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CASE REPORT

Primary Sjogren's syndrome presents as recurrent meningoencephalitis

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A B S T R A C T

Primary Sjogren's syndrome is an autoimmune disease mostly affecting middle-aged females. It usually involves the exocrine glands system resulting in dryness of the mouth and eyes. It rarely involves the Central nervous system. Here we present a case of Sjogren's syndrome with a rare presentation at an unusual age. An 80 years old patient who presented with 3 episodes of recurrent aseptic meningitis in a year, was diagnosed with primary Sjogren's syndrome after extensive workup. She responded well to immunomodulatory treatment. Clinicians should consider the rare presentation and age of autoimmune diseases when facing the challenging clinical situation.

Keywords: *Sjogren's syndrome, Meningitis, Central nervous system*

Introduction

Sjogren's syndrome (SS) is a chronic autoimmune inflammatory condition marked by impaired lacrimal and salivary gland activity, resulting in eye and mouth dryness. However, multisystem involvement can occur in it. The age of onset is 40-60yrs with female predominance.[1] Central Nervous System (CNS) manifestations occur in 25% of patients, ranging from diffuse symptoms to focal white matter lesions.[2] Meningoencephalitis and pachymeningits have been described in the literature as a rare manifestation of SS. [3, 4] Here we present a case of an 80-year-old female with recurrent episodes of meningoencephalitis secondary to Sjogren's syndrome.

Case report

An 80-year-old diabetic female was brought to Emergency Reception (ER) with a short history of fever and headache followed by depressed consciousness. There was no associated vomiting, photophobia, fits, body weakness, or falls. On examination in ER, she was febrile (100 F), and GCS was 11/15 with signs of meningeal irritation. The rest of the examination was normal. She experienced the same illness twice; 6 and 11 months back. In between the episodes, she remained normal. She was investigated extensively to look for the cause of recurrent meningoencephalitis (Table 1).

MRI brain with contrast showed diffuse Pachymeningitis. CTA showed no significant abnormality in vessels. In her previous admissions, she received IV acyclovir and a short course of steroids, she responded well within 48hrs and had full functional and cognitive recovery within a week. This time she was given only supportive care, and her improvement was slightly delayed

but like her previous admissions, she regained her prior functional status. To look for the cause of this recurrent aseptic pachymeningitis, Antineuronal and anti-paraneoplastic antibodies (CSF), Whole body CT scans were done that was unremarkable except for enlarged parotid glands on CT scan as an incidental finding. After excluding other causes, her autoimmune profile was sent that showed strong Positive ANA, Anti SSA, and SSB antibodies. The test results were confirmed by two laboratories 15 days apart. So the cause of her recurrent aseptic meningitis/pachmeningitis turned out to be Sjogren's syndrome (Figure 1a, b).

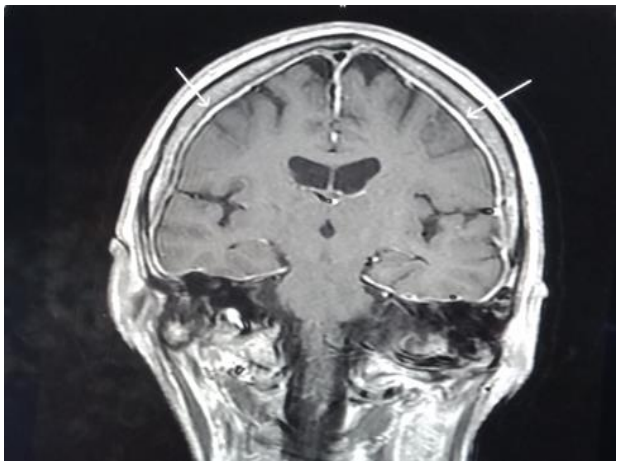

Figure 1a: Contrast-enhanced MRI brain coronal section. Arrows pointing towards enhanced pachymeninges.

