Beyond Dry Eyes and Mouth: Cramp Fasciculation Syndrome Revealing Primary Sjogren's Syndrome



Anandvardhan Sharma¹, **Varun Kumar Singh**¹, Rameshwar Nath Chaurasia¹, Pratishtha Sengar², Deepika Joshi¹, Abhishek Pathak¹, Vijaya Nath Mishra¹, Anand Kumar¹

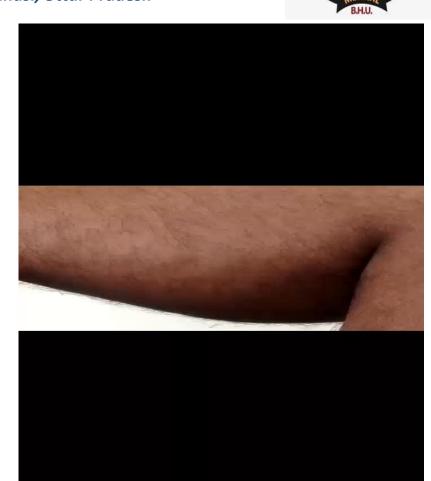
¹Department of Neurology, ²Department of Pathology, Institute of Medical Sciences, Banaras Hindu University, Varanasi, Uttar Pradesh

Case Description

- ✓ 26-year-old male, with no comorbidities presented with grittiness of eyes, dryness of mouth, bilateral calf pain, cramping type, worsened by walking and relieved partially by massaging/ compresses for 2 months.
- Muscle twitching started from calves progressing to arms, forearms, and trunk.
- Social impact: stopped wearing shorts and half sleeves shirts due to stigma

Examination

- √ Vitals: normal
- General examination: unremarkable.
- CNS examination: Continuous fasciculations in thighs, calves, arms, forearms, back.



Investigations		
CBC, LFT, RFT, TFT ESR, CRP, Lipid Profile, Calcium, Phosphorus, Vitamin D, B12	Normal	1.2
Viral markers	Negative	100ms 50p
Autoimmune encephalitis panel (LGI1, CASPR2)	Negative	
ANA profile	anti Ro-52 / SSA and anti La/ SSB 2+ positivity	Diagnosis Cramp Fasciculation Syndrome (CFS) secondary to Primary Sjogren's Syndrome (pSS)
Complements	Normal	
Shirmer test	< 5 mm in both eyes (positive)	Clinical Course ✓ Patient was managed with pulse dose of methylprednisolone (1gm per day) followed by oral steroids (1mg/kg body weight) and tablet
Electrophysiology		
Nerve Conduction Study	Normal	
Electromyography	 Spontaneous activity: Fasciculation potentials present (doublets, triplets and quadruplets) MUAPs: Neurogenic pattern 	 oxcarbazepine 600mg/day ✓ Pain and twitching improved after 1 week of treatment. ✓ Treating primary illness?

Discussion

- ✓ CFS: Rare disorder, part of peripheral nerve hyperexcitability spectrum
- ✓ Pathogenesis: Autoantibodies against VGKC/CASPR2
 → Nerve hyperexcitability
- ✓ Sjogren's: autoimmune connective tissue disorder; neurological involvement in 8.5-70% cases
- ✓ Usual neurological manifestation: sensory polyneuropathy
- ✓ Our case: rare presentation of CFS as initial manifestation of pSS

Conclusion

✓ Always evaluate systemic autoimmune disease as possible cause of CFS

References

- Sawlani K, Katirji B. Peripheral nerve hyperexcitability syndromes. CONTINUUM: Lifelong Learning in Neurology. 2017 Oct 1;23(5):1437-50.
- Ahmed A, Simmons Z. Isaacs syndrome: a review. Muscle & nerve. 2015 Jul;52(1):5-12.
- Katirji B. Peripheral nerve hyperexcitability. Handb Clin Neurol. 2019;161:281-290.