Hydatidiform mole with clinical and biochemical evidence of hyperthyroidism.

Sir,

Hyperthyroidism is one of the non-neoplastic complications of hydatidiform mole. We present here a case with clinical and biochemical hyperthyroidism which resolved soon after evacuation of the mole.

A 19 year old Saudi female gravida, para 1 presented at the emergency room with a 2 day history of slight vaginal bleeding after amenorrhea for three months. There were no other relevant symptoms. Her periods were regular. On examination, she had a stern, steering look, her pulse rate was 132 beats per minute and her blood pressure was 140/80 mm Hg with a temperature of 37.2 degrees centigrade. The thyroid gland was not enlarged. Abdominal examination revealed a uterus that was about 20 weeks gestational size. No fetal heart sound was heard with doppler ultrasound. The cervical os was closed on pelvic examination. Ultrasound scan revealed a uterus without any fetal echoes but typical snow storm appearance of hydatidiform mole. There were bilateral ovarian cysts which measured about 5.5 cm diameter each. She was taken to the operating room where under general anesthesia, suction evacuation was performed with syntocinon infusion intravenously. Initial laboratory results were as follows: hemoglobin of 9.2 gm/dl, white blood cell count of 6,300/mm3, thyroid stimulating hormone (TSH) < 0.1 mIU/L, (normal range = 0.2-6.0), free T4 (FT4) > 101 pmol/L, (normal range = 11.7-26.0), free T3 (FT3) 38 pmol/L, (normal range = 4.3-7.6). The serum B-HCG was > 1000000 mIU/mL. Histology confirmed hydatidiform mole with trophoblastic hyperplasia. The chest x-ray was normal. She was seen weekly at the out patient clinic where she reported good health. There was no clinical evidence of hyperthyroidism and at each visit a serum B-HCG was requested. At the fourth visit, the thyroid function test was back to normal levels while the serum B-HCG was 100.8 mIU/mL. By the seventh visit, the B-HCG was 2.4 mIU/mL.

One wonders about the source of increased thyroid hormone activity in patients with trophoblastic hyperthyroidism. It has however been shown that HCG is a very weak thyrotropin and therefore is capable of stimulating the thyroid when its concentration is increased. This is in keeping with the finding that the hyperthyroid syndrome occurs in patients with very elevated levels of HCG although, occasional discordance between elevated thyroid levels and the clinical states of the patient with molar pregnancy do occur. The treatment of trophoblastic hyperthyroidism associated with hydatidiform mole is emergency evacuation preceded by supportive cardiovascular measures.

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Schwartz - Jampel syndrome in two siblings.

Sirs,

Schwartz Jampel syndrome (SJS) is a rare disorder. An association of congenital blepharophimosis and myopathy was first described by Schwartz and Jampel. Affected patients, usually children, have narrow palpebral fissures (blepharophimosis/blepharospasm), micro-gnathia, typical facial appearance, generalised muscle hypertrophy and stiffness, percussion and electromyographical myotonia, joint contractures, skeletal abnormalities and short stature. There may be associated myopia and cataract in some patients. Most of the patients have the above typical features with onset in early infancy, however cases have been seen in 2nd to 5th decade. Mild forms of the disease and absent percussion and electromyographical myotonia have been reported. SJS children have normal intelligence though 20% cases have been reported to be mentally retarded. The symptoms are usually not progressive. Myotonic stiffness produces feeding and mobility difficulties. Some therapeutic benefit has been reported with phenytoin sodium and procainamide. Carbamazepine was found most effective in a recent report. The condition is usually autosomal recessive though a few cases of dominant pattern have been reported. Recently locus to chromosome 1 34-36.1 by homozygosity mapping has been confirmed.

We report SJ syndrome in two siblings. Generalized hypertrophy and stiffness of muscles, blepharophimosis, typical myotonic facial appearance and short stature were seen in the living child. There were no bony changes. Percussion and electromyographical myotonia was absent. In addition the child had generalized areflexia. Carbamazepine improved the myotonia and the stiffness.

A 17 month old female child presented with generalized stiffness and weakness of about one year’s duration. She was 5th by birth order, born to consanguineous parents and developed normally until 6 months of age. The mother then noted initial stiffness in the lower limbs, gradually increasing over time. The upper limbs became involved 4 months previously. About a month prior to hospitalization the child was noted to have difficulty in sucking, closing her eyes and unusual facial features. There was no intellectual involvement. The child recognized her mother at 3 months and started speaking Ma-Ma, Ba-Ba at 11 months of age. She was initially able to sit at 6-7 months of age and later with the onset of symptoms could neither