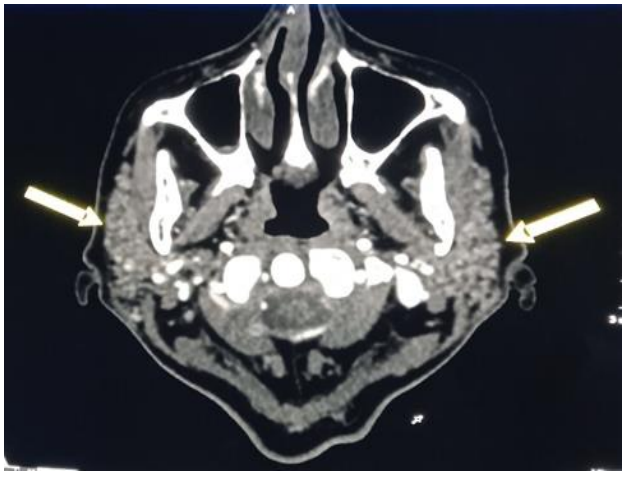

Figure 1b: Arrow indicating diffuse hypertrophied parotid glands

On exploring, the patient gave a history of easy fatigability, weight loss, occasional oral soreness, and arthralgia. However, there was no H/o head and neck radiation, organ transplant, or any drug use other than gliclazide. Normocytic normochromic anemia and parotid gland hypertrophy also favored the diagnosis of SS. Furthermore, she had a dry tongue, and multiple dental caries, a slit lamp examination showed dry and keratinized conjunctiva and cornea, and Schirmer's test was positive in both eyes. As her CNS symptoms had resolved completely when the diagnosis was made, and there was no indication requiring high-dose steroids so she was started on Hydroxychloroquine 200mg once daily. The patient is still under follow-up and has neither developed any further episodes of aseptic meningitis nor any other disease flare in the last 1year.

Discussion

The case under discussion describes a rare presentation of Sjogren's syndrome (recurrent aseptic meningoencephalitis / pachymeningitis) at an unusual age. We diagnosed the case after extensive laboratory workup because the presentation and age both were very rare for any autoimmune disease. We used the 2016 American College of Rheumatology / European League Against Rheumatism (ACR-EULAR) criteria for our diagnosis of SS, our case satisfied the criteria for diagnosis with a score of 4 [positive anti-SSA/Ro antibody (score 3), positive Schirmer's test (score 1).[5]

In literature, Lee et al. reported a young patient with recurrent aseptic meningitis secondary to SS who could not meet ACR-EULAR criteria, and their diagnosis was supported by diagnostic scintigraphy.[6] Rossi, R et al. also describe the case of a 58-year-old female presenting with signs of meningism along with CN IV deficit, SS was confirmed by salivary gland biopsy and was resolved with steroid therapy with dexamethasone.[7] Both cases had diffuse leptomeningeal enhancement on brain imaging,[6, 7] whereas our case had diffuse pachymeningeal enhancement. On the other hand, only a couple of cases of SS-related pachymeningitis have been reported, those presented with a variable focal deficit but none had recurrent aseptic meningitis. [4, 8, 9]

Unusual age, and rare clinical and radiological presentation made the case peculiar. Secondly, we excluded all other possible causes in addition to fulfilling ACR/EULAR criteria. Furthermore, follow-up after a year with no recurrence of neurological symptoms also strengthened our case.

