Optic Neuritis: A Rare Paraneoplastic Phenomenon of Hodgkin's Lymphoma

Yasir Khan 1, Waqas Khan 1, Nishanth Thalambedu 1, Ammar A. Ashfaq 1, Waqas Ullah 2

1. Internal Medicine, Abington Hospital - Jefferson Health, Abington, USA 2. Internal Medicine, Abington Hospital-Jefferson Health, Abington, USA

Corresponding author: Yasir Khan, yykhan@gmail.com
Disclosures can be found in Additional Information at the end of the article

Abstract

Hodgkin's lymphoma (HL) is a hematological disorder that has a high cure rate. It usually presents as asymptomatic lymphadenopathy or a mass on chest radiograph along with constitutional symptoms (‘B’ symptoms such as fever, night sweats, or unintended weight loss) in less than half the cases. Optic neuritis is a demyelinating condition that is rarely associated with HL. We present a case of HL that presented with optic neuritis as a paraneoplastic syndrome.

Introduction

Hodgkin's lymphoma (HL) is an uncommon disorder with a low incidence rate, comprising only 0.5% of all cancers that will be diagnosed in 2019. HL shows two peaks—the first one in the third decade and the second peak after age 50. It is considered a potentially curable disease with a five-year survival rate of almost 87%. HL is classified by the World Health Organization as nodular sclerosing, mixed cellularity, lymphocyte depleted, lymphocyte rich and nodular lymphocyte-predominant. Patients with HL usually present with asymptomatic lymphadenopathy, unexplained weight loss, and drenching night sweats.

Paraneoplastic syndrome is a group of symptoms that occur with increased frequency in cancer patients. These are not caused by metastasis, direct infiltration of the tumor, or known indirect mechanisms such as toxicity, ectopic secretion of hormones, or induced coagulopathies. Paraneoplastic syndrome is thought to be caused by an immune mechanism against the antigens that are normally present in the body.

Optic neuritis is an acute demyelinating condition of the optic nerve. It usually occurs in patients in multiple sclerosis but can present as an isolated neurologic finding. It typically presents with a sudden loss of vision, which can vary from a small defect in the field of vision to total loss of light perception followed by improvement over several months [1]. Rarely, it appears as a paraneoplastic phenomenon associated with small cell lung cancer (SCLC). We are presenting a case of a 21-year-old man whose optic neuritis was a paraneoplastic phenomenon of his underlying HL.

Case Presentation

A 21-year-old man with no significant past medical history was seen in his college student
health department with complaints of bifrontal headaches and blurred vision in the left eye. He
was noted to have 20/50 vision in his left eye. He denied any history of fever, chills or night
sweats but did report a weight loss of 6 lbs. over the past month. He was referred to the
emergency room (ER), and a magnetic resonance imaging (MRI) of his brain showed abnormal
elevated T2 signal involving the left optic nerve along the leftward aspect of the chiasm in the
left prechiasmatic and intracanalicular portions of the left optic nerve. He was treated with IV
methylprednisolone for three days, followed by oral prednisone taper. Whereas his headache
responded to the steroid treatment, his vision remained the same. An MRI of the cervical and
thoracic spine was obtained to see the lesions of multiple sclerosis. This MRI showed a normal
spinal cord with no findings consistent with multiple sclerosis. However, incidentally, we noted
bilateral level five cervical adenopathy measuring approximately 3 cm and mediastinal and
right hilar adenopathy also measuring up to 3 cm. A lumbar puncture was also done that
showed glucose of 79 mg/dl, protein of 29 mg/dl, and 2 UL white blood cell (WBC). The results
of his cytology examination were negative. A computed tomography (CT) of his neck and chest
revealed extensive bilateral adenopathy in the lower neck, supraclavicular areas, right
paratracheal region, right hilum, and an anterior mediastinal mass measuring 3.1 cm x 5.8 cm.

The patient was discharged from the hospital with an outpatient follow-up for a hematological
evaluation. His positron emission tomography (PET) scan showed increased tracer activity
associated with lymph nodes in the neck and chest, which was consistent with a clinical history
of lymphoma. However, no tracer activity was identified below the diaphragm. He then
underwent right cervical lymph node biopsy that showed large nodules encircled by fibrosis
with scattered classic and lacunar Reed–Sternberg cells. The patient continued to have blurred
vision that had remained the same in the initial outpatient follow-up. He underwent
echocardiogram and pulmonary function testing and was started on ABVD (adriamycin,
bleomycin, vinblastine, dacarbazine) chemotherapy with a plan for a PET scan after two cycles with a total
of four cycles with involved-field radiation therapy (IFRT) depending on the chemotherapy
response. Optic neuritis was a paraneoplastic phenomenon. The patient decided to receive
chemotherapy at a different hospital and did not follow up with our clinic. Upon calling the
patient later, he informed us that he was undergoing chemotherapy, and his symptoms of
blurred vision have resolved completely.

Discussion
Paraneoplastic syndromes are rare with lymphomas. The probability of paraneoplastic
syndrome associated with HL is even lower. Demyelinating neuropathies are found more
commonly with non–HL [2].

The presence of neurological pathology of unknown origin at the time tumor diagnosed can be
entirely coincidental and may be the result of two different but concurrent events. The
neurological symptoms are considered definite paraneoplastic syndrome if the patient has well-
characterized onconeural antibodies along with the path of the tumor. The paraneoplastic
syndrome can also be considered definite in the absence of onconeural antibodies if the
neurological symptoms improve after the treatment of the cancer [3].

Paraneoplastic optic neuritis has been reported but usually does not occur as an isolated
finding. It mostly happens in association with other neurological syndromes such as retinitis,
posterior cerebellar degeneration, or encephalomyelitis [4]. Optic neuritis associated with
malignancy has clinical and radiographic features indistinguishable from optic neuritis
associated with multiple sclerosis. Because of identical clinical manifestation generated by
both conditions, the exact determination of its etiology is critical for proper treatment of the
underlying disease. Occasionally, it presents as the initial finding of underlying cancer [5].
Paraneoplastic optic neuritis is most commonly associated with SCLC, but there have been
cases reporting it in the setting of non-SCLC, thymoma, renal, and thyroid cancer [6-8]. Many of these patients have antibodies to collapsin-responsive mediator protein-5 [9]. To the best of our knowledge, there have been no reports of optic neuritis occurring as paraneoplastic syndrome with HL.

Patients usually present with loss of vision that may be painless. This finding may be unilateral or bilateral. This may be associated with other neurological symptoms or may occur as an isolated finding. MRI is done to evaluate the cause of optic neuritis and may show enhancing and swollen optic nerve. Treatment of the underlying cancer either through surgical excision or chemotherapy usually improves the symptoms of optic neuritis.

Conclusions

Although rare, optic neuritis can present as paraneoplastic syndrome associated with HL. Recognition of this paraneoplastic syndrome can lead to early detection and treatment of cancer, which is a crucial initial step of management.

Additional Information

Disclosures

Human subjects: All authors have confirmed that this study did not involve human participants or tissue. Conflicts of interest: In compliance with the ICMJE uniform disclosure form, all authors declare the following: Payment/services info: All authors have declared that no financial support was received from any organization for the submitted work. Financial relationships: All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. Other relationships: All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

References