WEBER SYNDROME SECONDARY TO TUBERCULOMA MIMICKING A VASCULAR INFARCT.

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ABSTRACT

Background: Central nervous system tuberculosis is rare but carries a high mortality and morbidity. We report a case of tuberculoma mimicking a vascular infarct presenting as Weber Syndrome.

Materials and Methods: Case report

Result: A 46-year-old lady with presented with history of drooping of the left eye and left sided weakness of the body. She had history of sudden onset of loss of consciousness two weeks before. On examination, patient had complete ptosis of the left eye with dilated pupil and right sided hemiparesis. Patient also had altered memory and disorientated. Visual acuity in both eyes was 6/6. Both anterior segments and fundus were normal. An urgent CT scan of brain was performed and showed a left thalamus and external capsule infarct. Subsequently MRI of brain revealed post contrast ring enhancement in the left thalamus and midbrain. A magnetic resonance spectroscopy (MRS) showed elevated lipid peak at the site of the lesion. ESR was elevated 74mm/hr. Mantoux test was positive 17mm. CSF profile was normal. A diagnosis of cerebral tuberculosis with Weber Syndrome was made. Patient was started on anti-tuberculosis treatment. She was also started on oral prednisolon for one month. Patient showed improvement in her cognitive functions. Her left eye ptosis and extra-ocular movements also improved after 3 months. Patient was continued to be on anti-tuberculosis treatment for one year.

Conclusion: Intracranial tuberculosis remains to be a diagnostic challenge to physician. MRI and MRS play an important role as a non-invasive tool for diagnosing supported by other investigation.

Keywords: Tuberculoma, intracranial tuberculosis, Weber Syndrome
1.0 Introduction

Tuberculosis is a disease that is caused by Mycobacterium Tuberculosis. This granulomatous infection is still becoming an important communicable disease in developing countries and carries high morbidity and mortality. The prevalence of tuberculosis cases in 2008 is estimated to be 11.1 million worldwide which is equivalent to 164 cases per 100 000 populations (Phyper, 2006). Central nervous system tuberculosis can be classified on a basis of clinical features, CSF findings, radiological or pathological investigations into tuberculous meningitis, intracranial tuberculomas, and tubercular abscess Intracranial tuberculosis can also mimicked other conditions such as cerebrovascular accident and intracranial tumour. Our objective were to report a case of intracranial tuberculosis which mimics a vascular infarct; Weber Syndrome which is 3rd cranial nerve palsy with contralateral hemiparesis.

2.0 Materials and Methods

Case Report.

3.0 Case Report

A 46-year-old Malay lady was referred by general practitioner for diplopia and right hemiparesis. Patient was apparently well until the day of presentation where she had sudden onset of giddiness and loss of consciousness for about 30 minutes. The incidence occurred while she was accompanying students for camping at the lake side. She was brought to a nearby hospital. Upon regaining her consciousness she was started to have diplopia and right sided body weakness. It was noted that her blood pressure was 176/111 mmHg with high blood sugar at of 11 mmol/L and she was treated as hypertensive emergency. An urgent CT scan of brain was performed and showed ill-defined hypodense lesion at the left thalamus and left external capsule. There was no intracranial bleeding or thrombosis of the cavernous sinus. Patient was suspected to have left thalamus and external capsule infarct. She was admitted for optimisation of her blood pressure and discharged home after 3 days after while still having residual right hemiparesis.

The right sided body weaknesses were getting worse over the course of few days as she was unable to walk properly and holding for support. She was also noted to have slurred speech and gait imbalances. Family members noticed that the patient having memory impairment and become disorientated. However, there was no headache, nausea or vomiting.

There was however no history of prolonged cough, fever, night sweat and loss of weight or appetite. There was no positive history of tuberculosis (TB) contact. It was the first episode of diplopia and haemiparesis and she had never experience any episode of transient ischaemic attack previously. She was then referred to neuro-ophthalmology team in our centre for further evaluation due to worsening of her conditions.
On general examinations patient was lethargic and disoriented. The speech was slurred. She had complete ptosis of the left eye with fixed and dilated pupil. The left eye was in outward and downward position with restricted movement in all directions more markedly in adduction. The right eye movement was slightly restricted in upward gaze only. Visual acuity in both eyes however was good. Both anterior segments examinations were unremarkable.

