COMMENT

This case provides several features of unusual interest in the understanding of the lenticular changes due to copper deposition.

Rarity of Copper Deposition in the Lens in Wilson's Disease.—The factors governing the deposition of the copper are not known. This case appears to show that the deposition is not necessarily a late manifestation of the disease, as claimed by Boudin and Pépin (1959). Although rare, sunflower cataracts in Wilson's disease have been reported by Fleischer (1922), Siemerling and Oloff (1922), Vogt (1926, 1929a, 1929b), Jess (1929), Rohrschneider (1934), and Thiel (1934).

Classical Appearance of Sunflower Cataract.—As described by Duke-Elder (1954), the appearance of sunflower cataract is that of a central green disc in the anterior capsule of the pupillary area, with radiating petals thought to correspond to the folds on the posterior surface of the iris. When posterior capsular deposition occurs, it is said by the same author to be uniform and without pattern. In support of his contention that the radiating petals are due to the folds on the posterior surface of the iris he cites the observations of Jess (1929, 1930), that if the pupil is kept dilated the central disc increases in size; of Hillemanns (1896), who showed that with a deformed pupil the central disc took the shape of the deformed pupil; and of Belz and Bonnet (1948), who observed a case with a hole in the iris in which a disc appeared behind the false pupil as well as behind the true pupil.

As seen in Fig. 2, the anterior lens capsule showed a simple disc of chalcosis, while the posterior lens capsule showed frond-like petals of deposition. This observation suggests strongly that the iris is not responsible for the characteristic sunflower pattern of discoloration, since, as in the above case, this may be present in the posterior lens capsule.

Clearing of Copper from the Lens.—The disappearance of copper from an avascular region like the lens following the recovery from the central nervous system lesion further shows the close relation of copper to the pathogenesis of the disease. This late disappearance of the metal from the lens suggests that after the body-stores of copper have been depleted by treatment with D-penicillamine the serum falls below a critical level, which then permits the relatively rapid resorption of the copper deposits in the lens.

The spontaneous clearance of copper deposition from the lens has been reported in chalcosis due to retained copper-containing foreign bodies by Zur Nedden (1903), Oloff (1926), Clausen (1930), Záhŏr (1930), Blake (1931), Müller (1931), Cordes and Harrington (1935), Marner (1945), and Belz (1952).

In all reported cases the process of clearing has been slow, and in some it has been incomplete. No case report has come to our notice recording the disappearance of copper from the lens in a case of Wilson's disease. The disappearance of Kayser–Fleischer rings is, however, well recognized. In the present case the rings survived the lens deposits, but were much attenuated.

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Appendicitis Presenting as Infection of Right Thigh

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CASE HISTORY

Ten days before his admission to hospital a 68-year-old man visited his doctor complaining of pain in the right flank. The pain had started four days previously and was described as like heart-burn. There was no vomiting or any urinary symptoms. Tenderness was present well out in the right iliac fossa, with some guarding. Rectal examination showed no abnormality. Four days later he developed anorexia and vomiting. His bowels were regular. After three further days he was visited at home on complaining of pain in the right femoral triangle. There was no pyrexia and the pulse rate was not raised. Three days later he was admitted to hospital with an obvious infection of the right thigh with subcutaneous crepitus.

Examination showed an obese man who did not look particularly ill. His temperature was 98° F. (36·7° C.), pulse rate 96, and blood pressure 120/80. The abdomen was soft with no localized tenderness. The right thigh was swollen, red, and tender, and there was diffuse palpable crepitus. All movements of the right hip-joint were limited and painful.

He was given 8 ml. of anti-gas-gangrene serum, and penicillin 500,000 units six-hourly and streptomycin 1 g. twice daily were started. Under general anaesthesia multiple incisions made into the right thigh produced only a little foul-smelling pus from the region of the adductor muscles. Aerobic and anaerobic culture resulted in a growth of non-haemolytic streptococcus.

The patient recovered well from the anaesthetic, and the following day his condition was entirely satisfactory. On the second day

after operation he suddenly collapsed with dyspnoea, rapid pulse, and slight cyanosis. He was treated with heparin, digoxin, and hydrocortisone and placed in an oxygen tent, but he died 17 hours later.

At necropsy no purulent or other exudate was found in the peritoneal cavity. The appendix was retrocaecal, 5 cm. long, and gangrenous in its proximal half. On removing the appendix a small circular hole about 0·5 cm. in diameter was visible in the iliopsoas sheath and adjacent appendix. On cutting into the iliopsoas muscle a greenish necrotic track was found extending inferiorly beneath the inguinal ligament to present among the adductor muscles of the thigh. There was a soft non-adherent post-mortem clot in the right femoral vein, and a large adherent ante-mortem clot was blocking the right main pulmonary artery. The coronary arteries were widely patent.

