"... THIS STRANGE EVENTFUL HISTORY"

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The history of an unusual case is considered worthy of record as it illustrates many features of interesting pathology. The events take place over a number of years, from the original occurrence of a gunshot wound in 1917, age 21, to the patient's decease in 1953, age 57, and are set out in sequence with the avoidance of unnecessary detail. Short discussions are interposed where most relevant, to avoid a more lengthy discourse at the end of the article.

The condition starts with a gunshot wound of the left hip in 1917 which resulted in disorganization of the hip with shortening of the leg. The pelvis in the region of the left hip joint showed sequestra and retained foreign bodies radiologically, and clinically several discharging sinuses were evident. The discharge persisted, varying in character from muco-purulent to frank pus and varied in amount. At no time did it cease altogether. In addition, there were acute "flares" occurring with some regularity at approximately six-monthly intervals and requiring admission to hospital. These incidents subsided satisfactorily with conservative treatment and no further surgery was thought advisable. Amputation would have had to be at the hip joint and would not have cured the condition nor permitted a satisfactory prosthesis. As described, this is a typical case of old bony injury with chronic osteo-myelitis, and a large number of such were seen between the two wars. The story told is a typical one of persistent discharge and recurrence of acute attacks of swelling and inflammation. These acute phases are interesting because in one's own experience they do not seem to have any causative factor. Unnoticed trauma may occur at any time, but over a large number of cases seen, no such incident has ever been noted and one would expect that there would be some association in at least some of them. Moreover, unlike this particular case, the majority do not have any regularity in the cycle of their attacks. One has seen a recurrence within a matter of weeks followed by fifteen years' remission. One has also seen an acute exacerbation occurring for the first time more than thirty years after the original wound. They are cases in the treatment of which masterly inactivity is the undoubted line to be pursued and attempts at radical drainage do no good and may even cause harm. Cases have been seen where an enthusiastic surgeon has attempted to lay open the seat of mischief, and although his extensive incisions were worthy of better reward, the abscess has pointed just alongside one of them! The explanation of this is considered to be that the disorganized tissues permit drainage to occur through a track of least resistance and that, such having been formed, it is quite effectively walled off. A further incision would only lead to further scar tissue formation, and the greatest efficiency is obtained by easing of pain until the place of pointing can be clearly seen beneath the skin, when it can be assisted in its final exit. Treatment by chemotherapy or any agent introduced via the blood-stream is disappointing, and the explanation of this is somewhat similar to that of the
tracking of the pus. The bone can be seen radiologically to be thickened and dense and can have but little blood supply. Any substances administered parenterally, therefore, simply do not reach the site. The factor causing the flare is totally indefinite. It has been described as a breaking down of the balance between resistance and infection, and while this is so, the reason for such is still not ascertained. Sound advice to give to the patient is not to worry about a recurrence which may never take place, but to have treatment for it at the first sign of acute mischief. Cases have been noted where well-intentioned instruction has been given to avoid any possible trauma and men have made their lives very restricted and very unhappy, only to see a flare occur in spite of all their precautions, and such is only adding further limitation of life to one already afflicted. Chronic bone sepsis is not seen so frequently in World War II cases as in World War I. There are several factors that may account for this, but in general the explanation is considered to be that World War I was largely fought in trenches with constant shelling and wounding by fragments. Everything was, therefore, favourable to infection, by no means least being the clothing taken into the wound, and there were, of course, no antibiotics available at that time. While clothing would invariably be grossly contaminated, it is an interesting contra-reflection that one has personally removed a sizeable piece of the tongue of an Army boot from the dorsum of a foot, twenty years after wounding, and at no time had sepsis been present!

Reverting from general discussion of such cases to the individual case in question, the cycle of acute phase and remission went on until June, 1945, when a new factor was described, that being severe clinical anæmia. This was confirmed by blood count and it was noted that all constituents of the blood were reduced, with red cell count of approximately 2,000,000, white cells 5,000 and colour index unity. Following transfusion with concentrated red cells, considerable improvement resulted and the count was raised to 3,500,000 red cells, 11,000 white cells, colour index unity. The first transfusion was given in July, 1945, and thereafter the cycle has added to it one of progressive anæmia. This was not related to the acute exacerbations of the hip condition and seemed to be a progressive state. Frequent admissions to hospital at approximately six-monthly intervals now became necessary in order that the blood picture could be brought up to a reasonable state and two counts are quoted as being illustrative. In May, 1948, red cells were 1,500,000, white cells 6,000, colour index slightly over unity. After repeated transfusion, six months later (December, 1948) the count was red cells 3,250,000, white cells 11,000, colour index unity. In no count was there any evidence of grave disturbance of the proportions of white cells. From the diagnosis of anæmia in 1945 until 1953, the treatment became almost routine. The hip condition was now much more stable as a simple chronic osteo-myelitis with constant discharge but without flares, and the symptoms were those of progressive anæmia. It was clear that the man was literally living on “borrowed blood” and that his own bone marrow was incapable of production to a degree adequate for his needs. He was considered to be a case of secondary aplastic anæmia. As one would expect, no improve-
ment followed liver therapy tried in many forms. In October, 1953, transfusion was followed by a reaction of headache, pyrexia and rigors. This was the first time such had occurred, although no fewer than forty-three pints of blood had been given. In 1953, during in-patient treatment lasting five months, seven pints of concentrated red cells had been administered. The blood count in the middle of November was: red cells 1,600,000, white cells 1,000, haemoglobin 39 per cent., colour index 1.15, and it was obvious that the general condition was deteriorating. On 28th November, oedema in the left leg was noticed and the occurrence of this coincided with a diminution in the amount of urine passed. Within three days both lower limbs had become severely oedematous and there was abdominal distension. The veins over the chest wall were noticed to be prominent and pigmentation of the skin was evident. This was of dark brown colour and strongly resembled sunburn. There was marked ascites and the spleen was readily palpable as being grossly enlarged. The urinary output was low and urine contained a considerable quantity of albumin. It was clear that the end was approaching and further transfusions were not undertaken, nor was any detailed investigation likely to reveal anything which would enable any therapeutic measures to be instituted. The condition now resembled amyloid disease except for the pigmentation, and caused considerable discussion which raised a further point of interest. No member of the hospital staff could recall a case of chronic bone sepsis producing amyloid disease, although one would expect such to be a textbook cause, and it was ascertained that in a period of thirty-two years no such complication had arisen. The reason one would suggest for this is that the bone sepsis is so shut off from the circulation as not to cause lardaceous change. This may well be so, but in this case there is no doubt that the absorption was sufficient to produce failure of bone marrow and blood formation. Apart from the emphasis on the absence of amyloid complications in other cases, one must leave the matter open.

The patient died on 28th December, becoming progressively weaker. Autopsy showed all the diagnoses to be correct save the last. The condition was one of aplastic anaemia attributed to chronic bone sepsis and the final developments were due to haemosiderosis. This in turn was attributed to the repeated transfusions which had been necessary. One can, therefore, conclude by saying, although this was a case of the utmost interest, he did not form the first case of amyloid disease seen in bone sepsis and such has yet to be encountered.

The condition of "transfusion haemosiderosis" has been described in several cases in recent medical literature. The post-mortem appearances resemble those of haemochromatosis and clinical diabetes has occurred from involvement of the pancreas. The reason for the deposit of haemosiderin is not understood except in the somewhat vague explanation that under certain circumstances the body seems to retain all the iron it can.

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