Table 1: Summary of lab investigations in three admission

Lab parameters	1st admission (Aug 2019)	2nd Admission (March 2020)	3rd Admission (September 2020)
Blood film	Normocytic normochromic anemia	Normocytic normochromic anemia	Normocytic normochromic anemia
C-reactive protein	0.5	0.4	0.5
ESR	-	-	53
Renal profile	WNL	WNL	WNL
Liver profile	WNL	WNL	WNL
HBA1C level	8.9%	8.1%	8%
B12 level	-	-	WNL
Serum Folate	-	-	WNL
Serum Ionized Calcium	-	-	WNL
ACE levels	-	-	WNL
Vit D levels	-	-	WNL
TSH	Normal	Normal	WNL
Anti-HIV Antibodies	-	-	Negative
Anti HCV antibodies	-	-	Negative
Shyphilis serology	-	-	Negative
Urine routine analysis	Protein +1	-	Normal
Cardiac workup (ECG, ECHO)	Normal	Normal	Normal
CSF Routine analysis	Proteins 111mg/l Cells< 5 (100% L) Glucose 112mg/dl (more than 50% of serum blood sugars at that time) LDH 35	Protein 67mg/l Cells 29 (100% L) Glucose 157mg (more than 50% of serum blood sugars at that time) LDH 28	Proteins 95mg/dl Cells 36 (100% L) Glucose 157mg (more than 50% of serum blood sugars at that time) LDH 121
CSF gram stain	negative	negative	negative
CSF Mycobacterium TB PCR and culture	negative	negative	negative
CSF for fungal stain and Culture	negative	negative	negative
CSF for HSV I & II PCR	Target not detected	Target not detected	Target not detected
CSF bacterial Culture	No growth	No growth	No growth
CSF cytology for malignant cells	No malignant cells seen	No malignant cells seen	No malignant cells seen

WNL= within normal limit

Conclusion

Clinicians should consider the rare presentation and age of autoimmune diseases when facing a challenging clinical situation.

References

1. Witte T. Sjögren-Syndrom. Zeitschrift für Rheumatologie. 2019; 78(6):511-7.
DOI: https://doi.org/10.1007/s00393-019-0625-8
2. Moreira I, Teixeira F, Martins Silva A, Vasconcelos C, Farinha F, Santos E. Frequent involvement of central nervous system in primary Sjögren syndrome. Rheumatol Int. 2015; 35(2):289-94.
DOI: https://doi.org/10.1007/s00296-014-3097-9
3. Akiyama T, Oiwa H, Hayashi M, Sugimoto T, Nomura E, Yamawaki T. Aseptic meningitis as an initial presentation of Sjögren syndrome: a report of two cases and literature review. Nagoya J Med Sci. 2020; 82(3):595.
DOI: https://doi.org/10.18999/nagjms.82.3.595
4. Heo Y, Kim T. Case report: hypertrophic pachymeningitis associated with Sjögren's syndrome. Neurol Sci. 2019; 40(4):879-82.
DOI: https://doi.org/10.1007/s10072-018-3637-2.
5. Shiboski CH, Shiboski SC, Seror R, Criswell LA, Labetoulle M, Lietman TM, et al, Scofield H, Vitali C, Bowman SJ, Mariette X. 2016 American College of Rheumatology/European League Against Rheumatism classification criteria for primary Sjögren's syndrome: a consensus and data-driven methodology involving three international patient cohorts. Arthritis Rheumatol. 2017; 69(1):35-45.
DOI: https://doi.org/10.1002/art.39859
6. Lee DH, Lee SJ. Recurrent aseptic meningitis as an initial clinical presentation of primary Sjögren's syndrome. J Neurocrit Care. 2019; 12(1):46-50.
DOI: https://doi.org/10.18700/jnc.190077
7. Rossi R, Saddi MV. Subacute aseptic meningitis as a neurological manifestation of primary Sjögren's syndrome. Clin Neurol Neurosurg. 2006; 108(7):688-91.
DOI: https://doi.org/10.1016/j.clineuro.2005.05.015
8. Nakano Y, Yamamoto M, Komatsu K, Yagita M, Fujita M. Hypertrophic pachymeningitis in Sjögren's syndrome. Intern Med. 2018; 57(3):413-5.
DOI: https://doi.org/10.2169/internalmedicine.9406-17
9. Abdelhedi H, Khammassi N. Pachyméningite et syndrome de Gougerot Sjörgren. Pan Afr Med J. 2015; 22:88.
DOI: https://doi.org/10.11604/pamj.2015.22.88.7840