Fundus examinations were normal for both eyes. Her blood pressure on presentation was 145/85 mmHg and her pulse rate was 82 per min.

Peripheral nervous system examinations revealed muscle power was reduced over the right upper limb (power of 4/5) and lower limb (power of 4/5).

ESR was elevated 74mm/hr with positive Mantoux test reading of 17mm. Chest radiography was normal. Lumbar puncture was performed and the CSF profile was normal.

Subsequently MRI of brain was performed. There was hypointense lesion in the left thalamus and midbrain on T1 and slightly hyperintense in T2 with high signal on DWI but low signal in ADC. There was post contrast a ring enhancement in the left thalamus and midbrain. A magnetic resonance spectroscopy (MRS) showed elevated lipid peak at the site of the lesion, highly suggestive of tuberculoma. A diagnosis of intracranial tuberculosis with Weber Syndrome was made.

The patient was started on anti-TB regime. The intensive phase of 2 month with maintenance phase of the anti-TB regime to complete one year. Oral prednisolon 50mg daily was also given for one months durations over tapering dose.

On follow-up after one month, patient showed some improvement. She was able to walk with help and able to move the hand. The left ptosis getting better although the lid was still covering her visual axis. The extraocular movement also showed some improvement. MRI brain was performed and showed smaller size of the lesion.

On follow-up at 3, 6 and 12 months, the left ptosis and extra-ocular movement in both eyes improved. However, the left hemiparesis still persist with power of 3/5 and patient needed walking aid to ambulate. She completed her anti-tb treatment for one year and had no worsening of her condition. Unfortunately she had to undergo medical board due to unfit to continue with her work as a teacher.
Figure 1: Limited extra-ocular movement with partial residual ptosis over the left eye at 3 months post starting anti-tuberculous drugs. (No pre-treatment photos available).

Figure 2: MRI showing ring enhancement post contrast with MRS showing lipid peak of non-tumoral origin.

4.0 Discussion

Central nervous system tuberculosis has a highly devastating outcome and accounted for approximately 1% of all the tuberculosis cases (Phyper, 2006). It carries a high mortality as well as neurological morbidity partly contributed by delayed of diagnosis and treatment.
Tuberculoma accounts for 10%–30% of intracranial masses in TB endemic areas (Modi, 2013).

Clinical manifestations of tuberculoma depend largely on their location. Patients often present with headache, seizures, papilloedema, or other signs of increased intracranial pressure. Our patient presented with sudden onset of diplopia, right sided hemiparesis, confusion and disorientation, which mimicked a common cerebrovascular accident. The extra-ocular movement more markedly affect the left eye with the eye in an outward and downward position in primary gaze (typical features suggestive of III cranial nerve palsy although the IV and VI were also involved. Coexisting contra-lateral (right) hemiparesis initially suggested that patient having Weber Syndrome, which commonly due to vascular infarct involving the mid brain area. Restriction of extra-ocular movement of the right eye also present although there was no deviation in primary gaze. Lack of fever, loss of appetite and weight as well as no history of contact with tuberculosis patient further deviate the initial attention towards infective causes.

General processes that produced the subsequent neurological pathology were believed to be secondary to obliterative vasculitis, adhesion formation and encephalitis or myelitis (Dastur, 1995) with characteristic vasculitis of the blood vessels traversing the exudates. Intraluminal caseation necrosis causes cellular changes which leads to infarct. There is some evidence that early in the course of the disease vasospasm may mediate strokes and later strokes is due to proliferative intimal disease (Lammie, 2009).

Intracranial tuberculosis remain to be a challenging diagnosis. It may not have positive cerebral spinal fluid examination due to small number of bacilli available in the fluid. Identification of bacilli in CSF through smear and culture methods is still the most important and most widely available means to diagnose CNS tuberculosis (Rock, 2008). In our patient, the lumbar puncture revealed an acellular smear. Therefore, in current situations, we have to turn to other option for diagnosing intracranial tuberculosis that include radiological investigations involving the CT Scan and MRI along with other blood investigation (ESR) and Mantoux test. Significantly high ESR and positive mantoux test highly suggestive of tuberculosis.

