Acquired coarctation of the aorta—a long-term complication of irradiation

Many cardiac complications of mediastinal irradiation have been described. Lesions of the great vessels are very much less common as they have higher resistance; nevertheless, in animal studies damage has been confined to the irradiated area. This article describes an unreported vascular long-term complication of irradiation in man.

Case report

A 35-year-old man with viral pericarditis, confirmed by serial viral studies, was referred for an opinion on a widely heard murmur. The only cardiovascular abnormalities were a widely heard ejection systolic murmur, maximal in the aortic area (with associated thrill), radiating to the neck, apex, and widely over the posterior chest, and blood pressures of 140/60 mm in the right arm and 120/60 mm in the left, with no femoral delay. The medical history was of Hodgkin's disease at the age of 16, of nodular sclerosing type (clinical stage IIa), which had been treated by mantle irradiation of 3000 rads. Hilar regression and upper lobe fibrosis had been seen over two months; the patient had remained disease-free for 15 years; and had been discharged from follow-up. No murmurs were heard during follow-up or in subsequent medical examinations.

Cardiac catheterisation and arch aortography showed normal right and left heart pressure with no aortic valve gradient. There was a 20 mm peak systolic gradient across the aortic arch on withdrawal of the catheter. Arch aortography (see figure) showed irregularity of the aortic arch between the origins of the left carotid and subclavian arteries, with notching of the inferior surface at that point (arrowed). When seen three months after catheterisation the patient was developing early obstruction of the superior vena cava.

Discussion

Though several cardiac and pulmonary complications of mediastinal irradiation have been reported, the effects on the great vessels are seen rarely. In one case of aortic irradiation damage features were localised to the irradiated area but had produced no signs or symptoms during life. The histological features were similar to those produced experimentally. Damage to other major vessels has occasionally been reported, sometimes with many years before presentation.

In this case thickening and fibrosis of the aortic arch, with subsequent buckling, were probably the major factors in producing the clinical and haemodynamic findings. Possibly with the current extended survival of patients with Hodgkin's disease treated by irradiation this complication may be met with more frequently in future.

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Malignant hypertension secondary to idiopathic arteritis of the aorta

Arterial disease in the tropics assumes a different pattern from that seen in the West. Unlike the degenerative disease of developed countries, the most common pathological process appears to be inflammatory and of unknown and possibly varying aetiology.

Case history

A 16-year-old Zambian boy was admitted with a two-month history of throbbing headaches, latterly associated with vomiting, increased urinary frequency with nocturia, and progressive visual deterioration. He denied haematuria, loin pain, or abdominal trauma. He was well nourished, with blood pressure 240/130 mm Hg in each arm; 230/130 mm Hg in each leg. There was a high-pitched bruit over the epigastrium, a bruit over the left femoral artery, and the pedal pulses were palpable only with difficulty. There was definite accentuation of the aortic second sound, but no enlargement of the heart. The fundi showed grade IV hypertensive changes, with acuity on the right of 6/9, and on the left of 6/60, with macular exudates. Investigations showed repeatedly normal haemoglobin; white cell count; urine examination; and serum concentrations of urea, electrolytes, creatinine, cholesterol, serum proteins, alkaline phosphatase, creatinine clearance, and urinary vanillyl mandelic acid. Haemoglobin electrophoresis was AA. The results of serological tests for antinuclear factor, rheumatoid factor, and syphilis were negative. Antistreptolysin-O titre, chest x-ray film, and electrocardiogram were within normal limits. Mantoux test was positive 1-1000. Erythrocyte sedimentation rate ranged between 25 and 37 mm per hour. An abdominal x-ray film showed a well-demarcated area of calcification overlying the body of the second lumbar vertebra. An intravenous pyelogram showed delayed nephrogram phase and loss of cortical volume at the left upper pole, with the right kidney measuring 12.5 cm and the left kidney 10.5 cm. Aortography confirmed aortic disease and showed the lower pole of the left kidney being supplied by an accessory vessel.

Surgical exploration showed calcification and patchy narrowing and dilatation of the aorta from 5 cm above the origin of the renal vessels to its bifurcation. The left kidney was supplied by two distinct polar arteries. That supplying the upper pole appeared fibrosed, while the lower polar vessel was widely patent. The upper pole of the kidney was atrophic. The right kidney and its blood supply were normal. Left nephrectomy was performed. Two months postoperatively his blood pressure remained controlled at 120/80 mm Hg on a “loop” diuretic alone. Fundoscopy changes regressed with residual exudates only. Histological examination confirmed an ischaemic left upper pole. The left lower pole and a right renal biopsy were normal. The upper polar artery showed degeneration and fibrosis in the media, with diffuse thickening of the intima secondary to connective tissue proliferation.
Discussion

Primary arteritis of the aorta is a distinct entity. Earlier descriptions of Takayasu’s disease, brachial arteritis, and aortic arch syndrome cover only one facet, and may be misleading. More recent reports based on necropsy or angiographic findings suggest that occlusive changes may affect any part of the aorta and its branches.

