Libyan International Medical University

Faculty of Basic Medical Science

Student name : noha hussin elsawadi

Role number : 1437

Dctore: sara elmegerhi

Sjogren's syndrome

Abstract:

Sjogren's syndrome is an autoimmune disease. It is a chronic and slowly progressive disease. The main pathology being, lymphocytic infiltration of the exocrine glands, resulting in xerostomia and dry eyes. Though dryness of eyes, dryness of mouth and parotid enlargement are considered as triad of sjogren's syndrome, rarely it can present as hypokalemic periodic paresis with renal tubular acidosis. Here, is a case report of patient having recurrent hypokalemic paresis, metabolic acidosis, type 1 renal tubular acidosis occurring secondary to sjogren's syndrome.

introduction:

Sjogren–Larsson syndrome nor Marines –Sjogren syndrome. is a long-term autoimmune disease in which the moisture-producing glands of the body are affected. This results primarily in the development of a dry mouth and dry eyes. Other symptoms can include dry skin, vaginal dryness,{1} a chronic cough, numbness in the arms and legs, feeling tired, muscle and joint pains, and thyroid problems. Those affected are at an increased risk (5%) of lymphoma. While the exact cause is unclear, it is believed to involve a combination of genetics and an environmental trigger such as exposure to a virus or bacteria. It can occur independently of other health problems (primary Sjogren syndrome) or as a result of another connective tissue disorder (secondary Sjogren syndrome). The inflammation that results progressively damages the glands. Diagnosis is by biopsy of moisture-producing glands and blood tests looking for specific antibodies.[On biopsy there are typically lymphocytes within the glands. Between 0.2% and 1.2% of the population are affected,{1} with half having the primary form and half the secondary form. Females are affected about ten times as often as males and it commonly begins in middle age; however, anyone can be affected. Among those without other autoimmune disorders, life expectancy is unchanged.

Case report 1:

The patient was a 50-year-old woman with newly diagnosed Sj?gren syndrome. She was positive for anti-SSA and anti-SSB autoantibodies and presented with stinging of the eyes, the sensation of a foreign body in the eye, and reduced salivary secretion (dryness of the mouth and altered sense of taste).

Discussion:

According to the European Community Study Group, the diagnosis of Sj?gren syndrome should be based on the results of: 1) biopsies of the minor salivary glands and/or lachrymal glands; 2) examination of the oral cavity consisting of sialography or the Saxon test (in which a sponge is chewed for 2 min by the patient and then weighed) combined with salivary-gland scintigraphy;{2} eye examination consisting of the Schirmer test plus the Bengal rose test or the fluorescein test; and 4) assays for anti-Ro/SSA or anti-La/SSB antibodies

For a definitive diagnosis, at least two of the four criteria must be met .Biopsy of the minor salivary glands is highly specific for Sjogren syndrome if the sample includes 5-10 glands and the connective tissues surrounding them. In recent years, however, MR-sialography and sonographic assessment of the major salivary glands have been playing increasingly important roles

case report 2:

After seven years of consulting rheumatologists, otolaryngologists, dentists, radiologists andophthalmologists, a 27-years-old female visited our unit of sonography, department ofradiodiagnosis, Giza hospital with bilateral swelling of parotid glands , dryness of mouth, eye and vagina, burning sensation of the tongue and oral mucosa, concomitant pain with swallowing and history of rheumatic fever.{3} The undiagnosed patient reported a regular treatment for her cardiac condition that is monitored and followed up by scheduled laboratory investigations.She was a mother of two daughters. Consanguineous marriage has been very frequent in herancestors for three generations according to her narration— a casual or causal finding that votesfor inheritance if it plays a vital part in providing a backcloth for the development of Sjogren's syndrome.

Discussion:

The patient was formally referred to the department of maxillofacial surgery and diagnosis, Shubra hospital for it enviablediagnostic reputation. A tentative diagnosis of Sjogren's syndrome (SS) was made based on the aforementioned signs and symptoms. Under local anesthesia, the surgeon harvested four minor salivary glands from a normally appearing labial mucosa. The extract was immersed immediately in 10% formalin to be submitted for microscopic examination. Meanwhile, some laboratory investigations including complete blood picture, erythrocyte sedimentation rate, antinuclear acid, rheumatoid factor,{3} anti-Sjogren's syndrome-A (SS-A) and anti–Sjogren's syndrome-B (SS-B) were performed. The clientele was also referred to a gynecologist to treat and check for any pertinent findings other than the vaginal dryness. Lab results revealed an elevated ESR, positive ANA, strong positive SS-A and positive SS-B. The rheumatoid factor was positive and CBC showed lymphocytopenia, absolute neutropenia, hypochromic microcytic anemic and thrombocytopenia. The histologic examinations viewed a confluence of lymphocytic infiltrate..

case report 3 :