Tuberculomas may have variable appearance on CT scan. It could appear as both hypo or hyperdense lesion on CT scan. In few case series, tuberculoma appeared as a ring-enhancing lesion with a hypodense centre post contrast, which was actually a central calcification. This is known as the ‘target sign’. Some study concluded that target sign is pathognomonic of an intracranial tuberculoma while other study concluded that it is non specific sign (Van Dyk, 1998; Shah, 2000; De Castro, 1995; Kazner, 1978). The initial CT scan done in our patient showed ill defined hypodense lesion at left thalamus and external capsule. However, no contrast study had been done initially as the lesion and presentation mimic a cerebrovascular infarct.

MRI appearance of intracranial tuberculomas is usually more specific than the CT appearance. With regards to its superior soft tissue contrast, MRI features of tuberculoma depend on the lesion whether it is non-caseating, caseating with a solid centre, or caseating with a liquid centre (Shah, 2000). However, in MRI it will still show a ring enhancement post contrast (Kazner, 1978).
In this case, although the MRI findings help to localise the site of the lesion, we were still left out with several differential diagnosis as the ring enhancement lesion may also be due to infections, infarction or malignancy. In the case of infarct, there is not much can be done apart from controlling the risk factors. Malignancy also pretty much unlikely in this case as the patient has been well previously and had no sign and symptoms suggestive of the condition.

However, in the case of infection, there are several possibilities but high on the list with regards to this part of the world, tuberculosis is still very common. In order to aid in diagnosis, we did a MRS study (Magnetic Resonance Spectroscopy). The metabolites level is of non tumoral lesion. The lipid peak is elevated and highly suggestive of tuberculosis. One of the metabolic marker for Mycobacterium tuberculosis is the productions of lipids, (Garg, 1999; Jinkin, 1995), in which may be detected by MRS if produced in significant amount (Gupta, 2001; Mishra, 2004; Semlali, 2008). Santy (2011), reported that Proton magnetic resonance spectroscopy (1H-MRS) enables to clearly discriminate tuberculous from other infectious cerebral abscesses in toddlers.

The lacunar infarct, that was present in our patient may be part of the disease itself whereby stroke can occurs in 45% of patients with Tuberculous meningitis either in early or later stage, mostly in basal ganglia region, and is a predictor for poor outcome at 3 months (Kalitha, 2009). Although it is difficult to say that meningitis occur in this patient due to lack of signs, however, at times the patient appear to be having cognitive impairment and irritable, this suggests the possibilities of some degree of encephalopathy also takes place.

To date, there were no randomized controlled trial that has established an optimal regimen for Central Nervous System Tuberculosis because of its relative rarity and difficulty to come to an early diagnosis. Most guidelines suggest a 2-month intensive therapy regimen of isoniazid, rifampicin, pyrazinamide, and ethambutol (EHRZ), followed by 10 months of continuation phase of isoniazid and rifampicin (Rock, 2008; Thwaites, 2009). Our patient completed her one year course of anti-tuberculosis but there were still residual neurological deficits on her. There were no further worsening of her symptoms.

Although no randomized controlled trial of steroids has been performed for patients with tuberculoma, most experts recommend co-administration of steroids with anti-tuberculous therapy (Hernandez, 2000; Naidoo, 2005; Jain, 2005). Usage of corticosteroids is widely accepted as adjuvant therapy for CNS TB, especially for TB meningitis. A meta-analysis of 7 randomized controlled trials concluded that corticosteroids should be used routinely in HIV-negative people with tuberculous meningitis to reduce death and disabling residual neurological deficit among survivors (Prasad, 2008). In our patient, she was started on oral steroid of 1 mg per kilogram of body weight over a slowly tapering dose.

5.0 Conclusion and recommendation

Intracranial tuberculosis remains to be a diagnostic challenge to physician. Intracranial biopsy is an invasive procedure to attain the diagnosis. MRI and MRS play an important role as a non-invasive tool for diagnosing when supported with blood investigations, CSF studies and...
Mantoux test. High index of suspicion needed to come to a diagnosis in this case as the initial presentation mimicking a cerebrovascular accident.

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Declaration

The authors declare that the above has not been published or submitted for publication in any other journal. There is no conflict of interest on this article.

Authors contribution

The 1st, 2nd and 4rd author involved directly in the management of the patient. The 1st author prepared the manuscripts with supervision of 2nd and 4rd author. The 3rd author provide consultation with regards to the imaging studies

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