Introduction

Opsoclonus-myoclonus is a rare neurological disorder characterised by intention tremor, unsteady gait, myoclonus and opsoclonus. Lethargy and irritability are also described. The abnormal eye movements are distinctive, being multidirectional and of high amplitude. Its recognition helps in the diagnosis of this clinical syndrome, the correct identification of which leads to the detection of an underlying neuroblastoma or other disorders.

Case report

An 18-month old, previously healthy girl presented with progressively worsening unsteady gait of 6 weeks’ duration. There was no preceding viral infection. On examination she was irritable, tremulous and unable to stand or walk without help. She had a staggering gait, continuous myoclonus of neck and limbs, and difficulty in speaking and swallowing. She had rapid, irregular, oscillatory eye movements. Cranial nerves, optic fundi, muscle power, tone, reflexes and sensation were normal.

Blood pressure was within the normal range for her age and the rest of the physical examination, including abdomen, was normal. Her growth curve showed flattening over the past 3 months.

Investigations with EEG and CT scan of the brain earlier had not shown any abnormality. A calcified suprarenal mass (2.9 x 2.5 x 2.8 cm) encasing the inferior vena cava on the right side, and enlarged para-aortic lymph nodes were found on ultrasound examination and were confirmed by computerised axial tomography (Figure 1). Bone marrow examination was normal.

The abdominal tumour and lymph nodes were surgically excised. Histopathological diagnosis was a ganglioneuroblastoma arising from the sympathetic chain and secondary deposits in lymph nodes. Surgery and chemotherapy were followed by marked regression of the neurological symptoms.

Discussion

The syndrome of opsoclonus, myoclonus, ataxia and encephalopathy has been termed “Kinsbourne syndrome”, “dancing eyes and dancing feet syndrome” and “opsoclonus-myoclonus syndrome” [1]. When they occur separately, opsoclonus and myoclonus have many causes, but when in combination, the cause is either post-viral (50%), or associated with an underlying tumour (50%).

Epstein Barr, Coxsackie B, mumps, polio and other enteroviruses are among the virus infections documented to precede this syndrome [2]. However, the search for a tumour should not be prevented by evidence of a viral infection. In children the syndrome usually presents in the first 2 years. Both sexes are equally affected. Tumours that cause opsoclonus-myoclonus are those arising from “neural crest cells”, i.e. neuroblastoma or ganglioneuroblastoma [3, 4].

An autoimmune basis is the likely explanation for this syndrome, because the brain and tumours of neural crest origin share the same embryonic forerunners, and have similar antigenic properties. The brain is therefore, “an innocent bystander caught in the cross-fires” between the immune system and the tumour. Facilities to test for anti-Hu antibodies in the serum were unavailable to us [5].

Removal of the tumour does not always result in complete regression of the neurological symptoms. Other treatment options include ACTH or steroid therapy, intravenous human immunoglobulin therapy, or azathioprine. Better survival rates have been reported in children who manifest opsoclonus-myoclonus [6, 7]. Awareness of this uncommon clinical entity is important for early recognition and prognosis of neuroblastoma, the commonest extracranial solid tumour of childhood.

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References


Pregnancy after excision of a yolk sac tumour of the ovary followed by chemotherapy

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(Index words: Chemotherapy, histopathology, surgery)

Introduction

A malignant ovarian tumour was excised and a course of chemotherapy given. Within 20 months of treatment, conception was achieved.

Case report

A 26-year old woman presented with a 2-day history of severe abdominal pain, distension, breathlessness and fever. Clinical examination revealed a large tender abdominal mass arising from the pelvis.

She was previously treated for subfertility with three consecutive cycles of clomiphene citrate. An ultrasound scan of the abdomen showed a large solid ovarian mass with a few cystic areas. Urine for beta-HCG was negative. The alpha fetoprotein level was over 350 000 ng/mL. A chest xray showed small pleural effusions on both sides.

Laparotomy revealed a large friable fungating mass, 15 x 15 cm, rupturing its capsule at many places, and a moderate amount of ascites. This mass originated from the left ovary. The left fallopian tube could not be visualised as it was entangled in the mass. The entire mass was excised, conserving the right ovary and the tube, which appeared normal. No adhesions were found. A sample of ascitic fluid was sent for cytology. No lymph node involvement was detected. The liver appeared unaffected. A sample of omentum was also sent for histology. The patient’s postoperative recovery was uneventful and she was discharged from hospital after 3 days.

The pathologist reported a germ cell tumour, composed of vacuolated network of embryonal cells, hyaline globules and Schillar-Duval bodies. The appearance was that of a yolk sac tumour of the ovary. Omentum was congested and infiltrated with chronic inflammatory cells with no malignant cells present. Ascitic fluid contained inflammatory mesothelial cells. No malignant cells were present.

The patient was subsequently referred to an oncologist, who arranged for a CT scan of the whole abdomen. This showed normal abdominal viscera and the remaining right ovary showed a cyst within. Alpha fetoprotein following surgery had declined from 350 000 ng/mL to 5 ng/mL.

After confirming the diagnosis, a course of chemotherapy consisting of five cycles of intravenous infusions of cisplatin, etoposide and bleomycin was instituted. The patient tolerated chemotherapy well. Serum alpha fetoprotein was further monitored. A subsequent ultrasound scan reported a normal uterus and the right ovary 20 x 28 x 38 mm with multiple small follicles. She was discharged from the hospital and told to expect normal menses in a few months. Regular menstrual cycles resumed 6 months after chemotherapy. She was advised to take 5 mg of folic acid daily, anticipating conception.

She conceived 20 months after chemotherapy and delivered by caesarean section a healthy baby girl near term.

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