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Hand — Schüller — Christian Disease (Case Report)

by

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Introduction

Eosinophilic granuloma of the bone, Hand-Schüller-Christian disease and Letterer-Siwe disease have formerly been considered as a distinct entity (Siwe, 1949 and Otani, 1957). Conversely, Wallgren (1940), Farber (1942), Jaffe and Lichtenstein (1944) and Lichtenstein (1953) have concluded that all three conditions represented variations of the same basic disease process. Lichtenstein in 1953 grouped them under the term "histiocytosis X" referring to the basic underlying proliferation of histiocytes, while the letter X following the term histiocytosis was to emphasize the unknown etiology of the disease. Nevertheless, because the course and prognosis of these three entities are so different, it seems worthwhile to perpetuate the clinical divisions while recognizing that these may represent different phases of the same disease (Avery, et al., 1957).

Oberman (1961) arbitrarily distinguished three groups corresponding roughly to the three classic categories based upon the clinical and radiological findings at the onset of the disease, i.e.:

1. Eosinophilic granuloma of the bone, constitutes those cases in which the disease is completely confined to the skeleton as a single or multiple lytic lesion at the onset without evidence of involvement of skin, soft tissue and viscera. Histologically, the predominant tissue pattern is one of histiocytic proliferation with superimposed aggregate of eosinophiles.
2. Hand-Schüller-Christian disease comprises those cases in which not only skeletal involvement, singly or multiply, but also invol-

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vement of viscera, skin and soft tissue are present.

3. Letterrer-Siwe disease, constitutes those cases in which no bone lesions are present at the onset as determined by extensive examination; the disease being confined to skin, soft tissue and viscera. Histologically, a relatively uniform and monotonous histiocytic proliferation and lack of eosinophilic infiltrate are observed.

The following case represents a Hand-Schüller-Christian disease as evidenced both by the clinical course and by the examination of biopsy material.

Case report

H, an Indonesian boy, aged 2 years, was referred to the Department of Child Health, Dr. Tjipto Mangunkusumo General Hospital, Jakarta, on March 5, 1974 with the diagnosis of tuberculosis.

Anamnesis taken from his parents revealed that 4 months prior to admission the child suffered from intermittent fever. Two months afterwards, several swellings on his head were noted, which were followed by the protrusion of both eyes. In the mean time, there was limitation of movement of his neck especially to the left side and he began to walk inharmoniously. Initially, he was treated by his family physician as a patient with tuberculosis coxitis without any improvement. On the other hand the disease became progressive with the development of swellings described above.

Physical examination on admission revealed a conscious, walle-nourished boy with a body weight of 10.0 kg and a height of 82 cm. He was rather pale, neither dyspnoea nor cyanosis were found. Heart rate was 110/minute, pulse rate was 110/minute, regular and equal in quality, temperature was 36.5° C. Several soft, unpainful swellings were felt on his scalp and bilateral exophthalmos was seen. The head was slightly deviated to the right side associated with limitation in the movement of the neck. Small lymphnodes were felt in the submandibular and inguinal regions, which were rather firm in consistency, easily movable and were heard over both lungfields. Liver and spleen were not palpable. The extremities showed no abnormalities. Neurological examination showed no evidence of either cranial palsy or motoric dysfunction. Repeated tuberculin tests gave negative results. Laboratory examinations of the blood showed: hemoglobin 8 gm%, erythrocytes 2.8 million/cmm., hematocrit 24 volume%, reticulocytes 9‰, thrombocytes 296.000/cmm., leucocytes 10.000/cmm. with 3% metamyelocytes, 2% bands, 52% segmented neutrophils, 42% lymphocytes and 1% monocytes. Serum iron content was 90 ug%, the iron binding capa-

city was 260 ug% and the total iron binding capacity was 260 ug% and the total iron binding capacity was 350 ug%. Serum albumin 4.04 gm%, globulin 2.87 gm%, cholesterol 246 mg%, blood urea 42 mg%, alkaline phosphatase 14 Bodansky Units. Calcium and phosphor were 8.7 mg% and 5.3 mg% respectively. The Hijmans v.d. Bergh test either direct or indirect was negative. Immunoglobulin levels were within normal limits. Urine was yellow in colour, with a specific gravity of 1.008 and 750 ml. in amount. Neither protein, reducing substances nor bilirubin were detected and the sediment showed no particulars. The examination of stool showed neither ova nor parasites. Bone marrow aspirate disclosed a normal erythropoetic as well as granulopoetic activity and megakaryocytes were present too.

