Original Research Paper

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General Medicine

VARIED PRESENTATIONS OF SJOGREN'S SYNDROME

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ABSTRACT Sjogren's syndrome is one of the common chronic auto immune exocrinopathy. Apart from Glandular manifestations; extra glandular manifestations also seen with Sjogren's syndrome which include multitude of manifestations like simple myalgia to life threatening organ system and vasculitis. We are presenting three cases of Sjogren's syndrome which presented atypically.

KEYWORDS : Sjogren's, Rheumatoid Arthritis MCTD

CASE SERIES:

Case-1:35yr female; known case of seropositive RA diagnosed 6months ago now presented with complaints of parotid swelling and dysphagia for solids. Upon evaluation B/L parotitis with enlargement; Xerostomia & xerophthalmia evidenced by Schirmer's I test. Later found to have high titres of U1 RNP & met the criteria of Khan for MCTD treated accordingly.

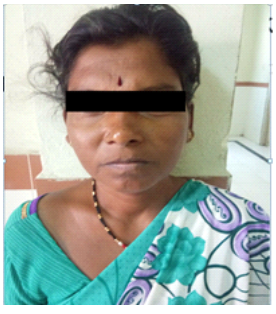
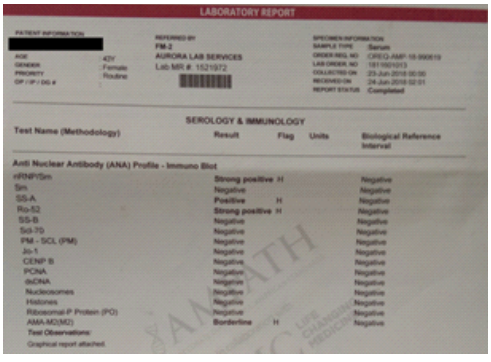
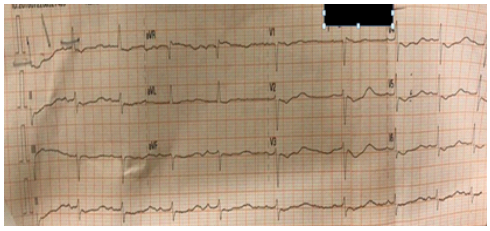


Fig-1-showing Bilateral parotid Enlargement

Investigations:



Case-2: 33y female presented with second episode of Hypokalemic paralysis; On evaluation found to have Hyperchloremic Metabolic Acidosis; High U. TTKG; distal RTA; serology confirmed high titers of Anti Ro-52; Anti SS-A antibodies & imaging showed evidence of Sjogren's syndrome and treated accordingly.



ECG showing Hypokalemia

Case-3: 35y F came with hematuria and petechiae. On examination gross pallor was present. Later on evaluation found to have coombs positive Hemolytic anemia with thrombocytopenia. On further work up she was found to be having Sjogren's syndrome with serological positivity of Anti SS-A antibodies in high titers without typical glandular manifestations.

Investigations:

- S. Bilirubin : 2.1mg/dl
- SGOT : 20 U/L
- SGPT : 51 U/L
- ALP : 74 U/L
- S. creatinine : 0.5mg/dl
- B. Urea : 17mg/dl
- S. Na+/ K+/ Cl- : 143 / 5 / 108
- Viral markers : Negative
- ESR : 10mm /1st hr
- CRP : +ve
- DCT: +ve
- ICT : +ve

DISCUSSION:

We are discussing a varied presentations of Sjogren's syndrome. (Case-1) Mixed connective tissue disease (MCTD) is an inflammatory disease characterized by combined features of systemic lupus erythematosus (SLE), systemic sclerosis (SSc), and polymyositis (PM) that is associated with high serum titers of antibody to extractable nuclear antigen, with a specificity for nuclear ribonucleoprotein[1]. The large spectrum of clinical signs and symptoms of MCTD includes sicca symptoms and Sjögren's syndrome (SS). However, the actual frequencies of dry eye and SS in MCTD patients are poorly characterized[2,3].

Case-2: Hypokalemic paralysis is caused by a number of underlying etiologies, namely, genetic, endocrine, gastrointestinal, and renal.Renal tubular acidosis (RTA) and thyrotoxicosis constitute the major causes of acquired hypokalemic paralysis. Distal RTA is the common pathway for potassium loss in a variety of diseases including connective tissue diseases such as Sjogren's syndrome (SS).SS is a chronic autoimmune inflammatory disease with an estimated prevalence ranging from 0.1% to 4.8%, affecting mainly middle-aged females and primarily involves the exocrine glands.The syndrome can present either alone (primary SS [pSS]) or in the context of an underlying connective tissue disease (secondary SS).The prevalence of renal involvement in pSS ranges from 18.45% to 67%. RTA with hypokalemic paralysis as a presenting feature of pSS is described in few case reports in literature. We present here a case series of 13 patients of pSS presenting as hypokalemic paralysis and review the related literature[4].

Case-3: Primary Sjogren's syndrome is an autoimmune disease wherein there is lymphocytic infiltration of salivary and lacrimal glands. This inflammation is thought to be caused by B-lymphocytes. The most common clinical feature of Sjogren's is dryness of the mouth and eyes, but rare complications can occur such as autoimmune cytopenias[5].

CONCLUSION:

High index of clinical suspicion is necessary to diagnose Sjogren's syndrome presenting with atypical manifestations. To develop a typical landular manifestation, it may take a median of 6 yrs from onset of 1st symptom. Patient may show serological positivity far ahead of actual typical / atypical manifestation to occur.

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