several other hormones. Measurement of the plasma concentrations of regulatory peptides in patients with both neurofibromatosis and pheochromocytoma may identify those with duodenal carcinoids and throw light on the functional capacity of these tumours.

This association of tumours appears to be important in the clinical management of patients. We believe that in any patient with neurofibromatosis or pheochromocytoma who also has diarrhoea, diabetes, or cholelithiasis the possibility of a duodenal carcinoid that produces somatostatin should be considered. Obstructive jaundice in a similar patient could also be caused by duodenal carcinoid, either owing to its obstructing the ampulla or as a result of cholelithiasis related to secretion of somatostatin.

Clearly, more information is needed about this rarely reported combination of tumours; it is probably genetically determined, but in only one of the cases of von Recklinghausen's disease was a family history obtained, and there was no evidence that any of the duodenal carcinoids or pheochromocytomas was inherited. We hope, however, that this preliminary report will lead to the discovery of more cases of this interesting syndrome so that its true importance may be assessed. There appear to be three broadly separate neuroendocrine tumour syndromes—namely, multiple mucosal neuromas with medullary carcinoma and pheochromocytoma (multiple endocrine neoplasia type IIb), neurofibromatosis with pheochromocytoma and duodenal carcinoid, and the von Hippel-Lindau syndrome with pheochromocytoma and islet cell tumours. If these latter two entities can be clearly established it may be possible to regard them as multiple endocrine neoplasia types IIIa and IIIb respectively.

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SHORT REPORTS

Case of ectopic pregnancy after postcoital contraception with ethinylloestradiol-levonorgestrel

A combination of 0.1 mg ethinylloestradiol and 0.5 mg levonorgestrel (two tablets of Eugynon 50 or Ovran) given within 72 hours after exposure to the risk of pregnancy and repeated 12 hours later has a contraceptive success rate of around 98%.¹ As this so called Yuzpe regimen is thought to act primarily at the uterine level rather than to inhibit ovulation an ectopic pregnancy is more likely when the treatment fails, especially if there is pre-existing tubal damage. There is also the possibility of an effect on tubal function which could contribute to the risk of ectopic pregnancy.

We report a case of tubal pregnancy after the Yuzpe regimen.

Case history

A 34 year old para 0+1 presented requesting postcoital treatment 34 hours after a single episode of unprotected intercourse on day 12 of her cycle. She had omitted using her cap on that occasion. She gave a history of salpingitis at the age of 20 and a termination of pregnancy at the age of 24. She was given a counselling leaflet and was counselled by an experienced family planning doctor. Vaginal examination showed no abnormality. She was treated according to the Yuzpe regimen and did not experience nausea or vomiting.

At follow up four weeks later she reported an episode of vaginal bleeding which was light but similar to her normal period, lasting for eight days. This had started 12 days after the postcoital treatment. Four days after finishing her "period" (24 days after treatment) she had started bleeding again and experienced severe lower abdominal pain, for which she attended a casualty department at 4 am. Although she mentioned the postcoital treatment, vaginal examination showed no mass or tenderness and she was discharged with no specific arrangement for follow up.

The pelvic pain persisted, and when seen by one of us (JG) 32 days after the original treatment, and 44 days after her last normal period, there was a tender left adnexal mass about 3 cm diameter. At laparoscopy a left tubal pregnancy was diagnosed. The whole tube was distended with a haematosalpinx complicating a pre-existing hydrosalpinx. There was a small haemoperitoneum. The right tube and ovary were normal. A left salpingectomy was done, and tubal pregnancy was confirmed histologically. The patient made an uneventful recovery.

Comment

Ectopic pregnancies have been reported after stilboestrol² and diethylstilboestrol.³ We also know of a case after ethinylloestradiol that occurred in Holland in 1974. Morris and Van Wagenen⁴ reviewed 9000 cases of postcoital oestrogen contraception (high dose, five day courses) and found 29 reported pregnancies: three were ectopic. Though in two of these inadequate doses of oestrogens had been given, the risk of an ectopic pregnancy was calculated as 10%. Yuzpe argues⁵ that the high risk of ectopic gestation quoted is misleading, as postcoital contraception appears to reduce the number of

intrauterine pregnancies but has little or no effect on the incidence of ectopic pregnancies, and therefore the total number of such pregnancies observed is roughly equal to what would be expected if no contraception had been used.

In the past three years we have treated 715 cases with the Yuzpe regimen and had 17 failures, including the present case of an ectopic pregnancy. This rate is compatible with the calculated risk of 10% for ectopic pregnancies.⁴

We believe that this is the first reported case of an ectopic pregnancy after the Yuzpe regimen. We suggest that patients requesting post-coital treatment should be counselled about this rare but serious complication and warned of the need for prompt examination if they develop severe pelvic pain. Until more evidence is available about the presence or absence of a causative relation between postcoital hormonal treatment and ectopic pregnancies, the history of previous tubal pregnancy would be a strong contraindication to postcoital treatment. A careful vaginal examination at the follow up visit is important, as vaginal bleeding associated with an ectopic pregnancy can be so easily mistaken for a period, as in our patient.

