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Extranodal Non-Hodgkin's Lymphoma presenting as Cavernous sinus syndrome

Authors

Uthaya Sankar M.K¹, Naveen Sathish.V², Sibi Chakravarthy.C³

¹Professor, ^{2,3}Post Graduate Residents, Department of General Medicine Sri Manakula Vinayagar Medical College and Hospital, Puducherry-605107

Abstract

Non-Hodgkin's lymphoma (NHL) is a monoclonal proliferation of lymphoid cells of B cell (70%) and T cell (30%) origin. Primary lymphomas of uterine cervix are rare. Central nervous system involvement can occur as a late manifestation of NHL and may include mass lesions and meningeal infiltration. This is a rare case of uterine cervical non hodgkins lymphoma presented as cavernous sinus syndrome.

Introduction

Non Hodgkin's lymphomas are cancers of mature B, T, natural killer cells. It constituted 4% of all cancers and it is the ninth most common cause of cancer related death. The incidence is 1.5-2% per year. About 90% are B-cell origin. T-cell lymphomas are more common in India. The five year survival rate is 72%.¹

The clinical presentation can be asymptomatic or the presence of B symptoms such as fever, night sweats, unexplained weight loss. Diffuse large Bcell lymphoma is the most common type. Upto 40% patients will have involvement of non-lymph node sites.¹

Case Report

This is a case of a 32 year old female who was apparently healthy 20 days back and then developed toothache for which her lower premolar was removed due to presence of caries. Two days later she developed headache which was progressive, dull aching, diffuse, not associated with vomiting and seizure. Associated with pain over the cheek for the past 10 days with low grade intermittent fever for 6 days. She came to our hospital for her main complaint of inability to open and move her left eye for past 4 days. No history of eye swelling, blurring of vision, eye pain, discharge, redness, local injury, double vision. No history of similar complaints on her left eye. History of altered sensation over the face is present. No history of any other cranial nerve, motor, sensory abnormality. On examination positive findings were left eye partial ptosis with dilated and non reacting pupil with nil movements and reduced sensation over the V1 and V2 distribution on the face. Abdomen examination revealed non tender hepatomegaly. Rest of the examination was normal.

Blood investigations showed deranged urea and creatinine. USG abdomen was done in view of view of elevated RFT. USG abdomen and pelvis revealed hepatosplenomegaly, bulky uterus and cervix with enlarged right ovary, right adnexal

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10*9.7*9 cm mass. MRI brain showed Granulomatous lesion affecting the paranasal sinuses with cavernous sinus and meningeal involvement. Lumbar puncture was done which revealed elevated protein and cell count with lymphocyte predominance. CA 125 - 598 U/ml. MRI abdomen and pelvis showed features suggestive of carcinoma cervix with parametrial extension and extension into upper $2/3^{rd}$ of vagina and bilateral ovaries and kidneys with moderate ascites. Ascitic fluid analysis came positive for malignancy probably Non Hodgkin's lymphoma effusion. Biopsy taken from cervix was suggestive of NHL infiltration (Fig 1). FESS was planned and it showed aspergillus growth in the paranasal sinuses. Screening chest CT was done and there was no evidence of lymph node involvement. She was diagnosed as primary extranodal Non Hodgkin's Lymphoma of the cervix with involvement of ovaries, kidneys with tumor thrombus spreading to cavernous sinus and aspergillus infection of the paranasal sinuses.



Fig 1: Biopsy of cervix showing large atypical lymphoid cells with hyperchromatic condensed nuclei with inconspicuous nucleoli with scant cytoplasm showing fine vacuoles with lymphoglandular bodies in a proteinaceous background.

Discussion

The incidence of NHL, especially extra nodal lymphoma, has increased in recent decades.

Lymphoma only accounts for 0.3% of all cervical cancers, making it a rare malignancy.² One of the common reasons for delayed diagnosis is frequent absence of bleeding per vagina and negative cytology due to subepithelial location of the tumor.

Cavernous sinus involvement is rare in NHL, and as the initial presentation of NHL it is extremely rare. Involvement of the cavernous sinus can be the inaugural sign of lymphoma and neurologic signs may appear rapidly as in our patient.³ The high-grade malignant NHL in this case is rapid progressive and considerably invasive, but can occur with symptoms ranging from minimal at the presentation to even cavernous sinus syndrome. The diagnosis rests largely on imaging and biopsy results. It is associated with poor prognosis and Aggressive combined modality treatment appears to improve survival.

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