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Case Report

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Nasopharyngeal Carcinoma with Atypical Ophthalmic Presentation- A Rare Case Report

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Abstract

Introduction: Nasopharyngeal carcinoma (NPC), a rare malignancy with unique and complex etiology, is most confused and commonly misdiagnosed disease and can present as painless enlargement of upper neck nodes, followed by nasal, aural, ophthalmic, neurological manifestations.

Case presentation: We present a case of NPC in a 24 year old unmarried female, who initially presented with painless bilateral loss of vision, initially treated empirically for optic neuritis and Tuberculosis for four years.

Conclusions: Isolated painless and bilateral loss of vision though unusual and rare manifestation of NPC should be considered as a possible differential.

Key words: nasopharyngeal carcinoma, bilateral loss of vision, optic neuritis, paraneoplastic effect, ophthalmic presentation.

Introduction

Nasopharyngeal carcinoma (NPC) is a rare malignancy with unique and complex etiology. NPC is one of the most confusing, commonly misdiagnosed, and poorly understood diseases. NPC has a remarkable racial and geographical distribution [1, 2]. NPC has attracted world-wide attention because of complex interactions of genetic, viral, smoking, environmental, dietary factors, occupational etc., which have been implicated in causation of NPC [1-9].

The clinical presentations of NPC may sometimes be insidious and nonspecific. They are usually related to the local, regional and distant spread or metastasis of the lesion. They may include cervical lymphadenopathy, nasal blockage, epistaxis, hyponasal speech and otologic and neuro-ophthalmic manifestations [2-10]. The clinical morphology may be infiltrative, ulcerative or exophytic [10]. The neuro-ophthalmic manifestations present usually late with the advancement of disease. Ophthalmic manifestations of NPC include ocular motility problems (cranial nerve involvement), blurred vision, proptosis, orbital

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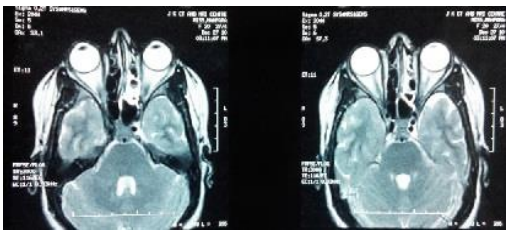


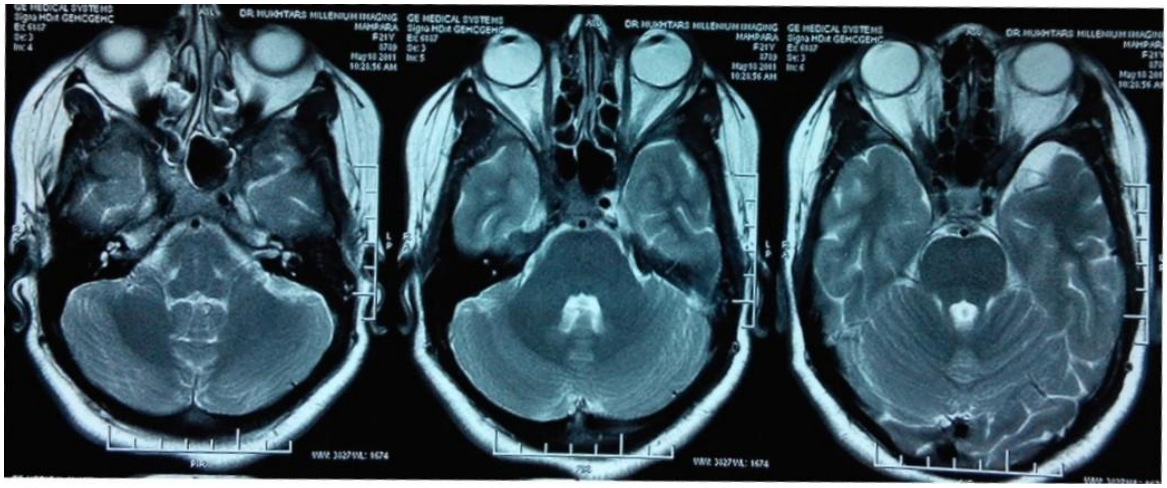
Figure 1- MRI Brain and Orbit showing normal study

pain, and optic disc edema [11]. Loss of vision as initial presentation is very rare and can be optic neuritis or/ as paraneoplastic manifestation of NPC [12-13].Here we present a case of NPC with atypical neuro-ophthalmic presentation misdiagnosed for four years.

Case Presentation

We present a 24 year old Asian female student presented with acute diminution of vision in both eyes 4 years back followed by complete loss of vision one year later in both eyes. There was no history of pain, redness in eyes, headache, blurring of vision, double vision, and inability to move eyeballs. Magnetic resonance imaging (MRI) of brain revealed normal study (Figure 1) and patient was started on intravenous methylprednisolone 1gram for three days followed by tapered dose of prednisolone but with no improvement in vision. Repeat MRI another six month later again revealed normal study (Figure 2) and patient continued on steroids and multivitamins without any benefit. Another MRI four months later showed localized enhancement of left optic nerve and cavernous sinus (Figure 3), followed by another MRI two months later which gave the impression of bilateral cavernous sinus infiltration and orbital apex syndrome (OAS) with left optic neuritis (Figure 4). The patient was continued on steroids. Another six months later MRI revealed peri-optic enhancing soft tissue involving optic nerve up to the optic chiasm, likely possibility of granulomatous etiology (likely Sarcoidosis, Wegener's Granulamatosis or Tuberculosis). Cerebrospinal fluid (CSF) analysis and chest roentgenogram (CXR) were normal. Rheumatoid factor (RF), anti-nuclear antibody (ANA), C - reactive protein (CRP) profile was negative. Ultrasonography (USG) evaluation of abdomen and pelvis was normal. She was put on anti-tubercular treatment for six months in view of MRI showing granulomatous lesion (likely tubercular) in bilateral optic canal. Nasopharyngeal biopsy revealed nasopharyngeal granular lesion.

