October 2010

Primary chiasmal sarcoid granuloma masquerading as glioma of the optic chiasm

Muhammad Zubair Tahir
Aga Khan University

Farheen Shaikh
Aga Khan University

Arshad Ali Siddiqui
Jinnah Postgraduate Medical Centre

CASE REPORT

Primary Chiasmal Sarcoid Granuloma Masquerading as Glioma of The Optic Chiasm

Muhammad Zubair Tahir[1], Farheen Shaikh[2] and Arshad Ali Siddiqui[3]

ABSTRACT

A 37-year-old woman presented with a 6 months history of headaches and memory impairment. Examination showed no neurological deficit with normal vision. MRI scans showed an enlarged optic chiasm. There was no dural or leptomeningeal enhancement or hydrocephalus. Open biopsy of the suprasellar mass showed non-caseating chronic granulomatous inflammation compatible with sarcoidosis. Systemic features of sarcoid were absent. Patient showed marked improvement on steroid therapy.

Key words: *Optic chiasm. Sarcoidosis. Magnetic resonance imaging. Steroid.*

INTRODUCTION

Neurosarcoidosis is a rare disease, seen in adults often with the evidence of systemic disease. Its clinical spectrum includes a diverse range of conditions including meningitis, cranial nerve palsies, epilepsy, hydrocephalus and transverse myelitis.[1,2] MRI is a useful radiological investigation for its evaluation active lesions show contrast enhancement.

Despite this, the MRI appearances of neurosarcoidosis are seldom distinctive and it may be mistaken for other diseases.[3,4]

We report a rare case of primary sarcoid granuloma of optic chiasm masquerading as chiasmal glioma.

CASE REPORT

A 37-year-old lady of Yemeni origin presented with headaches and memory impairment over the last 6 months. She denied any visual or other constitutional symptoms. Her neurological examination including visual acuity, cranial nerves and fundoscopy was unremarkable. There was no adenopathy. Chest radiograph, basic haematological investigations were normal. Her endocrine workup showed that serum T3 level of 0.05 n mol/L, T4 level of 4.2 microgm/dl, TSH level of 4.28 micro IU/ml, growth hormone level of 0.05 ng/ml, serum cortisol level of 0.89 micrograms/dl and serum prolactin level of 143 ng/ml. All these values were below normal and suggestive of hypopituitarism. Her serum angiotensin converting enzyme (ACE) level was marginally high at 28 U/L.

She underwent MRI scan of brain which showed an enlarged chiasma with swelling extending to the inferior part of the hypothalamus. The mass had a low signal on both T1 and T2 weighted images and showed strong contrast enhancement. No dural or leptomeningeal enhancement or hydrocephalus was present (Figures 1 and 2).

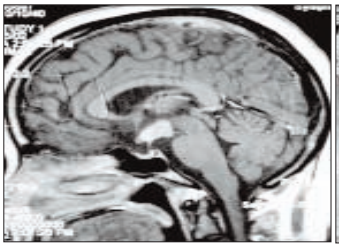

Figure 1: Postcontrast T1-weighted image shows sagittal section of the enlarged optic chiasm. The homogenous contrast enhancement is exclusively within the chiasma.

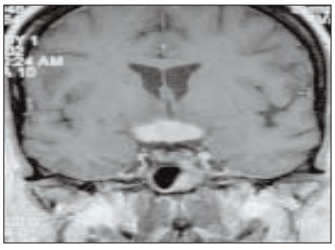

Figure 2: Postcontrast T-1 weighted (coronal section) shows enhancement of the chiasm, suggestive of an intrinsic mass lesion.

The patient underwent open biopsy of the suprasellar mass. Frozen section specimens revealed a granulomatous lesion. Histopathology showed multiple non-caseating granuloma and inflammatory cell infiltrate with perivascular cuffing but no acid-fast bacilli, a diagnosis of sarcoidosis was made. The patient was commenced on oral steroids 5 mg of prednisolone for 6 weeks. She showed a definite slow progressive improvement and discharged from hospital on the 14th postoperative day. Patient was advised to follow-up in her country of origin with her referring physician and have a follow-up MR imaging at least after 3 months from date of surgery. Unfortunately, she lost follow-up and repeat MR study could not be documented.

DISCUSSION

Neurosarcoidosis, accounts for approximately 5% of all cases of sarcoidosis.[5,6] Of these, have reported that

[1] *Department of Neurosurgery[1]/Opthalmology[2], The Aga Khan University Hospital, Karachi.*

[3] *Department of Neurosurgery, Jinnah Postgraduate Medical Centre, Karachi.*

***Correspondence:** Dr. Arshad Ali Siddiqui, Neurosurgeri Klinik, Hospital Sultanah Aminah, Jalan Abu Bakar, Johar Bahru-80100, Johor-Malayisa..*
E-mail: doc4brains@yahoo.com

Received January 30, 2010; accepted May 17, 2010.

48% initially present with neurological symptoms.[6] Ultimately majority of the patients develop other systemic manifestations of the disease.[6] Typically, sarcoid presents in other organs such as eyes, skin, lymph nodes, bones, lungs and parotid gland. When it is found in the nervous system it seems to affect cranial nerves, promote a basal arachnoiditis or meningo-encephalitis, or organize in posterior fossa nodules.[7] Histologically distinct, non-caseating granulomas with lymphocytic infilterates are found.[8] Radiological diagnosis of this entity is often indistinguishable from lymphoma, metastasis or primary malignant disease.[9]

Enlargement of optic chiasmal is commonly result of optic nerve glioma. It accounts for 2% gliomas in adults, and 7% in childern.[10] They occur commonly in females and up to 25% of cases are associated with neuro-fibromatosis. Histologically there are infiltrations of the optic pathways with glioblasts of various sizes and occasional astrocytes. Hyperplasia of the overlying arachnoid matter, known as arachnoid gliomatosis, is a well-recognized feature. Chiasmal tumours are generally not treated surgically except for biopsy, CSF shunting or to remove the rare exophytic component to try and improve vision. Chemotherapy or radiotherapy is used for chiasmal tumours especially in younger patients.[10]

Neurosarcoidosis affecting the optic chiasm may closely resemble chiasmal glioma. Chiasmal enlargement on MRI should suggest a diagnosis of sarcoidosis even in the presence of mass lesion and absence of systemic features. This case shows one of the many faces of neurosarcoidosis, which are almost impossible to diagnose correctly without open biopsy.

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