CORRESPONDENCE

The Diagnosis and Treatment of Idiopathic Facial Paresis (Bell's Palsy)

by Prof. Dr. med. Josef Georg Heckmann, Prof. Dr. med. Peter Paul Urban, Prof. Dr. med. Susanne Pitz, Prof. Dr. med. Orlando Guntinas-Lichius, and Prof. Dr. med. Ildikó Gágyor in issue 41/2019

Test Is Also Helpful in Clinically Unclear Cases of Varicella Zoster

Regarding the laboratory tests explained in the article, the importance of varicella zoster virus (VZV) IgA antibodies in the serological diagnosis of (VZV) infection should be emphasized (1). Whereas mostly IgM and often also IgA antibodies are detectable during the acute primary infection (chickenpox), the IgM antibody response is lacking in reactivated VZV infection (herpes zoster) in 50–65% of cases (2, 3). In this setting, however, in addition to the obligatory VZV-IgG antibodies, VZV IgA antibodies are found one to four weeks after disease onset (2, 3). These can persist for three to six months. In two studies, the sensitivity of the VZV-IgA antibodies in herpes zoster was 67–94% (3, 4), the specificity is as high as 94% (4).

For this reason, VZV serology is also helpful in atypical or clinically unclear cases of zoster, such as "herpes sine herpete," where the typical blisters are lacking, or in zoster encephalitis.

However, VZV-IgA antibodies and VZV DNA in the serum have been repeatedly found in asymptomatic persons (subclinical reactivation). The test for varicella DNA (PCR) in cerebrospinal fluid or in blister fluid is highly sensitive, but its use in the outpatient setting is not covered by the statutory health insurers.

A test for VZV specific CD4-T-cells, which in one study showed 100% specificity and almost 100% sensitivity, is still in the experimental stages (4).

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Essential Comments

I have three comments regarding this article (1):

- Cortisone should only be administered once an infection with borrelia bacteria has been ruled out. I remember a young patient who attended a neurology hospital with acute unilateral facial paresis, and he was administered cortisone. On the fourth day he came to me as a medical emergency. The facial paresis had become bilateral, and the patient had a headache and clear meningism. The cortisone had led to an "explosion" of neuroborreliosis, and the patient was admitted to intensive care.
- Electrotherapy is effective and shortens the disease course. It has to be administered early and on working days. Patients will have to perform the necessary facial exercises in front of a mirror three times every day.
- If symptoms are slow to recede or even persist, ultrasound scanning of the parotid region should be undertaken in order to rule out a possible tumor.

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In Reply:

We thank both correspondents for their contributions.

Of course, borrelia infection will have to be ruled out before corticosteroid treatment is given, as Beutner mentions. In our article we do not address this aspect directly, but we did emphasize that neuroborreliosis and herpes zoster oticus are the most common causes with a defined etiology and should be referred to diagnosis-specific treatment in the decision pathway (1).

Whether steroid administration at the recommended dosages generally brings neuroborreliosis to an "explosion" should be considered a clinical observation by our correspondent. In the individual case, antibiotic treatment is combined with steroid and cyclophosphamide administration in vasculitis associated with borreliosis (2). Regarding electrotherapy, the most recent meta-analysis does not confirm any scientific evidence for its use. Its prescription lies with the individual physician, according to the rules of drug prescription. Similarly, the literature does not provide evidence supporting facial exercises (3), even though these make sense psychologically speaking. In this context, we support the idea for patients to undertake such

exercises three times daily in front of the mirror. The comment that if the paresis recedes very slowly or persists, ultrasound scanning of the parotid region should be undertaken, is very important and commendable.

Regarding symptomatic treatment in lagophthalmos, we personally are in receipt of a letter from an ophthalmologist, who recommends occlusion by means of an adhesive dressing as an alternative to the nocturnal moisture-retaining eye shield. In his view, patients experience this as less disruptive and are able to conceal the dressing by wearing sunglasses during daytime. In the author's experience, such dressings are less stigmatizing than a moisture-retaining eye shield.

Maire mentions the importance of VZV-IgA antibodies and the parameter "VZV-specific CD-4-T-cells," which to date is experimental. VZV-IgA can be of great importance in routine clinical practice and possibly even close a diagnostic gap in cases of zoster sine herpete

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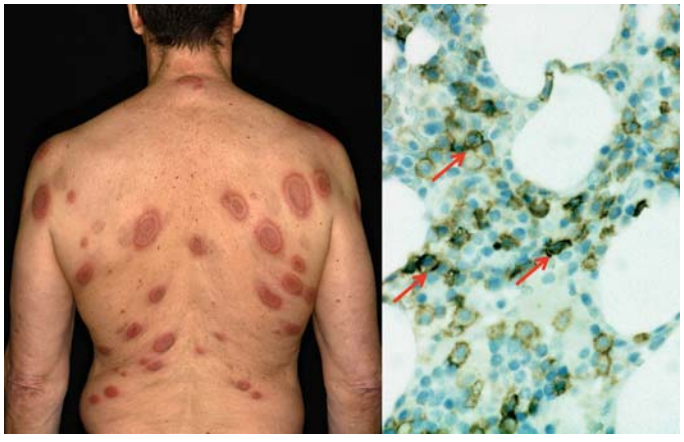
The authors of all contributions declare that no conflict of interest exists.

CLINICAL SNAPSHOT



Left: Multiple succulent erythematous plaques on the trunk
Right: Bone marrow infiltration with CD103-positive atypical lymphocytes (markers, magnification 400 ×; reproduced by kind permission of Prof. K.-F. Bürrig, Hildesheim)

Sweet's Syndrome as Precursor of Hairy Cell Leukemia

A 57-year-old man reported a 6-month history of skin lesions without systemic symptoms. We found succulent erythematous plaques concentrated mainly on the trunk. Based on the clinical findings and confirmatory histology, we diagnosed Sweet's syndrome. An extensive work-up to rule out malignancy (differential blood count, thoracic and abdominal computed tomography, esophagogastroduodenoscopy, and colonoscopy) revealed no abnormal findings. The patient was treated with prednisolone in varying doses accompanied by 100 mg/d dapsone, with moderate success. Over the ensuing 12 months he developed progressive pancytopenia. Examination of a bone marrow sample to exclude hematological disease showed infiltration by a B-cell non-Hodgkin lymphoma (CD103+) with inhibition of hematopoiesis, consistent with hairy cell leukemia. The patient received a cycle of treatment with the cytostatic drug cladribine and has been in hematological remission ever since. The skin lesions healed a short time later. Sweet's syndrome is often found together with infection, but in 10% to 20% of cases it is accompanied by a hematological malignancy, and occasionally a solid tumor. Chronic recurrence of Sweet's syndrome should prompt investigation for an associated malignancy.

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