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Margaret Pyke Centre, Soho Square, London W1

ALI A KUBBA, MB, MRCOG, deputy director

JOHN GUILLEBAUD, FRCS, MRCOG, medical director

Correspondence to: Dr Ali A Kubba.

Superior sagittal sinus thrombosis and essential thrombocythaemia

Both bleeding and thrombosis are complications of essential thrombocythaemia.¹ Common sites of thrombosis are the splenic vein¹ and the small vessels of the foot,² and the sagittal sinuses are rarely affected. Neurological symptoms such as amaurosis fugax, recurrent vertigo, and recurrent hemiparesis may occasionally be associated with thrombosis due to essential thrombocythaemia.³⁻⁴ We describe two patients with essential thrombocythaemia who presented with persistent headaches and chronic papilloedema. One patient was shown to have thrombosis of the superior sagittal sinus, and the second probably had a similar cerebral venous thrombosis.

Case reports

CASE 1

A 28 year old woman teacher presented to this hospital in 1980 complaining of severe headaches during the past six months made worse by coughing and bending. She had had two miscarriages, one in April 1979 at 16 weeks' gestation and the other in April 1980 at 24 weeks' gestation. She had stopped taking the contraceptive pill in April 1978.

On examination she was alert but had severe bilateral papilloedema. Visual acuity was 6/6 bilaterally and the visual fields were normal. There were no other abnormal findings. A gammascan and computed tomography of the head yielded normal results. Lumbar puncture showed a cerebrospinal fluid pressure of 3.15 cmH₂O, but further examination of the cerebrospinal fluid gave normal results. Left carotid arteriography showed superior sagittal sinus thrombosis with occlusion of the sinus at the vertex. Haemoglobin concentration was 13.9 g/dl and white cell count was $9.1 \times 10^9/l$ with a normal differential, but the platelet count was $748 \times 10^9/l$ and subsequently varied between this level and $934 \times 10^9/l$. The erythrocyte sedimentation rate was 15 mm in the first hour. Total cellularity of the bone marrow was normal but there were increased numbers of megakaryocytes. There was no cause for secondary thrombocytosis, and essential thrombo-

cythaemia was diagnosed. Platelet aggregation in response to adenosine diphosphate, adrenaline, and collagen was normal. She underwent bilateral decompression of the optic nerve in October 1980 and was treated with hydroxyurea. The platelet count fell to within the normal range and her symptoms resolved, but mild papilloedema persisted.

CASE 2

In 1977 a 38 year old Iranian woman presented with visual obscurations. Examination showed bilateral papilloedema but no other abnormal physical signs. She had never taken the contraceptive pill. Cerebrospinal fluid tests, a gammascan, and serial computed tomography of the head over three years gave normal results. Benign intracranial hypertension was diagnosed. She was found to have thrombocytosis and was referred to this hospital. Her platelet count was $1326 \times 10^9/l$. Haemoglobin concentration was 13.1 g/dl and white cell count $9.5 \times 10^9/l$ (6% myelocytes, 1% metamyelocytes, 50% neutrophils, 35% lymphocytes, 3% eosinophils, 5% monocytes). Total cellularity of the bone marrow was normal, but increased numbers of megakaryocytes were present. Essential thrombocythaemia was diagnosed. Platelet aggregation in response to adenosine diphosphate, adrenaline, and collagen was normal. She was treated initially with pipobroman and subsequently with one dose of radioactive phosphorus. The thrombocytosis and her symptoms resolved. Mild papilloedema persisted for six years but otherwise she remained well.

Comment

Spontaneous thrombosis of the superior sagittal sinus is rare⁵ and may present as isolated papilloedema. The classic predisposing factors such as sepsis, malignancy, polycythaemia rubra vera, pregnancy, and the puerperium are often absent. This condition was demonstrated in one patient in this report and was strongly suspected in the other; both patients had essential thrombocythaemia. Anti-coagulants were not used in either case because of the associated thrombocytosis.

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St Bartholomew's Hospital, London EC1A 7BE

M F MURPHY, MRCP, MRCPATH, senior registrar in haematology

C R A CLARKE, MB, MRCP, consultant neurologist

R L BREARLEY, MD, MRCPATH, senior registrar in haematology

Correspondence to: Dr M F Murphy.

Low protein diets in uraemia

Patients with chronic renal failure usually receive dietary advice as a palliative measure to reduce uraemic symptoms, but some start dialysis without having reduced their protein intake. In both experimental animals and man early protein restriction slows the rate of progression of renal failure.¹⁻² We reviewed our experience of low protein diets to alleviate uraemic symptoms in patients approaching end stage renal failure referred to this unit for maintenance dialysis.

Patients, methods, and results

Over the past five years (1977-82) 176 patients were accepted for dialysis; 20 continued to be managed conservatively and 68 received no dietary treatment because of late referral. Of the remainder, 12 had evidence of protein malnutrition—that is, low serum albumin concentrations or low ratios of serum urea to creatinine concentrations—and did not receive advice to reduce protein intake. Ninety six patients were placed on diets containing 35-45 g protein (about 0.6 g/kg body weight) and 19-23 mmol