Pregnancy should raise suspicion of improper placement, transmigration or expulsion of the IUD [5]. If the lost IUD is not found in the placenta and membranes at the time of delivery, imaging of the abdomen and pelvis must be done to locate it. When intraperitoneal, laparoscopic removal of the IUD is considered the first choice of therapy [1]. However, open surgery may be necessary in some [6].

A young lady with MELAS syndrome: a sporadic case

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Introduction

Mitochondrial myopathy Encephalomyopathy Lactic Acidosis and Stroke-like syndrome (MELAS) is a rare neurodegenerative disease, characterized by recurrent stroke-like episodes, lactic acidosis, bilateral occipitoparietal infarction and basal ganglion calcification. Only a few cases have been documented worldwide.

Case report

A 30 year-old lady presented with acute severe headache, tinnitus, blurred vision and weakness followed by two episodes of generalized tonic clonic fits. Symptoms worsened over a period of 48 hours. Her first presentation with a similar illness was two years ago and since then she has hearing impairment.

On examination, she was unconscious with grade III weakness in all four limbs, exaggerated deep tendon reflexes and right extensor plantar response.

Her random blood sugar, full blood count, arterial blood gas analysis and cerebrospinal fluid analysis were normal. Urine full report showed proteinuria (++) and no cells. Blood culture and urine culture were negative. ESR was 110 mm for the first hour but ANA and thrombophilic screening including lupus anticoagulant, anticardiolipin, antibodies IgG and IgM were negative.
CT scan of the brain showed bilateral basal ganglion calcification (Figure 1) and diffuse cerebral infarction involving occipito-parietal regions (Figure 2). The ECG showed Wolff-Parkinson-White Syndrome (WPW) type B but chest X-Ray and echocardiography were normal. Tests for HIV were negative.

**Figure 1 -** CT scan of the brain showing calcification of basal ganglia

**Figure 2 -** CT scan of the brain showing diffuse cerebral infarction involving occipito-parietal regions

A tentative diagnosis of meningoencephalitis was made and she was commenced on Aciclovir and IV ceftriaxone but there was no improvement. Serum lactic acid concentration was 110.4 mg/dL (Normal 4.5–19.8). CPK was normal. Muscle biopsy showed dysmorphic muscle fibers under the light microscope. Audiometry showed neurosensory deafness. She achieved a delayed partial recovery over a period of three months, but was dementic and mute most of the time. She was discharged on niacin and vitamin B₃.

**Discussion**

MELAS is a rare progressive multisystem disease, presenting either sporadically or among members of affected maternal pedigree. In USA no estimates of MELAS mutation are available. However, it is 10.2 per 100,000 in adult Finnish population and 1 per 13000 in Northern England. It has high morbidity and mortality. No sex predilection exists. About 80% of patients have a heteroplasmic A to G point mutation in Dihydouridine molecules and it is detected in mitochondrial DNA [1, 2]. Metabolic stroke-like episodes may be vascular or parenchymal due to a transient dysfunction of oxidative phosphorylation and consequently increased free radicals, vasoconstriction and decreased oxygen availability [3].

The myopathy manifests with weakness and easy fatigability. Encephalopathy may progress to dementia and apathy. The stroke-like episodes are the hallmark of the disease—associated initially with vomiting, headache and seizures and later with hemiplegia. The visual complaints are due to either ophthalmoplegia or blindness which may occur as a result of optic atrophy, lesions in the area of the brain concerned or retinal pigmentation. Diabetes mellitus is a common manifestation [4]. The dilated or hypertrophic cardiomyopathy and WPW may be present in some patients. Peripheral neuropathy, neurosensory deafness, gait ataxia, psychiatric disorder and neuroradiological features such as cerebral infarction, cerebral atrophy and calcification of basal ganglia have also been reported in some cases [3, 5].

The clinical diagnosis of MELAS is based on the presence of the following 3 major features;

1. Stroke-like episodes, typically before 40 years of age.
2. Encephalopathy with seizures or dementia.
3. Mitochondrial myopathy, evidenced by lactic acidosis or ragged red fibers and the presence of two of the following features;
   - Normal early psychomotor development.
   - Recurrent headaches.
   - Recurrent vomiting [5].
The muscle biopsy under H&E stain shows the changes due to myopathy. Ragged red fibers are the hallmark of MELAS, but it needs a special stain - Brilliant Red. The electron microscopy shows increase in number and size of mitochondria [3, 5].

Management is conservative. The vitamin supplements like Co enzyme Q10, Vit K1 and K2, riboflavin and niacin have been suggested to support respiratory chain enzymes in mitochondria, but no therapy is of proven efficacy. The genetic screening and counselling should be done at least in the first degree relatives [3, 5, 6].

This case highlights the need to be aware of uncommon conditions that can have a common clinical presentation.

References

Type I polyglandular syndrome patient presenting with metabolic encephalopathy

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Introduction
When two or more endocrine glands and other non endocrine immune disorders are present, the polyglandular autoimmune (PGA) syndromes should be considered. There are two major types of polyglandular autoimmune syndromes [1].

Type I polyglandular failure is characterised primarily by adrenal insufficiency, mucocutaneous candidiasis, hypoparathyroidism, and an array of other endocrine and non-endocrine disorders which occur with variable frequency [2].

PGA type I requires two of the three primary components for diagnosis. At the onset only one organ may be involved, but the number increase with time so that patients eventually manifest two to five components of the syndrome [1, 2].

Type II polyglandular failure (Schmidt syndrome) shares the high prevalence of adrenal insufficiency, but differs from the type I syndrome in that common features include autoimmune thyroid disease and insulin dependent diabetes mellitus. The onset of type II PGA is usually later than in type I, occurring primarily in adults [2].