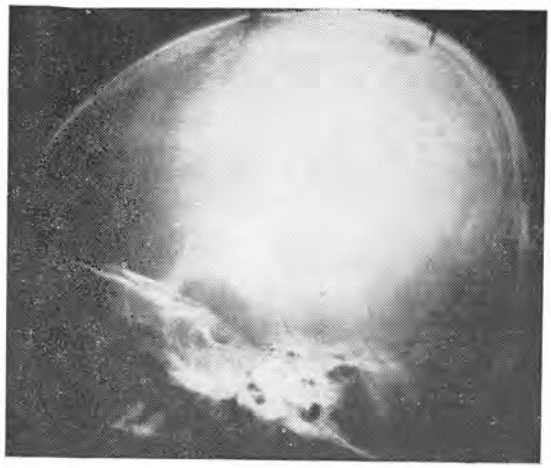
Radiographic examination of the bones revealed radioluscent areas presenting osteolytic processes within the skull, pelvic bone and femur (see figures 1a, 2 and 3).

The chest x-ray photo showed evidence of haziness in the right paracardial and retrocardial regions (see figure 4). Biopsy of the bone was carried out on the proximal part of the right femur where osteolytic lesion was present. Histopathologic findings of this specimen (P.A. No. 742216) strongly supported the diagnosis of Hand-Schüller-Christian disease (see figure 5).

The therapy consisted of prednisone in a dosis of 2 mg/kg of body weight and methotrexate, 75 mg twice a week; both were given orally. During his stay in hospital, improvement was gradually achieved. Complaint on the movement of the neck disappeared, his gait became normal and the protrusion of his eyes diminished. The swellings on his head and the enlargement of the lymphnodes also disappeared. Radiographic re-examination five and a half months later, showed diminution of the radioluscent areas in the skull, pelvic bone and femur (see figures 1b, 2 and 3), while the lung abnormality almost disappeared (see figure 4). The patient was discharged on request and the same treatment was continued at home. He was asked to come back for regular check up, but never showed up again.

Discussion

A 2-year-old patient with Hand-Schüller-Christian disease was described. The diagnosis was established on the clinical manifestations and histopathological findings. Diabetes insipidus as a symptom of the classic triad of Hand-Schüller-Christian disease was not present in this case. Actually, the triad consisting of exophthalmos, diabetes insipidus and skull defect does not necessarily exist simultaneously for the diagnosis (Love and Fashena, 1948; Blahd, et al., 1951; Bass et al., 1953). Mer-



