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Case Report

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Non-AIDS related multiple brain and orbital lymphoma mimicking Meningioma: A Case Report

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Abstract

Non-AIDS lymphoma, a type of primary central nervous system (CNS) lymphoma is an uncommon aggressive infiltrative malignant tumour involving several sites in the central nervous system such as the periventricular region and leptomeninges. In this article, the authors presented a 26-year old man with painless progressive right exophthalmos and scalp swelling with no symptoms and signs of intracranial hypertension and hyperthyroidism. Magnetic resonance imaging (MRI) done revealed isointense masses with homogenous brilliant enhancement on contrast administration resembling a meningioma, with a dura tail – like attachment as seen in meningioma. He had surgery for the right orbital tumour and histopathological diagnosis confirmed our suspicion of lymphoma (B type). Steroid was given in the post-operative period which led to significant regression of the tumours, hence its description as 'vanishing tumour'. He is presently receiving a methotrexate-based chemotherapy and subsequently planned for radiotherapy.

Keyword: Lymphoma, Orbital, Meningioma

Introduction

Primary central nervous lymphoma (PCNSL) is an uncommon tumour, highly infiltrating aggressive extranodal non-Hodgkin's lymphoma mainly confined to the craniospinal axis (the brain, the spinal cord, leptomeninges) and the eyes [1]. Ocular involvement occurs in 10-20%, [1,2]. Non-AIDS central nervous system lymphoma is a type of PCNSL. PCNSL represents about 2-6% of primary CNS tumour and 1-2% of all non-Hodgkin's lymphoma [3].

It is often diagnosed in the 5th-6th decade of life with median age at 60 – 65 years [4], but lower in immunocompromised patients. It tends to be multiple in about 20-40% of NON–AIDS related patients at the time of presentation [5], PCNSL is seen in both immunosuppressed (due to HIV, organ transplant, and congenital immunodeficiency syndromes) and immunocompetent patients but causes and behaviour differs [6]. The diffuse large B-cell lymphoma is the commonest histopathological subtype which is seen in about 90% of patients, T- cell lymphoma accounts for about 2% of PCNSL in western world, and about 8% in Japan South-east Asia. The exact cause is unknown but there is direct linkage with Ebstein Barr virus infection [7]. Common neurological features include headaches in some cases, focal neurological deficit, seizure, personality changes, intracranial hypertension and ocular symptoms such as progressive exophthalmos and visual deterioration. The Non–AIDS PCNSL imaging features on brain magnetic resonance imaging (MRI) or computed tomography (CT) scan reveals mainly a solitary or in some instances multiple supratentorial lesion in close contact with meninges or ventricle, deeply seated in the parenchyma, periventricular white matter deep gray nuclei, corpus callosum and superficially adjacent to CSF spaces. Atypical locations include the pineal gland, pituitary gland, brainstem, and cavernous sinuses [3].

On CT scan, it appears hyperdense on non-contrast and on MRI iso-hypotense signal on both T1WI & T2WI and shows homogenous contrast enhancement

with minimal to moderate perilesional oedema. This characteristic finding allude to the fact that it is also described as '' pseudomeningioma'' by some authors and could pose diagnostic challenges [8,9,10]. Brain tumour biopsy (stereotactic guidance) remains the gold standard of diagnosis but surgical resection does not improve outcome [2]. Methotrexate based chemotherapy remain the mainstay of treatment. This is also used in conjunction with radiotherapy as the tumour is highly chemo- and radiosensitive [11].

Corticotherapy has a role with rapid tumour regression within 2weeks of administration and this is useful in those with significant mass effect, hence the nickname the vanishing tumour or ghost cell or disappearing tumour [12].

Case Report

A 26-year old male Painter who resides in Souissi Rabat with no previous significant health challenges presented with 11-months history of painless progressive right exophthalmos and painless progressive right parietal scalp swelling. No symptoms suggestive of hyperthyroidism, intracranial hypertension and loss of consciousness. No history suggestive of retroviral diseases and bone malignancies in the past

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Examination findings at presentation showed a conscious man with intact higher mental and long tract functions. Eye findings revealed right eye exophthalmos grade IV, non-pulsatile, non –tender, immobile with chemosis. There was associated 3rd cranial nerve opthalmoplegia involving levator palpebrae superioris and medial rectus muscle and with no light perception. The left eye was essentially normal with no evidence of papilloedema. Retroviral screening was negative. Brain MRI (T1, T2 with FLAIR) done revealed findings as below in Figure 1.