Comment

The absence of clostridial organisms excludes gas-gangrene infection as being responsible for the gas in the thigh. The most likely explanation for the gas in the subcutaneous tissues of the thigh is escape of air and infected bowel contents from the ruptured appendix into the psoas sheath and thence to the thigh. The communication between the appendix and the psoas sheath was a direct one. Any increase in the intra-abdominal pressure—for example, defaecation—would force air direct from the hole in the appendix through the hole in the iliopsoas sheath and along the muscle fibres. It would seem likely that, by a series of such intermittent increases in pressure, air along with infected bowel contents gradually forced its way downwards, eventually breaking out in the iliopsoas sheath to present in the thigh. The remarkable feature in this case was the almost complete lack of systemic upset even when the presence of gas and pus in the thigh was evident. The patient was fully mobile for the first 10 days of his illness, though the infection must have been tracking down the psoas sheath. It is interesting to speculate on the final outcome had the patient not succumbed from a pulmonary embolus.

Similar cases of intra-abdominal infection presenting with gas in the thigh are rare. Dawson and Hardy (1948) reported the case of a 73-year-old woman who presented with an abscess in the left groin producing gas and pus. She died from a generalized peritonitis. Necropsy showed an inflamed pelvic colon with a fistula tracking into the left thigh. Barrington and Gardham (1932) reported a somewhat similar case. Their patient, a 52-year-old man, presented with a distended abdomen and crepitus in the left thigh. Necropsy showed perforated diverticulitis of the sigmoid colon with necrosis and gas in the psoas sheath. The case reported by Gordon (1936) presented as an appendix abscess which ruptured intraperitoneally, followed by the development of gas and pus in the right thigh. Culture of the pus grew *Clostridium welchii*. Necropsy revealed a large appendix abscess eroding into the psoas sheath. Clostridial infection similarly complicated the case reported by Wyman (1949), in which a 65-year-old man presented with an indurated area in the left thigh producing gas and pus. At necropsy a carcinoma of the descending colon ulcerating into the psoas sheath was found. Blood culture had grown *Cl. welchii*.

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Severe Aortic Stenosis Produced by Bacterial Endocarditis

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Valvar incompetence is a well-recognized sequel of active infective endocarditis (Cohen and Freedman, 1961). In contrast, the development of haemodynamically significant valvar stenosis is rare (Roberts *et al.*, 1967). This report describes a case of severe aortic valvar stenosis caused by vegetations of bacterial endocarditis.

Case Report

A 62-year-old white woman who had had rheumatic fever at the age of 14 years was admitted to hospital on 14 May 1968 with a three-months history of lassitude, anorexia, and loss of weight. She had previously enjoyed good health and had had a physically active life. Shortly after the onset of this illness an examination by her general practitioner had shown evidence of mild aortic and mitral rheumatic valvar disease. Chest *x*-ray examination confirmed the absence of cardiomegaly, and an electrocardiogram was normal. A blood count showed that she was anaemic, with a haemoglobin of 10·4 g./100 ml. Blood cultures were negative, and no specific therapy was given. Her symptoms became worse and attacks of night sweats supervened.

On admission she was mentally alert but looked pale and moderately ill, and had a pyrexia of 99·4° F. (37·4° C.). She had a sinus tachycardia of 104 per minute. The blood pressure was 110/70 mm. Hg. There was early clubbing of the fingers but no splinter haemorrhages were seen. The jugular venous pressure was normal. Crepitations were present at both lung bases. The apex beat was displaced to the sixth intercostal space just outside the midclavicular line, and clinical evidence of both left and right ventricular hypertrophy was found. The first heart sound was of normal intensity. A grade 3 pansystolic murmur of mitral incompetence and a fairly short mid-diastolic murmur were heard at the apex. A grade 2 short aortic ejection systolic murmur which radiated into the carotids and a short soft early diastolic murmur were heard at the base. The second heart sound was normally split, the aortic component being slightly decreased in intensity. An electrocardiogram showed sinus rhythm, a mean frontal plane QRS axis of −20°, and no definite evidence of chamber enlargement. Some left ventricular enlargement was apparent, however, on radiological examination. The chest *x*-ray film showed two areas of linear atelectasis, compatible with pulmonary embolism, in the right lower zone. The abdomen and extremities were normal.

Investigations showed haemoglobin 10·6 g./100 ml.; W.B.C. 10,500/cu. mm. (normal differential count); E.S.R. 48 mm. in one hour (Westergren); urea and electrolytes within normal limits; streptococcal antihaemolysin O titre 50 units; rickettsial complement fixation tests negative. Repeated blood cultures yielded *Streptococcus viridans* sensitive to penicillin.

The diagnosis was *Str. viridans* bacterial endocarditis superimposed on chronic rheumatic endocarditis with predominant mitral incompetence and associated mild aortic valve disease. Treatment with high-dosage penicillin and streptomycin was started, and the temperature settled within two days. Five days later the pyrexia recurred. This was attributed to *Escherichia coli* infection of the urinary tract, and it responded to treatment with nalidixic acid. Anticoagulants were also given in view of the radiological signs suggestive of pulmonary embolism.