FIG 1

was then grossly distended and similarly filled by organising thrombus.

The only other abnormalities were:—

- (1) A small infarct in the upper lobe of the right lung.
- (2) Fatty change in the liver.
- (3) Slight enlargement of the mesenteric lymph glands.

Histology. Renal, ulnar and mesenteric arteries—normal. Coronary arteries: The aneurysms were filled by organizing thrombus extensively infiltrated with polymorpho-nuclear leucocytes. The wall of the affected areas of the coronary arteries was grossly thickened, the outstanding lesion being splitting of the internal elastic lamina and extensive proliferation of fibroblasts

spreading through the remains of the media and adventitia into the peri-adventitial tissue. The whole lesion was widely infiltrated by polymorpho-nuclear leucocytes and eosinophils and, to a lesser extent, plasma cells. (Fig. 2).

Discussion

These appearances are virtually identical with the two similar cases described by Crocker and others (1957).

It would seem reasonable to conjecture that one coronary artery became occluded at the time of the collapse on 15.10.61 and the other on the day of death. No conclusion could be reached either from the clinical findings or the post-mortem appearances regarding the cause of this illness, but it is interesting that a reasonably large dose of steroids had no certain effect on the course of the illness as one might have expected if the basic abnormality had been that of a 'collagen disease'.

The two similar cases of Crocker and others (1957) were boys dying at 4 months and 8 months of age. Each fell suddenly ill with fever, blotchy rash, conjunctivitis, sore throat, tachycardia and leucocytosis. One had a large heart on X-ray and the E.C.G. pattern suggested myocardial infarction. He also had an eosinophilia. In spite of treatment the illness progressed with cardiac murmur, ectopic beats and progressive cardiac enlargement.

The illness of the second started with irritability, conjunctivitis, pharyngitis and a blotchy rash that waxed and waned for two weeks. There was neutrophil leucocytosis with left shift which subsided with treatment of an incidental urinary infection. The child went home, only to return after two months in heart failure which had developed rapidly over two days.

Summary

A case of aneurysmal coronary arteries with terminal bilateral occlusion in an infant is described. Two remarkably similar cases have been reported previously.

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CONGENITAL ABSENCE OF PAIN

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THE ability to react normally to painful or noxious stimuli serves to protect the body from damage. Interference with pain perception can occur from a variety of lesions at different anatomical levels

from peripheral nerve endings (as in progressive sensory radicular neuropathy) to cerebral cortical disorders (as in congenital absence of pain). In the latter condition there is no demonstrable lesion

in the sensory pathway to account for the unusual failure to feel pain although there is no analgesia or thermonæsthesia: the deep tendon and the corneal reflexes are present and the mentality is within normal limits. The nature of stimuli at normal thresholds are recognized correctly, but there is a failure to react in the usual defensive manner with withdrawal of the offended part, acceleration of pulse and respiration and elevation of blood pressure as in healthy subjects. The result is that the patient can sustain severe injuries without any conscious awareness that anything untoward has occurred as is illustrated in the case here described.

Although rare the condition is well recognized and reported in the literature: to date there are reports of some 46 cases in all. In order to comply with the condition as it is now recognized the absence of pain must be present from birth, the failure to react to pain must be generalized, the mentality must be within normal limits, and no organic lesion should be demonstrable. Winkelmann and others (1962) in the latest review of the subject propose a standard procedure for the examination of such patients based on separate demonstrations of perception of stimulus, integrity of peripheral receptors and afferent structures, and functioning of integrated involuntary reflex responses to pain.

Case History

P.S., a male child, was first seen when eight months of age by the casualty officer at the Memorial Hospital, Woolwich where his mother had taken him because he had bitten his fingers to such an extent that medical treatment was required. At a loss to explain the condition and how to deal with the persistent biting the casualty officer referred the child to the pædiatric department. Apart from these self-inflicted injuries the child had at no time shown any discomfort let alone pain and his mother recalled he had not reacted to his inoculations as other children do. Clinically he was a robust healthy child, the first born of healthy young unrelated parents: the pregnancy and delivery had been uneventful and until the time he began to injure himself they had had no concern about him. Routine examination revealed no abnormality apart from the demonstrable failure to react to the normally pain producing stimuli such as pressure on the tendo Achilles, testicular pressure or jabbing the skin with a sharp instrument. A presumptive diagnosis of congenital absence of pain was made which was confirmed by Dr. E. A. Carmichael who kindly saw him in consultation at the National Hospital, Queen Square. The parents were given to understand what the condition entailed, particularly for such serious conditions as an acute abdomen or a fractured limb. Supportive counselling was provided by the medical social case worker to help the parents to adjust emotionally to their child's condition.

FIG 1

Nothing untoward occurred till he was 21 months of age when he was noticed to have a swollen left leg and was seen at the hospital after it had been present for three weeks. A fracture at the lower end of the tibia and fibula was seen on X-ray with excessive callous formation (Fig. 1) presumably due to the unlimited and unrestricted activity. Although this was thought to be healing satisfactorily, an abscess developed two weeks later over the site of the fracture which turned out to be a discharging sinus from an underlying osteomyelitis necessitating prolonged chemotherapy and hospitalisation for almost two months. Since then he has sustained a fracture of a metatarsal which was discovered by X-ray when he was noticed to be walking strangely, likewise there was a complete lack of pain with a good deal of local swelling and thickening.

Summary

This unusual and uncommon condition is well illustrated by the patient described. This is undoubtedly a serious and hazardous disease since the absence of pain with the lack of protective warning when the body is in any way damaged means the tissues may be seriously damaged before being recognized and treatment given. There is no treatment for the disease itself and the management depends on the vigilance of the parents and later of the patient for any untoward bodily happening.

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