He was operated for the right orbital tumour. A right frontolateral incision made, had right fronto-orbital craniotomy, periorbita stripped, medial rectus muscle opened and right optic nerve identified, fibrous pale pink tumour excised from the orbital cavity, routine haemostasis secured and muscle apposed, bone replaced and scalp closed in layers. Frozen section done during the intraoperative period revealed non-hodgkin's lymphoma which was confirmed with the definitive histopathological analysis as non-hodgkin's lymphoma (type B).

Post –operatively, patient was placed on high dose methyl prednisone for two weeks and almost all the tumour disappeared as revealed on the Brain CT scan done 2week after steroid use, hence its nickname- the ghost cell tumour. The patient is being presently worked up for platinum-based chemotherapy and radiotherapy.

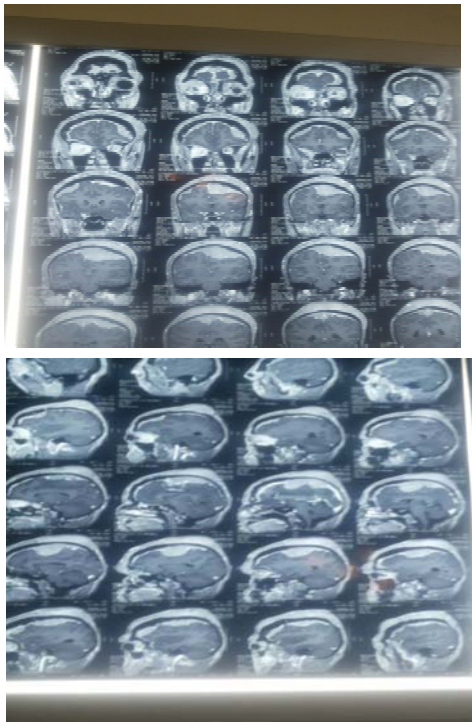

Figure 1: Multiple isointense lesions in the right orbit and left parietal parasagittal region on T1WI and T2WI and homogenous enhancement on contrast administration and minimal oedema on FLAIR

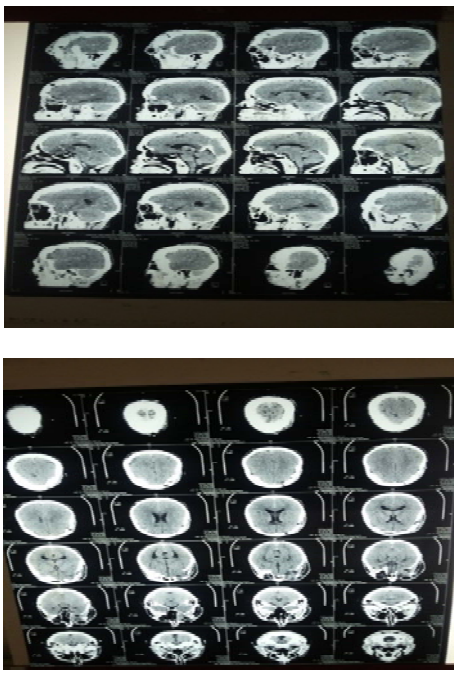

Figure 2: Brain CT scan of the above patient with evidence of vanished tumour

Discussion

The age of the patient is far less than the median age of the disease in non-AIDS CNS lymphoma as

reported in several literatures [1]. The usual age at presentation for patients with AIDS is lower with a mean age of 31-36 years when compared to immunocompetent patients. [13].

Some studies from South-east Asia countries such as Japan and Korea have shown no significant association of PCNSL with AIDS [16, 14, 15]. A recent study estimated that in USA about 13% of CNS lymphoma cases were associated with AIDS in the period 2001- 2007. A significant reduction is evident since about 48% of CNS lymphoma cases were associated with AIDS over a period ranging from 1990-1995 [16]. Despites this finding in the USA, studies from Japan and Korea have not shown any significant association of PCNSL with AIDS in the past or in the present [16, 17].

The clinical features of PCNSL in immunocompetent patient include focal deficits, neuropsychiatric symptoms, headache, nausea, vomiting suggestive of raised intracranial pressure, seizures and ocular symptoms. Patients with AIDS related PCNSL are more likely to present with mental status changes or seizures (and Magnetic Resonance Imaging (MRI) and CT scan usually show single or multiple, periventricular, homogeneously enhancing lesions. These lesions are predominantly supratentorial in location. Ring like enhancement is rarely seen in immunocompetent patients, but noticed frequently found in immunocompromised patients [10, 18].

