three decades. Hepatosplenomegaly is not always evident, though some patients have excess sphingomyelin in their livers. Sphingomyelinase levels are normal or marginally deficient. Dermatological manifestations of Niemann-Pick disease are uncommon and consist of a waxy texture and appearance of the skin with variable pigmentation of exposed areas and occasionally the appearance of eruptive xanthomata.

The clinical and histopathological appearance of the perianal lesions in our patient were entirely consistent with a diagnosis of Crohn's disease. The papular abdominal eruption with sarcoidal granulomata and without evidence of lipid deposition resembled in all respects the perianal changes.

McCallum & Gray (1976) reviewed the dermatological manifestations of Crohn's disease and divided them into two broad groups. The typical perianal, peristomal and oral lesions and flexural 'metastatic Crohn's' all have histological features characteristic of the disease, with collections of epithelioid and giant cells of the Langhans type without central caseation (the sarcoid reaction), infiltrated with and surrounded by variable numbers of lymphocytes and plasma cells (Gray *et al.* 1965). The second group show no characteristic histology and include erythema nodosum, erythema multiforme, pyoderma gangrenosum, palmar erythema, rosacea and polyarteritis nodosa. The term 'metastatic Crohn's' has been used to describe a granulomatous reaction occurring in the flexures and separated from the affected areas of the gastrointestinal tract by normal skin. Previous reports refer to such lesions affecting the groins, male genitalia, a flexure on the anterior abdominal wall of an obese patient (Mountain 1970), submammary skin (Parks *et al.* 1965) and the retroauricular area (McCallum & Gray 1976). Our patient had no clinical or radiological evidence of active Crohn's disease of the bowel, but the appearance of the skin manifestations of this disease several years before overt bowel involvement is well documented (Bishopric & Bracken 1964, Lockhart-Mummery 1964, Gray *et al.* 1965, Strathers *et al.* 1967).

There has been no previous recorded association between Niemann-Pick and Crohn's disease and the two conditions in this patient may well be coincidental. The presence of granulomatous papules on the abdominal wall of our patient in association with perianal lesions may represent yet another form of 'metastatic Crohn's disease'.

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References

Bishopric G A & Bracken J S
(1964) *Southern Medical Journal (Nashville)* **57,** 675–678
Gray B K, Lockhart-Mummery H E & Morson B C
(1965) *Gut* **6,** 515–524
Lockhart-Mummery H E
(1964) *Proceedings of the Royal Society of Medicine* **57,** 897–898
McCallum D I & Gray W M
(1976) *British Journal of Dermatology* **95,** 551–554
Mountain J C
(1970) *Gut* **11,** 18–26
Neville B G R, Lake B D, Stephens R & Sanders M D
(1973) *Brain* **96,** 97–120
Parks A G, Morson B C & Pegum J S
(1965) *Proceedings of the Royal Society of Medicine* **58,** 241–242
Strathers G M, Abbott L G & McGuinness A E
(1967) *Archives of Dermatology* **95,** 375–380

Investigation of brain death with Doppler-shift ultrasound[1]

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There has recently been considerable discussion about the diagnosis of brain death. Yoneda *et al.* (1976) used Doppler-shift ultrasound and reported that sonagrams (Figure 1) obtained from the common carotid arteries in patients who have developed brain death are characterized by considerable reverse blood flow and a single systolic peak.

We have used Doppler-shift ultrasound and spectral analysis with good directional resolution (Coghlan *et al.* 1974, Coghlan & Taylor 1976) to examine the carotid arteries in over 2000 subjects, which included asymptomatic volunteers and conscious patients who subsequently underwent carotid angiography for symptoms not confined to cerebrovascular incidents (Gosling & Lewis 1979). Doppler signals were displayed in the form of sonagrams which, provided cardiac abnormalities such as aortic incompetence were not present, only occasionally demonstrated a transient and minimal amount of reverse blood flow; this was usually observed at the end of cardiac systole in sonagrams obtained from the common carotid artery. Further, unless the sonagram

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Figure 1. Directional common carotid sonagram over one cardiac cycle from a healthy young volunteer, showing Doppler-shift frequency on the ordinate and time on the abscissa. Blood flow towards the brain is displayed above the horizontal line and any reverse flow is displayed below the line. The blackness of the trace at each point is related to the number of erythrocyctes generating the same frequency, i.e. moving at the same velocity. Two peaks are present during cardiac systole and no reverse blood flow is shown