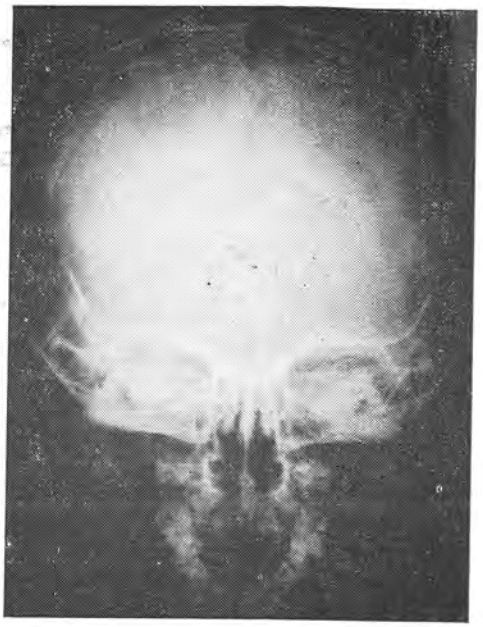
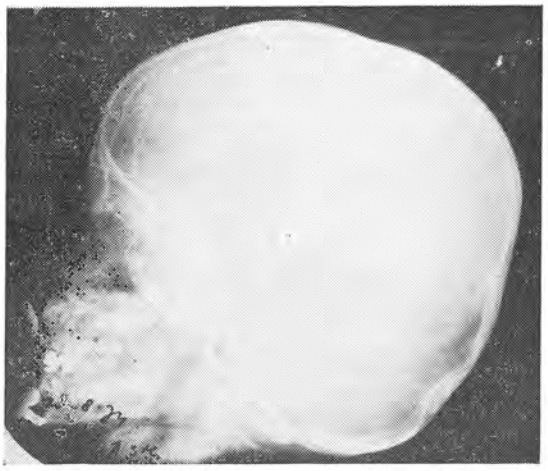


FIG. 1a : *X-ray photos of the skull presenting the osteolytic areas within the parietal and frontal bones.*



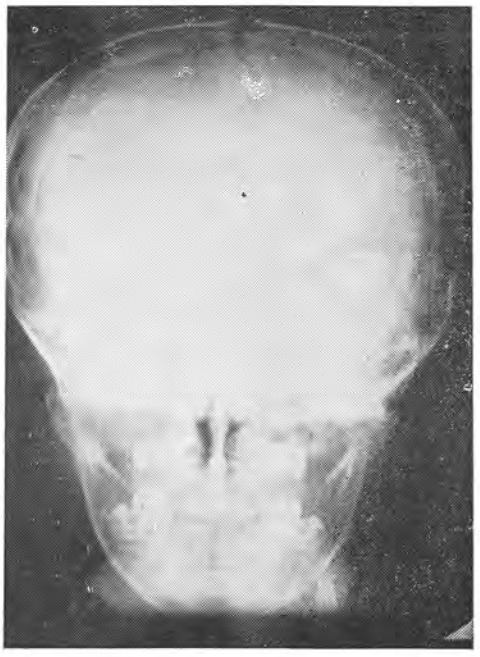


FIG. 1b : *Radiographic re-examination revealed the disappearance of the osteolytic lesions.*

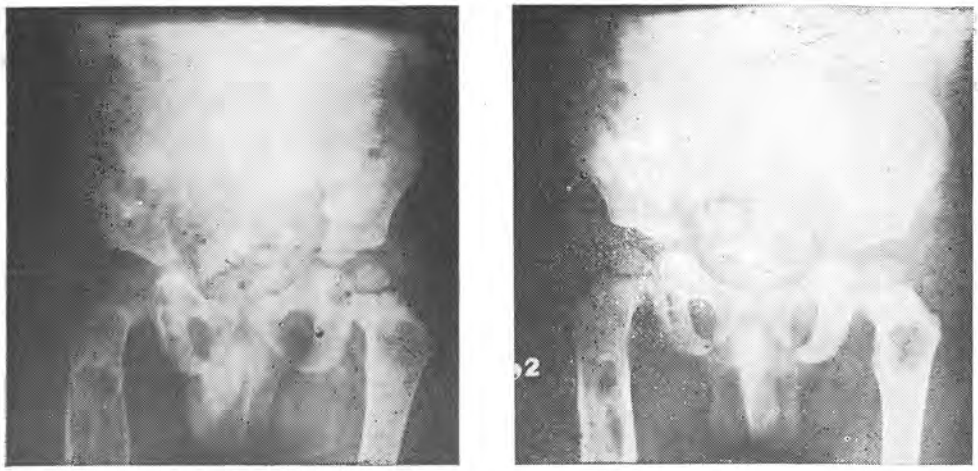


FIG. 2 : *X-ray photos of the pelvic bone, showing the osteolytic lesions (left) which diminished in size after therapy was instituted (right).*