38 years old female referred from private hospital on ventilator support secondary to quadriparesis with respiratory failure. On detail history taking she had history of recurrent quadriparesis. On examination she was drowsy and was responding to verbal commands in the form of eye opening{4}. She had hypotonia in all four limbs, power was grade two in all four limbs, diminished reflexes and mute plantars. On admission BP was 110/90 mm Hg, HR was 85/min, RR was 26/min and saturation was 99% on ventilator SIMV mode. Since she had spontaneous respiratory efforts,

she was taken on CPAP mode and gradually extubated in next 2 days. On admission blood biochemistry revealed Na+ 143 mmol/l/; k+2.2mmol/l; RBS 116mg/dl. liver function test; renal function tests and complete blood counts were normal. ABGA revealed metabolic acidosis with Ph. of 7.13. ECG showed prominent u wave. She was treated with intravenous potassium correction. Patient recovered completely in next 3 days. On enquiring she had history of dryness of mouth, eyes-features s/o sicca syndrome. Auto antibodies test were done and she was Ro/SS-A-positive and La/SS-B-Positive. She had positive schirmers test.

Dicussion:

Sjogren's syndrome usually presents with features such asxerostomia, kerato conjunctivitis sicca. and parotid enlargement. The Extra glandular manifestation includes, Arthralgia/arthritis-60%, Raynaud's phenomenon-37%, Vasculitis-11%, Renal-9%, Lungs/lymphadenopathy-6%, Splenomegaly-3%, Peripheral neuropathy-2% and Myositis-1%studied 31 cases of hypokalemic periodic paralysis where 3 cases had sjorgen's Syndrome. This indicates that 10% of people with hypokalemic periodic paralysis can have Sjogren' syndrome which if diagnosed early has more management options available. Patient was treated with hydroxychloroquin, bicarbonate and potassium supplement and supportive measures. Rheumatologist,{4} nephrologist, dermatologist, and gynecologist opinion taken and done accordingly. Patient recovered and was discharged within 8 days. Sjogren's syndrome rarely presents with hypokalemic periodic paralysis secondary to renal tubular acidosis with respiratory failure, therefore timely clinical suspicion and early diagnosis is important

Conclusions :

Sjögren syndrome is a systemic autoimmune disease that affects the exocrine glands. Diagnosis of the syndrome is generally based on biopsy of the minor salivary glands and/or lachrymal glands, examination of the oral cavity and eyes, and autoantibody positivity. More recently, MR-sialography and ultrasound studies of the major salivary glands have been playing more important roles because they are noninvasive and easily repeatable. Various criteria have been assessed for classification of sonographic findings: the most recent approach focuses on the echogenicity of the salivary gland parenchyma relative to that of the thyroid**,{5}** glandular homogeneity, the presence of hypoechoic areas, parotid and submandibular foci of hyperechogenicity, and characteristics of the margins of the salivary glands. Sonographic exploration of the salivary glands in patients with Sjögren syndrome is effective, noninvasive, and highly repeatable**.{6}** It provides accurate information on the vascularization of the glands, which reflects the activity of the disease.

References :

1. Obinata K., Sato T., Ohmori K., Shindo M., Nakamura M. A comparison of diagnostic tools for Sjögren syndrome, with emphasis on sialography, histopathology and ultrasonography. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2010;109:129–134. [PubMed]

2. Takagi Y., Kimura Y., Nakamura H., Sasaki M., Eguchi K., Nakamura T. Salivary gland ultrasonography: can it be an alternative to sialography as an imaging modality for Sjögren's syndrome? Ann Rheum Dis. 2010;69:1321–1324. [PubMed]

3. Niemelä R.K., Takalo R., Pääkkö E., Suramo I., Päivänsalo M., Salo T. Ultrasonography of salivary glands in primary Sjögren's syndrome. A comparison with magnetic resonance imaging and magnetic resonance sialography of parotid glands. Rheumatology. 2004;43:875–879. [PubMed]

4. Salaffi F., Peroni M., Ferraccioli G.F. Discriminating ability of composite indices for measuring disease activity in rheumatoid arthritis: a comparison of the Chronic Arthritis Systemic Index, Disease Activity Score and Thompson's articular index. Rheumatology (Oxford) 2000 Jan;39(1):90–96. [PubMed]

5. Hocevar A., Rainer S., Rozman B., Zor P., Tomsic M. Ultrasonographic changes of major salivary glands in primary Sjögren's syndrome. Evaluation of a novel scoring system. Eur J Radiol. 2007 Sep;63(3):379–383. Epub 2007 Mar 6. [PubMed]

6. Shimizu M., Okamura K., Yoshiura K., Ohyama Y., Nakamura S. Sonographic diagnosis of Sjögren syndrome: evaluation of parotid gland vascularity as a diagnostic tool. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2008;106:589–594. [PubMed]