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Paraneoplastic syndrome – An unusual presentation

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Abstract:

Paraneoplastic visual syndromes are rare group of disorders due to immune mediated pathogenesis. Paraneoplastic optic neuropathy is predominantly bilateral and present with progressive painless visual loss. In our case report, 63-year-old woman presents with sequential bilateral loss of vision and on investigation was found to have adenocarcinoma of the lung. Treatment with concomitant chemotherapy and immunotherapy resulted in significant stability in her vision. She had finger counting in the left and visual acuity for the right eye was 400/20. She also noted deranged color perception. Her neurological examination was unremarkable. The cranial nerves V1-V3 were intact, tongue was midline without any fasciculation. She did not had any facial asymmetry and Romberg's test was negative. Initial CT scan did not showed any intracranial pathology. On PET scan from June 2021 showed intensely avid 1.7 x 2.9 cm right upper lobe uptake with SUV of 7.

Keywords: Paraneoplastic usual case, Paraneoplastic visual syndromes, Paraneoplastic optic neuropath, chemotherapy and immunotherapy to treat visual paraneoplastic syndrome

1. Introduction:

Paraneoplastic syndrome is defined as signs and symptoms observed from cancer but having no direct correlation with the cancer tissue. Meaning that they are distant from the site of the real cancer. PNS result from the immune mediated cascade of events directed against tumor related antigen. The host detects tumor proteins as antigen to the body and directs antibody against them.

It has been reported that 7-15% of all cancers are associated with PNS.^{1,2} PNS is described in almost 10% of the lung cancers.¹ Usually, PNS is the first sign of the disease in majority of the patient's and almost 60% of all small cell lung cancer disease are already metastatic at the time of the diagnosis.³

2. Case report:

Sixty three-year-old woman, physically active complain of sudden loss of vision in the left eye on February of 2021. Ophthalmic examination revealed visual equity only for counting fingers in the left eye and visual acuity of 20/400 in the right. Investigations included funduscopy and CT head and brain did not reveal any abnormality. MRI from June 9, 2021 did not show any optic apart the compression or any venous thrombosis. CSF study was normal with normal pressures. CT chest abdomen pelvis from June 14, 2021 showed multiple ground-glass appearances bilaterally and right pleural base lesion measuring 16.7 mm x 9.3 mm in the right upper lobe.

A PET scan was done on June 2021 showed intensely avid 1.7 and 2.9 cm right upper lobe uptake with SUV of 7. Biopsy of the right upper lobe lesion showed invasive non-small lung cell cancer with strong immunoreactivity for TTF1 positivity. Chemotherapy was initiated in on August 2021 with cisplatin and gemcitabine. Immunotherapy, pembrolizumab was started from 2nd cycle onwards. Patient completed 4 cycles of combined chemo/immunotherapy on October 24, 2021 and is currently on maintenance immunotherapy.

CT scan from November 1, 2021 showed stable disease. She was prescribed prednisone 60 mg with the neurologist restarting the immunotherapy however she did not showed any improvement in her vision so far portion and more than 20 cycles and vision has improved slightly. Patient was extensively worked up however, paraneoplastic disease profile Plus was negative.

3. Discussion:

Usually, PNS is the first sign of the disease in the majority of the patient's and almost 60% of all small cell lung cancer diseases are already metastatic at the time of the diagnosis. Different antibodies have been isolated as a precursor of PNS.⁴ Antibodies against Recoverin, anti -alpha anolase, anti-transduction an anti-carbonic anhydrase are all implicated in the loss of vision, or sensitivity, ring scotomas visual field defects respectively.⁵ ⁶ Over the years numerous antibodies have been isolated causing these effects as a manifestation of the PN is spectrum ranging from decreased visual acuity to total blindness.

Early recognition of the PNS can be helpful in early detection and treatment of the cancer. There is strong correlation with specific antibodies and certain malignancies however, these can not be considered specific or sensitive marker for the malignancies.⁷

Painless, progressive visual loss are frequent complaints among patient with CAR and MAR antibodies. Endoscopic examination is initial step followed by imaging studies including CT/MRI PET scan. Other investigations include fluorescein angiogram and fundus photography. CSF analysis and immunohistochemical assay with retinal antibodies and western blots are part of investigation tools in such cases.

The main focus of the treatment is for underlying malignancy. Immune suppression is often employed using corticosteroid in an attempt to attenuate the autoimmune cascade. Use of intravenous immunoglobulins has shown mixed effect in improving visual symptoms secondary to PNS.^{8,9} There is no role of ocular surgery however the surgery for the tumor excisions were conducted treat underlying malignancy with variable results. Significant improvements are either anecdotal or in experimental settings, controlled human studies are yet to show any sustained improvement of visual symptoms by any means of treatment.

4. Conclusion:

Paraneoplastic ocular neuropathy has been documented in sporadic cases without association of the paraneoplastic neurologic syndrome. Usual presentation is painless visual loss which may be simultaneous or sequential. Investigation for occult malignancies should be undertaken as this may be their first manifestation. This patient showed benefit of the treatment with chemotherapy alone or with immunotherapy.

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