CASE REPORT

A rare case of alcoholic pellagra encephalopathy with startle myoclonus and marked response to niacin therapy: time for a new dictum?

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SUMMARY

We report a case of 56-year-old man, chronic alcoholic, presented to us with progressive weakness in all the four limbs with stiffness and gait disturbance since 1-year associated with cognitive impairment. On examination he had mild confusion, spastic quadriparesis with brisk reflexes, extensor plantars and cerebellar features. During the hospital stay myoclonus was noticed in the patient, which was startle in nature. He did not have dermatitis, ascites or any stigmata of liver failure. MRI of brain revealed bilateral subdural effusion, left focal subarachnoid haemorrhage at perisylvian area and diffuse cortical atrophy. He was treated with supportive measures including thiamine with which his condition worsened. His serum niacin was low. With a possibility of alcoholic pellagra encephalopathy (APE) the patient was treated with niacin. His clinical condition improved drastically over next 1 week and startle myoclonus disappeared, favouring the diagnosis of APE though multiple confounding factors were present.

BACKGROUND

Alcoholic pellagra encephalopathy (APE) can manifest as nervousness, insomnia and hallucinations commonly found in chronic alcoholics and can include confusion, oppositional hypertonus, myoclonus, ataxia and thiamine supplementation in these patients might aggravate or trigger the development of APE. APE may manifest without dermatological manifestations of pellagra. Niacin estimation should be considered in all alcoholics, malnourished patients presenting with encephalopathy, myoclonus and ataxia as niacin supplement can reverse this condition and prevent fatal outcome, if untreated.

CASE PRESENTATION

A 60-year-old man, a heavy alcoholic for more than 30 years, presented with history of progressive weakness in all the four limbs since 1-year and unsteadiness of gait since 6 months. He also had memory disturbances, apathy and emotional lability. There was no history of fever, diarrhoea, seizures or jerky movements and falls.

On examination he had confusion. Cranial nerve examination showed nystagmus. Motor examination revealed spastic weakness with power of 4/5 in all the four limbs. He had cog wheel rigidity at both the wrist joints. His deep tendon reflexes were brisk with ankle clonus and extensor plantars

bilaterally. Cerebellar features were present in the form of finger–nose incoordination.

INVESTIGATIONS

His haemoglobin was 9.8 mg/dl. Serum calcium was 6.3 mEq/dl but rest of the biochemistry including electrolytes, renal and liver function tests were normal. Serum parathormone and ammonia levels were normal. Ultrasound of the abdomen showed fatty liver. His head CT and brain MRI revealed bilateral old residual subdural effusions with focal subarachnoid haemorrhage at left perisylvian area (figure 1) without any evidence of corpus callosum demyelination, pontine hyperintensities or mammillary body atrophy/haemorrhages.

His EEG showed diffuse slowing, thyroid profile was normal and anti-TPO antibodies were negative. During hospital stay he developed a new symptom in the form of myoclonic jerks which were infrequent to begin with and occurred on startle. He was supplemented with thiamine and calcium. However, his condition worsened after thiamine supplementation as he became drowsy with increased frequency of startle myoclonus. He was shifted to ICU and CSF study was done, which was a normal procedure.

Figure 1 MRI of brain showing bilateral old residual subdural effusions with focal subarachnoid haemorrhage at left perisylvian area.

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DIFFERENTIAL DIAGNOSES

- ▶ Wernicke's encephalopathy
- ▶ Marchiafava-Bignami disease
- ▶ Central pontine myelinosis
- ▶ Hashimoto's encephalopathy
- ▶ Creutzfeldt-Jakob disease (CJD)

TREATMENT

Serum niacin level estimation was conducted, which was low. The patient was diagnosed with APE and treated with niacin (300 mg/day) through a nasogastric tube.

OUTCOME AND FOLLOW-UP

He showed dramatic improvement over next 3–4 days, myoclonic jerks reduced in frequency and eventually disappeared. Weakness and ataxia improved over next 1 month.