Contrast enhanced (CE) MRI another six month later gave the impression of bilateral bulky cavernous sinus with diffuse enhancing cavernaldural pachymeningeal thickening seen extending upto orbital apex, features suggestive of infective etiology likely tubercular pachymeningitis. Repeat biopsy nasopharynx was suggestive of sarcoma possibly rhabdomyosarcoma and was started on



- Figure 2- MRI Brain and Orbit showing Normal Study

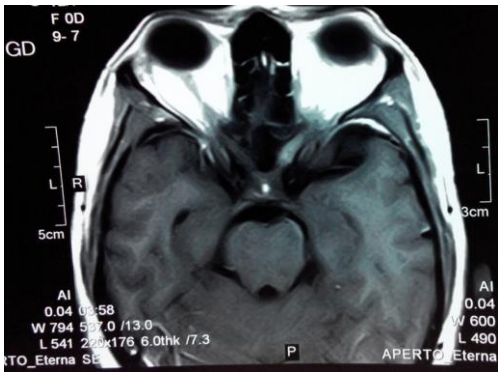


Figure 3 MRI showing localized enhancement of left optic nerve (ON) and cavernous sinus (CS)

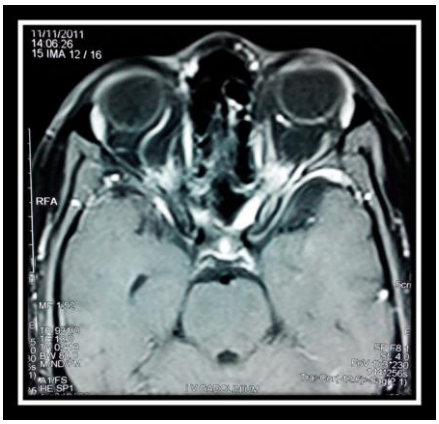


Figure 4- MRI showing Bilateral cavernous sinus (CS) infiltration and orbital apex syndrome (OAS) with left optic neuritis (ON)

adriamycin and ifosfamide based chemotherapy. Positron emission tomography – computed tomography (PET-CT) using fluorodeoxyglucose (FDG) radiotracer, revealed FDG avid thickening in nasopharynx, with mild FDG avid sclerotic metastasis involving clivus, cervical vertebrae and upper thoracic vertebrae. Non-FDG avid meningeal enhancement in cavernous sinus extending into bilateral orbital apices. Repeat nasopharyngeal biopsy revealed tumor composed of malignant epithelial cells. Immune histochemistry (IHC) was cyto keratin positive and leukocyte common antigen (LCA) and CD profile for lymphoma was negative confirming the diagnosis of NPC. Patient was advised but refused to undergo EBV DNA levels in view of terminal illness. She is presently on palliating treatment.

Discussion

The NPC is a rare malignancy worldwide, the most confusing, commonly misdiagnosed, and poorly understood disease [2]. The ophthalmic manifestation as only or the first symptom is very rare in NPC at presentation [14]. Similarly the isolated optic nerve involvement is rare [14] but usually indicates cavernous sinus invasion [8, 9, 14]. The ophthalmic involvement though not uncommon in patients with nasopharyngeal carcinoma in the late stages of disease [13] but isolated optic nerve involvement is rarely reported as an initial manifestation [13, 15]. The patients with the orbital or ocular manifestations also have lesser survival and poor prognosis [11]. The sudden onset of painless monocular blindness can be the initial manifestation of a NPC [16], but in our case, patient had acute and painless bilateral loss of vision.

The optic neuritis with sectorial field loss as a remote effect of NPC likely paraneoplastic effect as there was neither any tumor mass around the optic nerve nor any histological evidence of tumor infiltration [12].

The most common pathway of NPC spread as orbital invasion are through pterygopalatine fossa and inferior orbital fissure, followed ethmoid and/or sphenoid sinuses as can be seen best by Coronal sections on CT or MRI [5,7,9,11,17]. NPC can invade orbital apex and can present as OAS and the causes can be inflammatory, infectious, neoplastic, iatrogenic/ traumatic, or vascular processes [18, 19].

Conclusions

The present case description emphasizes that acute painless bilateral loss of vision can be a rare and unusual initial manifestation of NPC. The present report suggests that NPC can present as retrobulbar/optic neuritis with initial negative radiological findings most likely due to its paraneoplastic effect. In order to

reduce the delay in diagnosis, the patients presenting with acute painless loss of vision should be ruled out for NPC.

Ethical Considerations

Written informed consent was obtained from the patient for the publication of this case report and accompanying images.

Conflict of Interest

The authors declare no conflict of interest.

Author's contributions

SQW prepared and approved the manuscript, TK approved final manuscript, SYW also helped in writing manuscript, AH, and TRM, MML, and FA did the literature search and prepared the draft manuscript.

All authors read and approved the final manuscript.

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