Non-Hodgkin's Lymphoma of the Brainstem with Atrial Septal Mass


Abstract

Simultaneous occurrence of cardiac and central nervous system tumors have been rarely reported. A 23 years male presented to us with right cerebello-pontine (CP) angle symptoms and signs. Cranial imaging showed a mass lesion in the right pons infiltrating into the right and middle cerebellar peduncles. There was also a cardiac - atrial septal mass. The brainstem lesion was found to be a non-Hodgkin's lymphoma where as the cardiac lesion was not accessible. Central nervous system lymphomas are reported rarely and the prognosis is poor. The chances of the cardiac lesion in this patient also being a lymphoma are high, as it was an infiltrative cardiac mass, infiltrating the atrial septum.

INTRODUCTION

The association of cardiac and central nervous system tumors are rare and infrequent. These include sarcoma, myxoma, lymphoma, ectodermal tumours, secondaries etc. We came across a young male presenting with cerebral and atrial septal masses. The cerebral lesion was biopsied and found to be non-Hodgkin’s lymphoma (NHL) but the cardiac lesion was not accessible.

CASE REPORT

Twenty three years healthy male individual, developed insidiously right facial numbness two months ago which progressed slowly. He noticed defective hearing in the right ear and facial deviation to left side with inability to close the right eye since 20 days. He felt unsteady and swayed to right side on walking and noticed double vision on looking to the right side over the past 10 days. All the symptoms were slowly progressive. His vision was clear on closing one eye. There was no tinnitus, earache or ear discharge. He could swallow well both, solids and liquids. His limbs were of normal power and there was no numbness anywhere. Bladder and bowel were normal. He did not have headache, vomiting, seizures, or loss of consciousness at any time.

No recent or past history of prolonged fever, cough, loss of weight, exertional dyspnea, arthralgia or sore throat. There was no history of dogbite, rashes or vaccination. There was no history of DM, HT, PT, STD, alcohol or tobacco intake or substance abuse. Patient was a cloth weaver and had exposure to dyes. He could work well until recently without fatigue.

On examination

The patient was a young, thin individual, without anemia, jaundice, cyanosis, clubbing or lymphadenopathy. JVP was normal. Afebrile, pulse rate of 80/min, regularly irregular. His blood pressure was 110/80 mm of Hg, both supine and standing, in RUL. There was no significant variation of BP in other limbs. Vision was normal, with normal fields and fundii. There was no ptosis. Pupils were 2.5mm equal, reacting well to light. Extra-ocular movements were full but he complained of double vision on looking to extreme right. There was horizontal jerky gaze evoked 1st degree nystagmus on looking to right. Touch and pin prick were diminished on right side of the face corresponding to ophthalmic and maxillary divisions and slightly decreased over mandibular area. There was gross wasting of right temporalis and masseter, with weakness on clenching the teeth.

There was a LMN type of right facial palsy and sensorineural type of hearing loss on right side. The pharyngeal sensation and gag reflex were diminished on the right side with mild left palatal weakness. There was furrowing of both sides of tongue but no weakness. He had mild left hemiparesis with grade 4 power both in upper and lower limbs with mild hypertension. Deep tendon reflexes were sluggish in upper limbs, normal in lower limbs. Plantars flexor. Superficial reflexes were normal. Pin prick was diminished on right side below D6. Beevor’s negative and joint position sense was normal in all limbs. Head and spine were normal. Stance was normal, but he swayed to right side on walking and tandem walking was not possible.

Abdomen and lungs were normal. There was short systolic
mass lesion involving the anterior left atrium and adjoining superior, posterior right atrium.

**Treatment**

It was decided to operate on the posterior fossa mass and to establish the pathological diagnosis and to proceed with appropriate treatment. As regards the cardiac mass, resection was not possible and hence it was decided that biopsy could be done at a specialized cardiac center at a later date. The patient underwent a right suboccipital craniectomy under GA. There were problems of AV dissociation, bradycardia and hypotension during the surgery. However with required medication, during surgery and post-operatively there were no complications. Only partial excision of the intracranial tumour could be done.

**Histopathology**

The histopathological examination (Fig. 4) showed Non-Hodgkin’s Lymphoma (NHL) - large cell type infiltrating the brain. The tumour cells showed angiocentricity, active mitosis and focal necrosis. As the diagnosis was confirmed with this immunohistochemistry was not done.

The patient remained as he was in the post-operative period. It was decided to give him Chemo and Radiotherapy and to tackle the cardiac mass after this.

**DISCUSSION**

This young male presented with symptoms of a right CP angle mass lesion. He was found to have an infiltrative pontocerebellar mass and also a cardiac mass. The intracranial mass turned out to be a lymphoma. We do not have the pathology of the cardiac mass due to technical reasons. There was no evidence for lymphoma elsewhere in the body. Hence it may be assumed to be a primary CNS lymphoma.

The Histopathological examination revealed non-Hodgkin’s lymphoma of large cell type. Most of the central nervous system lymphomas are of T-cell in origin.1

According to the National Cancer Institute of USA, non-Hodgkin’s lymphoma is divided into three subtypes namely the small non-cleaved cell, lymphoblastic and large cell types.2 The most frequent primary sites are the abdomen (31.4%), the head and neck region (29%), and the mediastinum (26%) . The non-cervical lymph node involvement is usually 6.5% while skin, thyroid epidural space and bone accounts for the
reminder (7%). NHL of the brain are rare, representing less than 2% of all brain neoplasm. They present as single or multiple lesions involving the eye, leptomeninges or brain parenchyma. Parenchymal lesions are often adjacent to the ventricles or ependymal surface and 10-20% are infratentorial localization, primarily in the brainstem (3%).

First generation chemotherapy regimens used successfully in systemic NHL were ineffective in central nervous system lymphomas, in large part due to the existence of blood-brain barrier. Whole brain radiotherapy results in high response rates more than 90% of cases, but this treatment is associated with high relapses and delayed neurotoxicity. Methotrexate based chemotherapy is associated with fewer toxic effects. The combination of chemotherapy following radiotherapy has improved the survival upto 17.6 months or double the median survival time. However the recurrence is high upto 90% within 1 year, making the prognosis poor.

It can be that the patient also has a cardiac lymphoma. However this is very rare.

Cardiac lymphomas are usually made out at autopsy.

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References