This present case presented with renovascular hypertension. Patchy narrowing of the abdominal aorta with its accompanying calcification and partial occlusion of the left renal artery with non-specific histological findings strongly suggest the diagnosis of primary aortitis. Removal of the stenosed vessel and affected kidney restored the blood pressure to near normal levels.

The aetiology of this apparently rare condition remains obscure. The disease may well represent the end result of a variety of different initiating agents, the anatomical site of disease being modified by other unknown variables.

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Neurological manifestations in a patient with filariasis

Filariais, in particular infections with Loa loa and Depetalonema perstans, may occasionally develop some neurological complications. However, these are neither common nor well known. We believe it important to call attention to them because they can be mistaken for a neurological disease of different aetiology. For this reason we report following case.

Case report

A 28-year-old Negro male born in Rio Muni, West Africa, was admitted to hospital because 10 days previously he had started to complain of vomiting, unsteadiness, loss of hearing on the left side, and paralysis on the left side of the face. Clinical examination disclosed hepatosplenomegaly, nystagmus, sensorineural hypacusis, and signs of left facial paresis. Blood analysis showed eosinophilia of 18% and numerous microfilariae, later identified as Loa loa and D perstans. No plasmoidia were seen. Blood chemistry, urine analysis, stool examination, and serological tests for Echinococcus granulosus and toxoplasma were all normal. In a bone marrow aspiration a high eosinophil count was made. An electrocardiogram showed T wave inversions on V1 to V5. Lumbar puncture revealed clear fluid with 45 mg/100 ml (0-45 g/l) protein, 70 mg/100 ml (4-2 mmol/l) glucose, and 45 cells. The presumptive diagnosis of cerebellar microangiopathic or acute encephalitis was made. Cerebral angiographic, pneumoencephalographic, and echoencephalographic studies were all normal.

One month after symptoms began spontaneous recovery set in, and all the symptoms completely disappeared without treatment in two more weeks.

Discussion

This patient inadvertently took Ovral for the first three months of her pregnancy and gave birth to an infant with a tracheo-oesophageal fistula, an abnormality whose normal occurrence is 1 in 3000. This association was also reported by Oakley et al in a letter to the Lancet on the relation between hormonal pregnancy tests and congenital malformation.

Until it is proved that there is no relation between these preparations and congenital abnormality it is prudent to ensure that no patient is prescribed oral contraceptives unless pregnancy has been definitely excluded.


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Tracheo-oesophageal fistula associated with hormonal contraception during pregnancy

Progestogen/oestrogen preparations have been suspected as potentially teratogenic drugs for several years but reports are conflicting. It is therefore important to document all cases where congenital abnormality is associated with the ingestion of the drugs during pregnancy. I report here such a case.

Case report

A 23-year-old primigravida was booked for hospital delivery at about the 20th week of pregnancy. Her last menstrual period had been on 22 March 1975, thus making her expected date of delivery 29 December 1975. Nevertheless, she had taken the oral contraceptive Ovral (Di-norgenstril 0-5 mg, and ethinyl oestradiol 0-05 mg) regularly until June 1975. At this time a vaginal examination performed to elicit the cause of 12 weeks’ amenorrhoea confirmed a 12 weeks’ size pregnancy. The patient attended antenatal clinic regularly for a total of 14 visits. The size of the uterus was initially small for dates and later large for dates owing to hydramnios. The result of a glucose tolerance test was normal and an x-ray film showed a normal singleton fetus. The hydramnios, therefore, was of unknown origin. On 20 January 1976, when the pregnancy was post term by dates, there was a slight rise in blood pressure and induction of labour was considered. Clinically the fetus was of good size and the amniotic fluid gave a positive “bubble” test and contained 20%, fat staining cells with Nile blue sulphate reagent. Induction of labour was planned, and was performed on 23 January 1976. Hydro-nate and fetal water rupture of membranes resulted in the drainage of over 4000 ml of clear liquor. A syntocinon infusion at 1 mg unit/minute was begun and the dose doubled every 30 minutes to 4 mg/minute, when labour was established. The latent phase of the first stage of labour lasted four hours and the active phase six hours. A delayed second stage was terminated by an easy vacuum extraction and the third stage of three minutes was uneventful. The puerperium was normal apart from a slight rise of blood-pressure for the first 24 hours after delivery.

The baby girl weighed 3000 g and the Apgar score at one and five minutes was 8. A nasogastric tube could not be passed and an oesophageal stasis with a fistula between the distal segment and the trachea was confirmed on x-ray examination. A repair was performed on the same day. Her postoperative progress was stormy. A gastrectomy was performed on 29 January 1976 and closure of fistulous track on 19 February 1976. After the latter her general condition improved, but she died from bronchopneumonia on 28 March 1976.