Metastatic Acropachy in Lymphatic Leukemia

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Peripheral manifestations in lymphatic leukemia are well recognized. They may dominate the clinical picture and closely simulate other diseases. The unique and confusing acral signs in this illustrative case merit detailed description. The only similar case recorded is that of Schittenhelm. Rhinophyma was, however, an additional feature in our patient.

Case Report

W. J. M., a retired rag and bone collector, aged 70, was admitted to Whips Cross Hospital on June 6th, 1953, for investigation. He complained that in the past two years he had gradually become a freak. His finger tips and the tip of his nose had become swollen. No previous illness of note had occurred. The personal and family histories were essentially noncontributory.

Physical examination revealed moderate physique and good color. There was no pigmentation, purpura, or telangiectasia. The salient peripheral signs included rhinophyma (fig. 1), moderate cervical lymphadenopathy, and distinctly bizarre hands (fig. 2) and feet (fig. 3). The hands had the hybrid appearance of multiple enchondromatosis and those of Fallot’s tetralogy. Soft, elongated, subcutaneous masses (infiltrations) were present on the dorsum and lateral aspect of some of the fingers. The feet showed similar but less conspicuous changes. Sparse, shotty, non-tender lymph nodes were palpable in the axillae and inguinal regions. Apart from hepatic and splenic enlargement to about 3 cm. each below the appropriate costal margin in the mid-clavicular line, other systems were normal.

Investigations

Hematological

Examination on admission revealed: hemoglobin level of 77 per cent (11.4 Gm. per cent); erythrocytes, 4,750,000 per cu. mm. with a mean cell diameter of 7.0 μ; leukocytes, 142,000 per cu. mm. (polymorphonuclears 3 per cent; lymphocytes 96 per cent; monocytes 0.5 per cent; basophils 0.3 per cent); platelets 141,000 per cu. mm. The blood film was typical of lymphatic leukemia and showed 1 lymphoblast and 60 smudge cells per 100 white cells. The bleeding time, clotting time, and prothrombin index were all normal.

Biochemical

These results showed: blood urea, 51 mg. per cent; serum uric acid, 6.0 mg. per cent; serum cholesterol, 196 mg. per cent; serum protein, 6.7 Gm. per cent (albumin 4.0 Gm. per cent); thymol turbidity, 4 units; cephalin-cholesterol flocculation, 1 plus after 24 hours.

Radiological

The entire body was x-rayed but only significant findings will be mentioned. The x-ray film of the chest was normal apart from an apparently quiescent tuberculous lesion at the left apex. The hands (fig. 4) showed multiple translucent cysts in all the phalanges and destructive changes in the metacarpals and the terminal phalanges. The cystic changes resembled those of Jungling’s osteitis tuberculosa multiplex cystoides but other evidence...
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of sarcoidosis was lacking. The appearances in the feet were similar but less striking. Apart from infiltration at the lower ends of the radii, ulnae, tibiae and fibulae, the skeleton was normal.

Histological

Biopsies of subcutaneous deposits and subungual nodules from the fingers (fig. 5) showed the characteristic histological appearance of lymphatic leukemia or its variant, lymphosarcoma.*

FIG. 1.—Profile showing rhinophyma.

FIG. 2—Spatulate digits due to marginal and dorsal leukemic infiltration. Subungual deposits causing bulbous tips and splintered nails.

FIG. 3.—Swollen toes from invasion of the soft tissues. Hornlike overgrowth of nails to protect tender infiltrated quicks.

PROGRESS AND TREATMENT

Shortly after admission a course of urethane, 1 Gm. daily, was commenced. It was, however, abruptly terminated after two weeks when the leucocyte count had fallen to 51,000 per cu. mm. (polymorphonuclears 7 per cent; lymphocytes 90 per cent; monocytes 2 per cent; eosinophils 1 per cent). The hemoglobin value and platelet count remained virtually unchanged. A week later the white cell count showed a further fall to 30,000 per cu. mm. (polymorphonuclears 8 per cent; lymphocytes 92 per cent) but rose to 102,000 per cu. mm.

* We are indebted to Professor Crawford, St. George’s Hospital, London and Dr. Raeburn, Whipps Cross Hospital, London for these reports.
Fig. 4.—Skiagram of hands showing extensive invasion of phalanges and severe erosion of the distal ones.

Fig. 5.—Subungual deposit. Section shows the subepithelial tissue x 600 packed with large lymphocytes, showing occasional mitosis.
two months later. At the time of writing (July, 1954), this value is 138,000 per cu. mm. (polymorphonuclears 2 per cent; lymphocytes 97 per cent; basophils 1 per cent) while the hemoglobin level is 79 per cent (11.8 Gm. percent) and the platelet count, 84,000 per cu. mm.

This treatment, which was not resumed, produced no regression of the peripheral lesions. No other special treatment was given. Supportive treatment included surgical mani- cure of the onychogryphosis and occasional short courses of penicillin to control recurrent paronychiae. Since his discharge from the hospital on August 1st, 1953, he has remained well. No further striking lesions have occurred but the "freak" features have become modestly accentuated.

DISCUSSION

Dermal involvement in leukemia occurs particularly in the lymphatic type where a primary dermatological disorder may occasionally be erroneously diagnosed. Schittenhelm described a patient with subungual infiltration and co-existent high grade bony deformity of the hands. The isolated appearance of such digital or rhinophymatous changes is not mentioned in Forkner's monograph or in subsequent literature on leukemia, indicating that this combination of clinical features is probably unique. Histological verification of the presumed leukemic nature of the rhinophyma has not been ventured.

Skeletal invasion is common in juvenile leukemia, yet Craver and Copeland found radiological evidence of this in only 5–10 per cent of patients with lymphatic leukemia. Their order of frequency of affected bones was femora, humeri, pelvis, metacarpals, ulnae and vertebrae. They observed that gross changes in bone occur more often in low-grade lymphatic leukemia or in sub-leukemias than in cases with marked leukocytosis. They also considered that skeletal invasion did not imply a graver prognosis. Both osteolytic and osteoblastic changes occur, but in adult chronic leukemia the osseous lesions are generally erosive. These destructive changes tend to affect the metaphysis of long bones while the proximal metaphysis of the proximal phalanges and the distal metaphysis of the metacarpal bones may also be involved, as in our patient.

Urethane medication for 2 weeks produced a striking reduction in the leukocyte count of our patient. There was, however, no objective change and none could be expected from this brief period of therapy. Irradiation of the peripheral lesions was not resorted to as these were painless and so slowly progressive while the possibility of postradiation fibrosis producing frank ulceration was a more serious deterrent. Indeed, proof is lacking that, in certain very chronic low-grade leukemias, increased survival time follows this treatment. Even when untreated their survival period may approach a decade.

SUMMARY

A patient with chronic lymphatic leukemia exhibiting the unique combination of bulbous digits from metastatic invasion and rhinophyma is described. The associated dermal and skeletal changes are discussed. Although two weeks of urethane administration produced a prompt hematological response, the peripheral lesions were unaffected by this very short therapeutic course.

SUMMARIO IN INTERLINGUA

Es describite le caso de un paciente de chronic leucenmia lymphatic qui exhibiva le unic combination de digitos bulbose (resultante de invasion metastatic) con
rhinophyma. Nos discute le associate alterationes dermal e skeletic. Le administration de urethano durante duo septimanas resultava in un prompte responsa hematologic, sed le lesions peripheric non esseva afficite per iste breve curso therapeutic.

REFERENCES

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