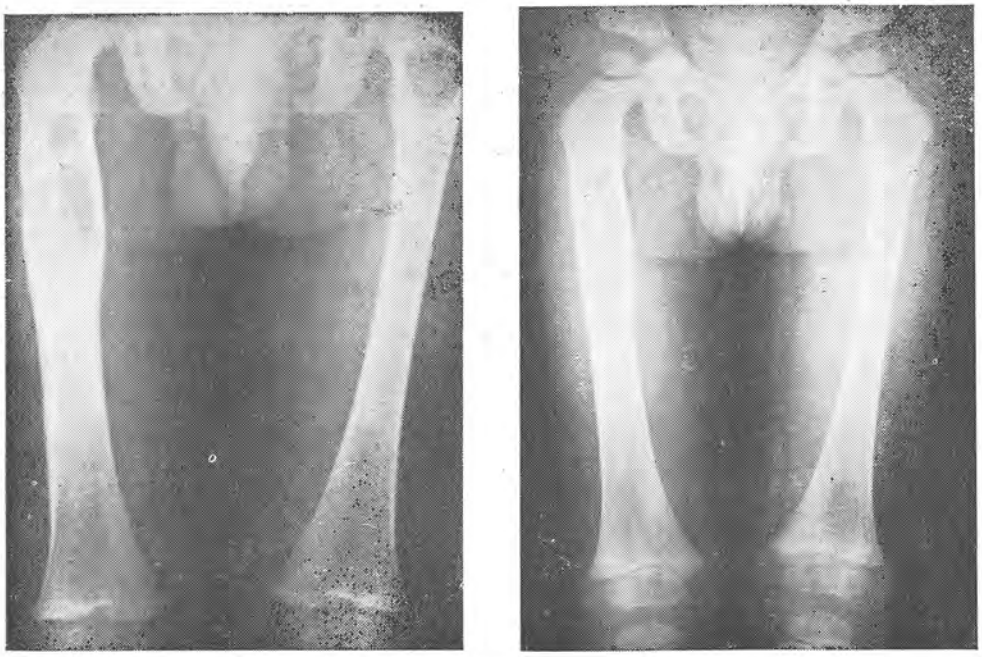


FIG. 3 : *X-ray photos of the femur obviously showed marked difference in size of the osteolytic lesions before and after treatment (left and right respectively).*

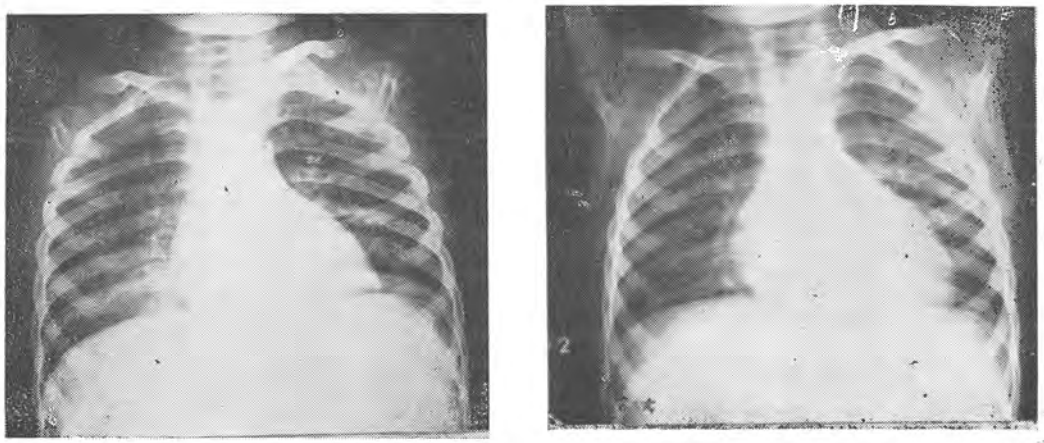


FIG. 4 : *Chest röntgenograms showed infiltrations in the right paracardial and retrocardial regions (left, before treatment), which became almost clear after treatment (right).*

