Original Research Paper

Volume-9 | Issue-12 | December - 2019 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Medicine

OVARIAN MALIGNANCY PRESENTING AS PARANEOPLASTIC GBS

Dr. Radhika S — MD Anaesthesia, FNB Critical Care, Hemraj Jain hospital, Pushpanjali Enclave, Pitampura, Delhi 110034

Dr. S Ajitkumar — MD Medicine, Department of Medicine, Hemraj Jain hospital, Pushpanjali Enclave, Pitampura, Delhi 110034

Dr. Gagan Jain* — MD Medicine, Consultant Physician, Department of Medicine, Hemraj Jain hospital, Pushpanjali Enclave, Pitampura, Delhi 110034 *Corresponding Author

ABSTRACT Paraneoplastic disorders are immune mediated cross reactivity conditions triggered by onconeuronal antigens expressed by tumors by mechanisms apart from metastasis and cancer related complications. Ovarian tumors commonly cause cerebellar degeneration, opsoclonus, brainstem encephalitis and encephalomyelitis, and demyelinating polyneuropathy. However, axonal variant of Gullian Barre syndrome with an ovarian lymphoma is extremely rare. We report a very unusual case of ovarian lymphoma presenting as asymmetric lower motor neuron type of quadriparesis. She showed rapid neurological recovery upon surgical excision of mass and chemotherapy.

KEYWORDS : Paraneoplastic syndrome in ovarian malignancy, polyneuropathy

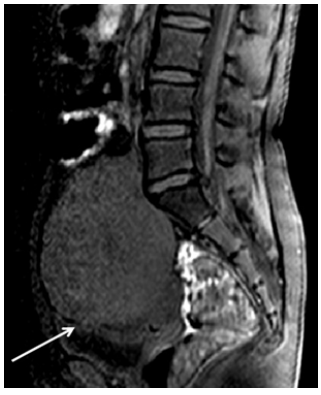
INTRODUCTION

A relentless search across the diagnostic spectrum can yield surprises. We encountered an ovarian malignancy manifesting as an unusual constellation of paraneoplastic features like thyroiditis and axonal predominantly motor neuronopathy. Gynecological manifestations were conspicuous by their absence despite the massive extent of tumor growth. The malignancy was indeed an accidental finding.

Case presentation

A 22-year-old female presented with acute onset of gradually progressive asymmetrical quadriparesis starting as asymmetrical paraparesis followed by distal predominant asymmetrical weakness of upper limbs since three weeks. She reported radicular pain from lower back up to the left foot, and in the neck radiating to both upper limbs since three weeks. She had no bulbar, facial, autonomic or respiratory symptoms. There was no antecedent history of fever, upper respiratory infection, and diarrheal illness. On examination, she had generalized hypotonia with neck weakness, asymmetrical areflexic quadriparesis including left wrist drop and left foot drop. Gait showed frequent buckling at the knee joint due to weakness.

Blood tests including thyroid function, serum creatine phosphokinase were normal. cerebrospinal fluid showed elevated protein level. Nerve conduction study and repetitive nerve stimulation test were normal. Electromyography showed denervation potentials in all involved muscle groups. Contrast enhanced MRI of brain and spine showed thickening and enhancement of the exiting C6 and C7 nerve roots in the cervical spine bilaterally. It showed an incidental finding of large pelvic mass [Figure 1].



Legend to Figure 1:

MRI Lumbosacral spine (plain and contrast) in sagittal view showing large homogeneous enhancing mass lesion (white arrow) in the pelvis

She was suspected to have an adnexal mass, probably a germ cell tumor as her serum LDH level was 280 mg/dL while the rest of the tumor markers were normal. Paraneoplastic antibody panel and autoimmune antibody panel were reported negative. Curiously, she never reported any menstrual irregularities or symptoms related to the adnexal mass.

A working diagnosis of atypical axonal variety of GB syndrome of paraneoplastic etiology secondary to unclassified ovarian tumour was formulated. Intravenous immunoglobulin (IVIG) was started at standard doses. No response was noted after five consecutive days of IVIG. Subsequently, the patient underwent exploratory laparotomy. A fertility preserving surgery i.e right adnexectomy with wedge biopsy of the left ovarian mass was done. Intra-operative findings showed a right adnexal solid mass completely replacing the right ovary, and a smaller but similar mass in the left ovary. Histopathological examination was suggestive of T cell lymphoblastic lymphoma. Patient was started on chemotherapy as per Berlin-Frankfurt-Munster protocol. On subsequent visits, her neurological status showed significant improvement.

DISCUSSION

Subacute motor neuronopathy in PNS manifests as subacute, progressive, painless, and often asymmetric lower motor neuron weakness. While pain may frequently accompany motor neuronopathy, few or no objective sensory deficits are found on examination.[1,2,3] There are only sporadic reports on nerve root enhancement on contrast enhanced MRI of spine in GBS. Contrast enhancement at the dorsal ganglia and exiting nerve roots can be explained by the absence of an effective blood nerve barrier at these points.[4] Furthermore, the demyelination process in GBS can also worsen the myelin integrity allowing infiltration by inflammatory cells.[5] The presence of nerve root enhancement can only supplement electrophysiological evidence and cerebrospinal fluid analysis favouring GBS. Treatment includes tumor excision, chemotherapy and immunomodulation for GBS with IVIG, steroids and plasma exchange. Response of GBS to immunomodulation in patients with co-existent malignancy is same as that in non-malignant conditions.[6]
To conclude, one must remember that ovarian malignancies may lie gynaecologically dormant. A clinician should suspect a paraneoplastic etiology in cases not responding to standard treatment. Investigations should cover the entire diagnostic spectrum from common to rare.

REFERENCES

1. Sladky JT. Guillain-Barré syndrome in children. J Child Neurol 2004; 19:191.
2. Devos D, Magot A, PerrierBoeswillwald J, et al. GuillainBarré syndrome during childhood: particular clinical and electrophysiological features. Muscle Nerve 2013; 48:247.
3. Mulkey SB, Glasier CM, ElNabbout B, et al. Nerve root enhancement on spinal MRI in pediatric GuillainBarré syndrome. PediatrNeurol 2010; 43:263.
4. Breger RK, Williams AL, Daniels DL, et al. Contrast enhancement in spinal imaging. AJNR Am J Neuroradiol 1989;10:633-637.
5. Mc Farlin DE. Immunological parameters in Gullian-Barre syndrome. Ann Neurol 1990;27(suppl):S25-S29.
6. Baran GA, Sowell MK, Sharp GB, Glasier CM. MR findings of Gullian-Barre syndrome . Radiat Med 1998 Mar-Apr;16(2):133-5.