DISCUSSION

Pellagra, first described by Gaspar Casal,¹ is a nutritional disease owing to niacin deficiency characterised clinically by four Ds: diarrhoea, dermatitis, dementia and death.² It is related mainly to malnutrition, chronic alcoholism, those subsisting mainly on niacin-deficient diets like jowar.^{1–3} APE can manifest as nervousness, insomnia and hallucinations.^{1–3} APE includes oppositional hypertonus, ataxia and thiamine supplementation might aggravate or trigger the development of APE.⁴ Rarely they can have Babinski sign and startle myoclonus.⁵ Central chromatolysis of neurons in pontine nuclei and Clarke's column of the spinal cord are characteristic of APE.⁶

The term 'pellagra' is derived from an Italian word meaning rough skin⁵ and caused by niacin deficiency. The neurological syndrome of pellagra is not well-defined and frequently confounded by other coexisting central nervous system disorders; especially in patients with alcoholic pellagra. The primary early symptoms are neuropsychiatric and may progress to stupor and coma.⁵

APE can present as a rapidly progressive dementia syndrome with myoclonic movements and ataxia resembling CJD.⁵ The cases of APE are rare and there are only few case reports. Though Serdaru *et al*⁴ described the clinical spectrum of APE in 22 cases, it was a retrospective postmortem study which showed characteristic chromatolysis of neurons suggestive of neurological pellagra.

APE can present with other alcoholic encephalopathies in combinations like Wernicke's disease.⁶ Serdaru *et al*⁴ described APE associated in 13 cases with Marchiafava-Bignami disease and/or Wernicke-Korsakoff disease. Park *et al*⁶ reported a case of APE with mental deterioration, myoclonic jerks and external ophthalmoplegia a feature of Wernicke's disease.

Isolated cases of pellagra encephalopathy been rarely reported. Prakash *et al*¹ reported a case with rare presentation of delusional parasitosis with pellagra whose delusion resolved after niacin therapy. Pitsavas *et al*³ reported a case of APE who was administered vitamin tablets that did not contain niacin presented with myoclonus, insomnia and recovered after treatment with niacin. Sakai *et al*⁷ reported a case with myoclonus and

ataxia in an alcoholic patient with cirrhosis who showed improvement after niacin therapy.

Our patient also had extrapyramidal features (rigidity) which has not been reported so far to the best of our knowledge. Symptoms of APE may worsen after thiamine supplementation. In 9 of 22 cases, Serdaru *et al*⁴ found that there was aggravation of symptoms after administration of thiamine.

In conclusion, it should be remembered that the neurological syndrome of APE is not well-defined and is frequently confounded by other coexisting factors. APE is an underestimated and under diagnosed clinical entity and one should keep high index of clinical suspicion in any alcoholic patient who presents with unexplained progressive encephalopathy, ataxia and myoclonus even other features of pellagra are not found. Though it is a dictum that one should supplement thiamine prior to the administration of dextrose in alcoholics, we like to stress that one should supplement niacin in all alcoholic patients before administration of thiamine to prevent the development or precipitation of APE as these patients may worsen with thiamine.

Learning points

- ▶ Alcoholic pellagra encephalopathy (APE) is underestimated clinical entity and confounded by multiple coexisting factors.
- ▶ APE can manifest rarely with ataxia and myoclonus mimicking various neurological conditions.
- ▶ APE can manifest even without other features of pellagra like dermatitis and diarrhoea.
- ▶ Though thiamine is empirically given to all alcoholic patients to prevent Wernicke's encephalopathy, one should be aware that it can precipitate APE.
- ▶ Niacin should be administered in all alcoholic patients with encephalopathy particularly if he is having myoclonus, ataxia and pyramidal signs and not improving with thiamine.

Competing interests None.

Patient consent Obtained.

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