In this index case presentation, multiple extra-axial lesions were seen on MRI are isointense on T1WI and T2WI which show homogenous enhancement on gadolinium administration hence the initial diagnosis of multiple meningioma prior to surgical intervention [20]. PCNSL simulate a variety of lesions like malignant gliomas, metastasis, demyelinating disorders, abscess and other infections. Toxoplasmosis is an important differential in immunocompromised patients. CT and MRI scan generally provide results suggestive but not conclusive for PCNSL [19]. Majority of the cases are of B-cell phenotype. The occurrence of T-cell PCNSL is very rare in Western as well as Asian studies [15, 17]. A study done in japan of patients with PCNSL between 2000-2004 showed that T-cell PCNSL accounted for 1.7% of all cases [14], whereas a similar study done earlier between 1985-1994 from Japan showed a higher figure of 8.5% [16] From this case presented, the lymphoma is of the B subtype and this is in line with majority of the lesions from several studies discussed above. PCNSL is both a highly radiosensitive and chemosensitive infiltrative tumour. Combination of chemotherapeutic agents such as cis-platinum and vincristine have been noticed to be very useful, as PCNSL is often multifocal with a propensity to involve the subarachnoid space. Stereotactic biopsy is the preferred procedure for obtaining tissue for pathological diagnosis. However, for this patient under review, surgical intervention was aimed at both diagnosis and cytoreduction thereby reducing tumour burden. Therefore, an intraoperative diagnosis is necessary before proceeding with surgical resection if PCNSL is suspected, as was done for this patient. The use of frozen section (FS) of CNS neoplastic lesions is usually accurate. However, cytologic preparations are often superior to FS in demonstrating the morphology of malignant lymphoid cells. PCNSL usually consists of a central area densely packed with tumour cells. The surrounding brain tissue shows gliosis, inflammation and scattered tumour cells hence the site of the biopsy, especially stereotactic biopsy is important. Difficulties can arise when the central hypercellular zone is not sampled adequately. The presence of reactive astrocytes can sometimes raise the possibility of a glial tumor or an inflammatory lesion.

This index patient was placed on high dose steroid after surgery for two weeks and tumour was noticed to have significantly reduced almost disappearing. The role of steroids in the treatment of PCNSL have been reviewed in several literatures.

Steroids are generally not administered until the diagnostic in suspected cases of PCNSL is confirmed. Previous reports have shown that apart from their therapeutic anti-oedematous effects, they can cause rapid lympho-depletion, most especially useful in patients whose tumour produces mass effect. They can also produce reactive gliosis with a variable infiltration of B- or T lymphocytes and macrophages, thus obscuring the diagnosis in some cases. This is a major limitation to use of steroids pre-operatively. However, a recent study at the Mayo clinic showed that corticosteroid administration before biopsy did not significantly affect the histopathological diagnosis of PCNSL cases [21]. Similarly, steroid administration did not significantly affect the intraoperative diagnosis/ final diagnosis in the present study.

Conclusion

Non-AIDS CNS lymphoma, a relatively uncommon infiltrating extranodal non-hodgkin's (mainly type B) lymphoma which may be solitary or multiple located in varied location in the CNS may present with varied clinical manifestation such as features of intracranial hypertension and eye signs. It is occasionally mistaken for meningioma due to its homogenous enhancement on gadolinium administration. Operative intervention is mainly for diagnostic purpose. The tumour is both chemo sensitive and radiosensitive which when given post-operatively, improves the quality of life of the patient. Steroids do have a role but could create diagnostic difficulties if giving pre-operatively due to the disappearing nature of the tumour.

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List of abbreviations

CNS, central nervous system; PCNSL, Primary central nervous lymphoma; CT, computer tomography; MRI, magnetic resonance imaging

Declarations

Ethics approval and consent to participate

Not provided

Consent for publication

Not applicable.

Availability of data and materials

The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

Competing interests

No conflict of interest associated with this work.

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Contribution of Authors

We declare that this work was done by the authors named in this article and all liabilities pertaining to claims relating to the content of this article will be borne by the authors. All authors contributed equally for this work.

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