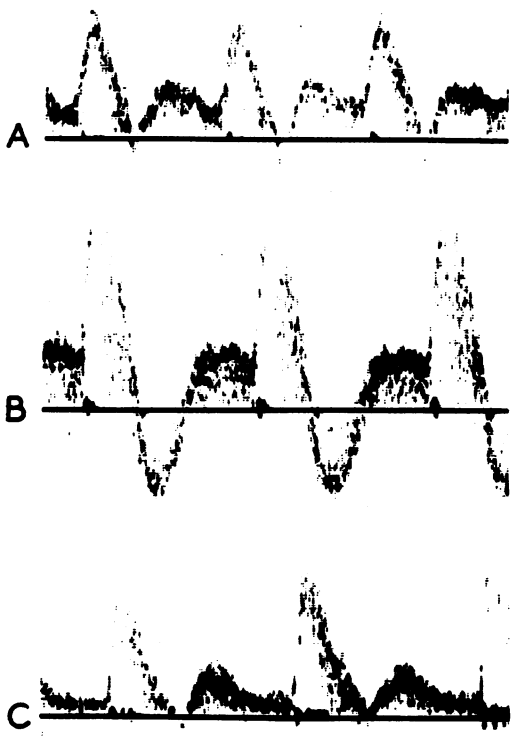

Figure 2. Directional sonagrams over three cardiac cycles from the right common carotid artery obtained from the patient on the third (A), fifth (B) and seventh (C) days following the cardiac arrest. Considerable reverse blood flow at the end of systole had developed by the fifth day. This had decreased to a minimal amount (C) when the patient had improved and was tried off the ventilator. A single systolic peak was present on the third and fifth days and an indistinct second peak had developed by the seventh day

waveform was distorted by signals from blood flow disturbance such as turbulence or vessel wall movement, two distinct peaks were noted during systole in all cases.

We have confirmed that marked reverse blood flow and a single systolic peak are present in common carotid sonagrams from patients who have developed brain death. However, this finding is not pathognomonic, as demonstrated by the following case report.

Case report

A 32-year-old male insulin-dependent diabetic was admitted to hospital having developed bronchopneumonia and ketoacidosis. He had no history of cardiac disease and on examination no cardiac abnormality was noted. Soon after admission he suddenly became confused and had a cardiac arrest. Following resuscitation he was hypotensive and in deep coma, with no response to painful stimuli and no spontaneous respiration. Artificial ventilation was commenced and he also required peritoneal dialysis for acute renal failure. His pupils were fixed at 6 mm and the corneal and pharyngeal reflexes were absent. An electroencephalogram was not performed.

Doppler examination of his common carotid arteries on both sides revealed a single systolic peak and the development of considerable reverse blood flow by the fifth day of ventilation (Figure 2A, B). No definite internal carotid signal was detected at that time and sonagrams obtained from the external carotid, superficial temporal, facial and supraorbital arteries also showed a single systolic peak and reverse blood flow. The patient's condition improved and after a week he was taken off the ventilator, at which time his common carotid sonagrams showed only minimal reverse blood flow (Figure 2C). He was, however, unable to maintain adequate spontaneous respiration and needed a further week of ventilation. Following this his steady improvement was complicated by the development of a severe peripheral neuropathy; when discharged from hospital he had no evidence of mental impairment.

Comment

The precise cause of the cardiac arrest was uncertain. It is likely the patient subsequently developed raised intracranial pressure from acute cerebral oedema, since the reverse common carotid blood flow and single systolic sonagram peak were most probably due to reflected pressure waves from increased impedance of the cerebral vascular bed. In an unconscious patient these features are therefore not exclusively associated with brain death and would appear to be caused by any condition which increases

vascular impedance of the brain; most importantly, they can be associated with normal mental function following improvement of the underlying condition.

References

Coghlan B A & Taylor M G
(1976) *Ultrasound in Medicine and Biology* **2**, 181–188
Coghlan B A, Taylor M G & King D H
(1974) In: Cardiovascular Applications of Ultrasound. Ed. R S Reneman. North Holland, Amsterdam; pp 55–65
Gosling R G & Lewis R R
(1979) In: Progress in Stroke Research 1. Ed. R M Greenhalgh and F Clifford Rose. Pitman Medical, Tunbridge Wells; pp 247–269
Yoneda S, Nishimoto A, Kukada T, Kuriyama Y, Katsurada K & Abe H
(1976) *Stroke* **5**, 707–713

Figure 1. Facial lesions; note elevated lesions on the chin

Erythema elevatum diutinum[1]

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Erythema elevatum diutinum, a rare disorder of the skin, was found to occur in association with an IgA paraproteinaemia. The rarity of both these conditions and previous reports of erythema elevatum diutinum occurring in association with IgA monoclonal gammopathy and IgA myeloma suggest the association may be more than by chance. A hitherto undescribed defect of monocyte migration and a defect of T-cell blastogenesis in erythema elevatum diutinum are described. Dapsone produced a characteristic clinical response and corrected defective monocyte migration and T-cell blastogenesis.

Case report

A 45-year-old woman presented with a red, raised, persistent rash which had been present for one year. Individual lesions were smooth-surfaced, erythematous and often brownish or purplish patches or plaques, some of which had a purpuric element. Several lesions, particularly the most elevated, were circinate or annular and had a yellowish hue with a surrounding border of erythema. The lesions were marked and extensive over the bony prominences of the face, forehead (Figure 1), extensor aspects of the wrists, upper back (Figure 2), and anterior chest wall. There were a few lesions on the dorsa of the hands. The lesions were always painful but only the lesions on the dorsa of the hands were tender. The lesions fluctuated in intensity, being more raised

Figure 2. Erythematous purplish and brownish patches on the back. Lesions are most marked over the vertebral column

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