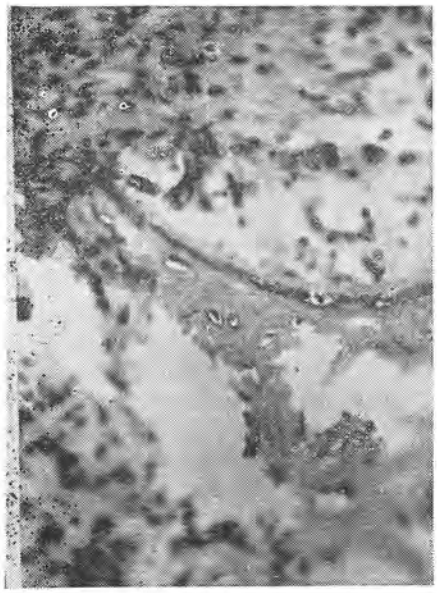


FIG. 5a

FIG. 5b

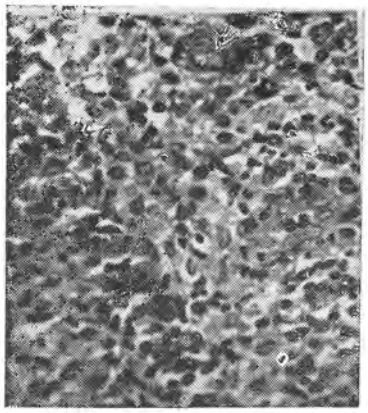


FIG. 5c

FIG. 5a, 5b and 5c : *Micrographs of the bone, showing the osteolytic areas beside histiocytic proliferations (Fig. 5a). Eosinophilic cell as well as foam cells are visible on fig. 5b and fig. 5c respectively (P.A. No. 742216).*

mann and Dargeon (1955) found only 2 cases with a complete triad out of 10 patients, whereas Avery and co-workers in 1957 found 3 out of 29 cases. Abbassy et al. (1958) found a case with skull defect which developed 3 months after the clinical onset of disease, followed by the appearance of exophthalmos and diabetes insipidus only two and a half years later. The bone lesions in this child were scattered in the skull, pelvic bone and proximal part of the femur. In spite of exophthalmos no lesions in the orbital bones could be detected.

Avery et al. (1957) found 2 cases with exophthalmos but without orbital lesions. On the other hand, one patient with multiple small destructive lesions in the orbita showed no exophthalmos. This case showed also bilateral pulmonary haziness, which almost completely resolved on the following chest photo. Pulmonary involvement in histiocytosis X has been reported earlier (Blahd et al., 1951; Avery et al., 1957; Renzetti et al., 1957; Green and Flaherty, 1960; Nadeau et al., 1960 and Lucaya, 1971; McNeill and Cameron, 1955).

Considering the benign nature of this disease, multiple courses of irradiation might result in a high cumulative dosage which might damage normal tissues as a consequence. Avery et al. (1957) preferred conservative treatment, while other workers (Blahd et al., 1951; Bass et al., 1953) reported good results with cortisone. Folic acid antagonists may successful use of vinblastine sulfate. cases treated by Mermann and Dargeon (1955). Beier and co-workers (1963), Siegel and Coltman (1966) were among the first to report the successful use of vinblastine sulfate. Recently, Starling et al. (1972) reported the use of vincristine, vinblastine and cyclophosphamide resulting in a complete remission rate of 50%, 20% and 36% respectively.

In our case, although subjective symptoms obviously disappeared on steroid and methotrexate treatment, it is very difficult to assess how long the result will last, since the patient never showed up again.

Summary

A case of Hand-Schüller-Christian disease was reported. The diagnosis was based on the clinical manifestations and histopathological findings. Subjective improvement was achieved on treatment with prednison and methotrexate.

Acknowledgement

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