis is rare in pSS as compared to tubulointerstitial nephritides (differential diagnosis of systemic lupus erythematosus or overlap syndromes) (4). The therapeutic possibilities mentioned (bromelain and rutin) have not been supported by adequate trial data. Prophylactic anticoagulation is not necessarily indicated for mild skin vasculitis unless other risk factors for thromboembolic events, such as immobilization and hospitalization, are also present (5).

DOI: 10.3238/arztebl.2017.0660c

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Conflict of interest statement

The authors of all contributions declare that no conflict of interest exists.

CLINICAL SNAPSHOT

A Rare Cause of Ileus in a Two-Year-Old Boy

A previously healthy two-year-old boy was admitted to the hospital because of abdominal pain and vomiting of two days' duration. He looked unwell, and there was guarding on examination of the abdomen. The leukocyte count was 13 000/μL, the C-reactive protein (CRP) concentration was 23.2 mg/dL, and the potassium concentration was 2.9 mmol/L. Ultrasonography revealed a large mass in the mid-abdomen on the left side, and a plain abdominal x-ray revealed fluid levels. At emergency laparotomy, a large Meckel's diverticulum, necrotic because of torsion, was found and removed. The diagnosis was histopathologically confirmed, and the postoperative course was free of complications.

Meckel's diverticulum, a remnant of the omphaloenteric duct of the embryo, has an incidence of approximately 2% of the population (ca. 8 of every 10 000 persons), but only 4% of the affected persons develop symptoms. Torsion of a Meckel's diverticulum is a rare cause of ileus in childhood that should always be included in the differential diagnosis of an acute abdomen in a child. Ultrasonography should be performed; if it does not demonstrate the intra-abdominal structures clearly, it should be supplemented by further diagnostic studies.

Figure: A Meckel's diverticulum, massive and necrotic because of torsion, just before resection

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Conflict of interest statement: The authors state that they have no conflict of interest.

Cite this as: Leonhardt J, Glüer S: A rare cause of ileus in a two-year-old boy. *Dtsch Arztebl Int* 2017; 114: 661. DOI: 10.3238/arztebl.2017.0661

Translated from the original German by Ethan Taub. M.D.