A RARE CASE OF CIDP SEEN IN YOUNG DIABETIC WITH UNUSUAL ASSOCIATION OF PERIPHERAL VASCULAR DISEASE AND COLOBOMA IRIDIS AND CHOROID

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PRESENTATION OF CASE

A 16-year-old girl presented to our OPD with h/o weakness of both lower limbs since 2 months. To begin with it involved proximal muscles and gradually involving the distal muscles over time. There was h/o associated paresthesias of limbs. However, no h/o bowel or bladder involvement was noted. She was diagnosed to have Type 1 Diabetes 2 years ago and started on insulin therapy. She was born out of consanguineous marriage. She also had h/o drooping of left eyelid since 1 year.

On examination HMF were normal, left iris coloboma present, fundi typical choroidal coloboma was seen. EOMs, mild restriction of upgaze present. Left eye ptosis present. Visual acuity was 6/18 in the right eye and 6/24 in the left eye, respectively. Other cranial nerves were normal. Generalised hypotonia and wasting present. Power at shoulder/ elbow was 4/5, fingers weak 2-3/5; hip, knee and toes 2-3/5. Sensations were normal. Plantars bilaterally mute, DTRs absent. Patient had an ulcer in the right foot with gangrenous changes. Peripheral pulses diminished in lower limbs bilaterally. Secondary sexual characters were found to be normal and without any menstrual abnormalities as ascertained by the Obstetrician. With this clinical background, a diagnosis of CIDP was thought and patient was evaluated thoroughly and opinion was sought from NIMHANS, Bangalore.

Chronic Inflammatory Demyelinating Polyradiculoneuropathy (CIDP) as the name suggests is an inflammatory neuropathy of autoimmune aetiology characterised by gradually progressive involvement of sensorimotor nerves. Classically, it causes progressive weakness of limbs, proximal-distal and symmetric in nature. Atypical forms mainly involving distal muscles are also described.¹ Age of onset is between 40 and 60 years with a mild male predominance. CIDP is seen in association with diabetes.² Unlike neuropathy seen in diabetics, CIDP is treatable and hence it is of paramount importance to identify CIDP in diabetics.

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We hereby report an interesting case of CIDP in association with Type 1 Diabetes in a 16-year-old female patient with unilateral ptosis, non-healing vascular ulcer in left foot and associated coloboma iris and coloboma choroid.

CBC/ RFT/ LFT/ Electrolytes were normal. Serum CPK and lactate and ammonia levels were normal. CSF examination revealed protein levels (123 mg/dL) without increase in cell count-

- HIV-NR, VDRL-NR, ANA- Negative.
- RBS- 300 mg/dL, HbA1c- 13.5.
- Thyroid profile- Normal.
- Nerve conduction studies revealed- demyelinating > axonal sensorimotor polyneuropathy.
- Electrophysiological study- VEP left side waves absent, ABR: normal, L/L SSEP absent.
- MRI Brain- Normal.
- Left biceps biopsy- neurogenic atrophy.
- Left sural nerve biopsy- vasculitic neuropathy with diabetic vascular changes.

These findings indicated presence of CIDP and patient was treated with Intravenous Methylprednisolone and small volume plasmapheresis. Her blood sugars were controlled with Insulin and regular dressings for the foot ulcer done.

Photo showing Ptosis on Left Side

Photo showing Foot Ulcer with Necrosis

Photo showing Choroidal Coloboma

DIFFERENTIAL DIAGNOSIS

In this case as there are multiple pathologies with single aetiology, many diagnoses were thought like diabetes with myasthenia gravis, diabetes with early manifestation of GBS. However, in view of varied presentation in our case, a syndrome was thought and proposed.

PATHOLOGICAL DISCUSSION

CIDP is a slowly progressive and sometimes relapsing neurological disorder with autoimmune aetiology. It is one of the causes of treatable neuropathies worldwide.³ Prevalence ranges from 1 - 9 cases per 100000.⁴

Typical presentation of CIDP includes involvement of both sensory and motor symptoms in the distal and proximal segments of the limbs areflexia, evolving over more than 8 weeks.

Raised CSF protein levels and slowing of nerve conduction are typical of the condition. There are atypical variants, which mislead and causes difficulty in diagnosis. Some of the phenotypic variants of CIDP are: Typical CIDP, Sensory CIDP, Chronic immune sensory polyradiculopathy, Lewis-Sumner syndrome, Focal CIDP, DADS, Acute onset CIDP and Motor CIDP.⁵ Assistance of muscle and nerve biopsy is very essential in making a definitive diagnosis of CIDP.

DISCUSSION OF MANAGEMENT

Steroids and intravenous immunoglobulin are effective, and plasma exchange can be helpful as rescue therapy. Role of immunosuppressants are still unclear. As such, there are no specific markers available for pinpointing the condition.⁶

In case reports presented by Husain and his colleagues, two of the diabetic patients were diagnosed to have CIDP with LMN facial palsy.⁷ Two case reports documented presence of CIDP with ptosis and third cranial nerve palsy,⁸ mostly after the onset of weakness of limbs. In a case report from Bradley and colleagues, they noted ptosis earlier in the patient and later on CIDP was diagnosed as similar in our case.⁹ In these case reports along with limb weakness, ocular manifestation was an important finding in CIDP.

FINAL DIAGNOSIS/ CONCLUSION

In our case, after extensive evaluation we concluded that the patient has various pathologies involving the endocrine (Type 1 DM), neuronal (CIDP) and vascular (PVD) probably of the same aetiology i.e. autoimmunity. And also, we have noticed patient having unilateral ptosis, which started early in the beginning as a part of CIDP. We are proposing PVD in this young patient as the foot ulcer remains unhealed and going in for gangrenous changes. Incidentally, we found the patient to have coloboma iridis and choroidal coloboma, though asymptomatic subjectively but has vision loss objectively. All these varied clinical manifestations with probably of same aetiology (Autoimmune) require even more research work to propose for a new syndrome.

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