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A CASE REPORT OF PERIPHERAL NEUROPATHY ASSOCIATED WITH PARANEOPLASTIC SYNDROME

Internal Medicine

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ABSTRACT

Paraneoplastic neurological syndromes (PNS) can be defined as remote effects of cancer that are not caused by the tumor and its metastasis, or by infection, ischemia or metabolic disruptions. PNS are rare, affecting less than 1/10,000 patients with cancer. Only the Lambert-Eaton myasthenic syndrome is relatively frequent, occurring in about 1% of patients with small cell lung cancer. PNS can affect any part of the central and peripheral nervous system, the neuromuscular junction, and muscle. They can be isolated or occur in association. In most patients, the neurological disorder develops before the cancer becomes clinically overt and the patient is referred to the neurologist who has the charge of identifying a neurological disorder as paraneoplastic. PNS are usually severely disabling. PNS are caused by autoimmune processes triggered by the cancer and directed against antigens common to both the cancer and the nervous system, designated as onconeural antigens. Due to their high specificity (> 90%), the best way to diagnose a neurological disorder as paraneoplastic is to identify one of the well-characterized anti onconeural protein antibodies in the patient's serum. In addition, as these antibodies are associated with a restricted range of cancers, they can guide the search for the underlying tumor at a stage when it is frequently not clinically overt. This is a critical point as, to date, the best way to stabilize PNS is to treat the cancer as soon as possible. As PNS are believed to be immune-mediated, suppression of the immune response represents another treatment approach.

KEYWORDS

INTRODUCTION

Paraneoplastic neurological syndromes (PNS) are a group of neurological disorders not directly caused by cancer metastasis, side effects of cancer treatment, nutritional deficiencies, metabolic derangements, or coagulopathies. Rather, PNS are secondary to an immune response triggered by the underlying tumor which affects the central or peripheral nervous system. These disorders can involve multiple levels of the neuraxis.

The most common PNS are Lambert-Eaton myasthenic syndrome (LEMS), subacute cerebellar ataxia, limbic encephalitis (LE), opsoclonus-myoclonus (OM), retinopathies (cancer associated retinopathy (CAR) and melanoma-associated retinopathy (MAR), Stiff-Person syndrome (SPS), chronic gastrointestinal pseudo obstruction (CGP), sensory neuronopathy (SSN), encephalomyelitis (EM) and dermatomyositis. Purkinje cell antibody type 1 (PCA-1, i.e. anti-Yo) and antineuronal nuclear antibody type 1 (ANNA-1, i.e. anti-Hu) were the most common onconeural antibodies in the studied population.

CASE REPORT

A 65-year-old male patient came to the department of general medicine, PDU Rajkot with the c/o of fever, cough with whitish expectoration in the last 10-15 days. Initially he noticed weakness in left lower limb then he gradually noticed weakness in right lower limb along with axial weakness.

On examination:

- HMF – Normal, Cranial nerves intact.
- Patient was fully conscious & oriented.
- Motor – Power Bilateral UL – 4/5 & LL – 0/5
- All four limb hypotonia
- Deep tendon reflex – Absent
- Babinski sign – Negative
- Bowel bladder reflex intact
- Sensory-Loss of all sensations

Investigation:

- HB – 13.10 g/dl
- WBC – 15,300 cmm
- Platelet – 2.3 lakhs cmm
- Creatinine – 0.8 mg/dl
- Sodium – 138 mEq/L
- Potassium – 4.1 mEq/L
- CSF – Protein 148 mg/dl total count 45, glucose 83
- Cxr -
- NCS- Primary severe exonal variant of pure motor poly-radiculoneuropathy (LL>UL)
- USG- NAD
- MRI BRAIN –Heterogeneously enhancing lesion is seen in the right parietal region with surrounding perilesional edema -Brain SOL (neoplastic Etiology). Diffuse cerebral atrophy. Ischemic demyelination changes are seen in the bilateral basal ganglia and periventricular region.
- CECT THORAX - A soft tissue density heterogeneously enhancing lesion of size 117*105*85 mm is noted in left upper lobe of lung; P/o bronchogenic carcinoma likely; multiples enlarged necrotic nodes; s/o metastatic mediastinal nodes. Osteolytic lesions in D1, D4, D5 vertebra and left 2nd rib. S/o bony secondaries.

DISCUSSION:

PNS are triggered by an immune response against onconeural antigens expressed by the tumor that are also expressed in the nervous system. These antigens are released after tumor-cell death and presented to the T cells by antigen-presenting cells in regional lymph nodes. This onconeural antigen-specific antibody or cell-mediated autoimmune response contributes to development of PNS.

The discovery of onconeural antibodies has led to the widely accepted nowadays hypothesis that PNS are immune-mediated disorders. The autoimmune hypothesis in PNS is also sustained by the inflammatory CSF findings and T cell infiltration in the affected part of the nervous system revealed by pathologic examination. However, the role of onconeural antibodies in neurological dysfunction is not clear. PNS pathogenesis is currently related to an aberrant expression in the tumor of an antigen that is normally expressed only in the nervous system. Even if the antigens in the tumor are identical in structure to the neural antigens, they could be recognized as foreign, leading to an immune attack directed against both the tumor and the nervous system. However, only few onconeural antibodies have been shown to play a direct role in the pathogenesis of the neurological symptoms.

PNS may affect any level of the nervous system (central or peripheral nervous system, including the neuromuscular junction and muscle). The severity of most PNS is due to the early and non-reversible destruction of neural structures by the inflammatory process. PNS are rapidly progressive, in many cases leaving the patients severely debilitated within weeks to months. A subacute progressive clinical course and a severe disability are highly suggestive of PNS. However, slow progression, relapses or a benign course do not exclude the diagnosis.

DIAGNOSIS::

PNS may affect any part of the central and peripheral nervous system, the neuromuscular junction and muscle. They can be isolated or occur in association. The incidence of malignancy in patients with potential PNS varies depending on the disorder and ranging from 5 to 60%. In almost 80% of patients, the PNS antedates the diagnosis of cancer by several months to several years. Most tumors are diagnosed within 4–6 months. It has been shown that several PNS are associated with onconeural antibodies. Unfortunately, almost 50% of patients with true PNS do not have any of the well-characterized onconeural antibodies. In these patients, early diagnosis of the tumor is frequently difficult, resulting in a significant delay in tumor treatment. To date, the best way of stabilizing a PNS is probably to treat the tumor as soon as possible. An early diagnosis of a neurological syndrome as PNS is thus crucial for the management of patients.

Detection of an onconeural antibody in a patient suspected to have a PNS is, at present, the most valuable diagnostic test.

CSF study-lymphocytic pleocytosis, elevated proteins, increased IgG synthesis, Oligoclonal bands.

MRI SCAN, FDG PET SCAN, CT SCAN, BIOPSY.

TREATMENT:

The two major principles for PNS management are treatment of the underlying cancer and immunotherapy initiation, both should be initiated as soon as possible to minimize irreversible neuronal loss and severe neurological disability.

IMMUNOTHERAPY- IV steroids, IV immunoglobulins, plasma exchange
Other- rituximab, azathioprine, cyclophosphamide.

Supportive therapy- analgesics, antiepileptics,3,4 diamino pyridine, physiotherapy.