CASE REPORT

Hand-Schüller-Christian Disease

by

*SJABAROEDDIN LOEBIS *, BISTOK SAING *, HELENA SIREGAR *, M. NAJIB LOEBIS ***

*(From the Department of Child Health * and Department of Histopathology ** University of North Sumatra, Medan)*

Abstract

A 14-month-old girl was admitted with signs of paleness, weakness and swelling of the lymphnodes in the neck and inguinal area. Histopathological examination of the lymphnodes showed histiocytosis X. X-photo of the skull and pelvic bones revealed several osteolysis.

Based upon clinical manifestations and histopathological examination this disease was diagnosed as the rare disease Hand-Schüller-Christian disease.

Received 4th September 1981.

Introduction

Eosinophilic granuloma of the bone, Hand-Schüller-Christian disease and Letterer-Siwe disease have formerly been considered as a distinct entity.

Lichtenstein (1953) however have concluded that these three conditions represented variations of the same basic disease process.

Lichtenstein 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.

This was a rare disease with an incidence of one to two million people, or one to three hundred fifty thousand children under the age of twelve years old (Cheyne, 1971).

It was first reported in Indonesia in 1974 by Widagdo et al.

Oberman (1961) distinguished three groups corresponding rougher to three classic categories based upon the clinical and radiological findings, namely :

1. Eosinophilic granuloma of the bone; constitutes those case in which the disease is completely confined to the skeleton as a single or multiple lytic lesions at the onset without evidence of involvement of the skin, soft tissue and viscera. Histologically the predominant tissue pattern is one of histiocytic proliferation with superimposed agregate of eosinophils.
2. Hand-Schüller-Christian disease comprises those cases in which not only skeletal involvement, present singly or multiply, but also involvement of the viscera, skin and soft tissue.
3. Letterer-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 tissue and viscera. Histologically a relative uniform and monotonus histiocytic proliferations and lack of eosinophilic infiltrate are observed.

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

Case report

M, an Indonesian girl aged 14 months was refered to the Department of Child Health Pirngadi Hospital on the 28th January 1980 with the complaint of paleness and weakness during the last two months.

Three months prior to admission there were swellings of lymphnodes in the neck and inguinal area which did not subsides after treatment, but become bigger with a diameter between 2 and 6 cm. (Fig. 1).

Birth History

She was born in Pirngadi Hospital with a birth weight of 2800 grams, the youngest of two siblings.

Immunization

She had been vaccinated against Tuberculosis, Smallpox and DPT.

Physical examination

A pale looking and undernourished girl, aged 14 months, with a body weight of 7,0 kg and a height of 70 cm. (Fig 1).

Pulse rate was 128x/minute regular and equal in quality.

The body temperature was 37° C.

Bilateral exophthalmus was found and the light and corneal reflexes were normal.

On the neck several swellings of the lymphnodes were felt with rather firm consistency. Diameter between 2 to 6 cm (Fig 3) the heart rate was 128/minute, regular, no murmur.

The respiration rate was 24/minute.

The liver was palpable 3 cm below the costal arch, with rather firm consistency but not painful.

The spleen was not palpable.

There were also several unpainful swellings of the lymphnodes in the left and right axilla and in the right inguinal area (Fig. 4).

The diameter of the enlarged lymphnodes were between 2 to 6 cm.

Laboratory examinations of the blood showed that the haemoglobin level was 4,5 gm%, thrombocytes 296000/Cmm, leucocytes 10.000/Cmm with 2% bands, 73% segmented neutrophils, 23% lymphocytes, 3% monocytes, anulocytes (+), anisocytosis (+), hypochrom (+). The serum albumin was 2,618 gm% (hypoalbuminemia), and serum globulin 3,63 gm%.

The urine was yellow in colour with a specific gravity of 1,008 and 750 ml in amount, neither protein nor reducing substances were detected and the sediment showed no abnormalities.

The examination of the stool showed no ova and parasites.

Bone marrow aspirate disclosed a normal erythropoetic as well granulopoetic activity.

Biopsy of the lymphnodes in the inguinal area was carried out, and showed a uniform monotonous histiocytic proliferation.

The histopathological diagnosis was histiocytosis X. (Fig. 7A, 8B). Radiographic examinations of the bone revealed radioluscent areas presenting osteolytic processes within the skull and pelvic bone with a diameter of 1 to 3 cm. (Fig. 2, 5, 6).

From the clinical symptoms, the patients diagnosis was Hand-Schüller-Christian disease with severe anaemia and moderate malnutrition.

Treatment

She was given a blood transfusion for the anaemia, and the Hb was elevated to 9 gm%.

She had also received prednison 2 mg/kg body weight.

After three days of admission the patient went home upon request of her own parents.

Though advised to come back for further treatment, she did not show up again.

Discussion

A 14 months old patient with Hand-Schüller-Christian disease was described. The diagnosis was established upon clinical manifestations, radiographic examinations and histopathological findings.

Diabetes insipidus as a symptom of the classic triad of Hand-Schüller-Christian disease was not present in this case.

The triad consisting of exophthalmus, diabetes insipidus and skull defect does not necessarily exist simultaneously for the diagnosis.

Mermann and Dargeon (1955) found only two cases with a complete triad out of ten patients.

Avery et al. (1957) found only three cases with diabetes insipidus in twenty nine cases with Hand-Schüller-Christian disease. Sims (1977) found forteen cases of diabetes insipidus in forty three cases. Hand-Schüller-Christian is a rare disease with an incidence of one in two million people.

In North England in twenty nine years only forty three cases were reported. Up till now the etiology of this disease is not clear, no microorganism was detected in the osteolesions. Some workers suggest that this disease
logical disease but it has not been proven.

The disease can spread to the other organs involving the liver, spleen as well as the brain which take several months to several years.

Medical research is still being done to solve the etiology of this disease, Blahd (1951) used steroid and reported good results.

Beier et al., (1963), Siegel and Coltman (1966) were among the first to use vinblastine Sulfate successfully. Starling et al., (1972) reported the use of vincristine, vinblastine and cyclophosphamide resulting a complete remission rate of 50%, 20% and 36% respectively. In England several methods have been used to treat Hand-Schüller-Christian disease including antibiotics for the secondary infections and surgery for the osteolytic lesions. Cytostatics was given to prevent metastasis to other organs, while radiotherapy was given to prevent lesions in the bones, swellings of the lymphnodes and spreading to other organs.

Pitressin was given to those patients with diabetes insipidus, and growth hormon in patients with growth retardation to promote growth.

On the other hand, spontaneous remission had also been reported in cases who had already metastasis to other organs.

Acknowledgements

The authors gratefully acknowledged the assistance and the kind cooperation of the Department of Histopathology and Radiology of the Medical School, University of North Sumatera.

REFERENCES

1. AVERY, M.E.; Mc AFEE, J.G. and GUILD, H.G.: The Course and prognosis of reticuloendotheliosis (eosinophilic granuloma, Schüller-Christian disease and Letterer-Siwe disease). Amer. J. Med. 22 : 636 (1957).
2. BEIER F.R.; THATCHER, L.G. and LAHEY, M.E. : Treatment of reticuloendoiheliosis with vinblastine sulfate J. Pediatr. 63 : 1087 (1963).
3. BLAHD, W.H.; LEVY, M.S. and BASSETT, S.H. : A case of Hand-Schüller Christian syndrome treated with cortisone. Ann. intern. Med. 35 : 927 (1951).
4. CHEYNE, C. : Histiocytosis X. Journal of bone and Joint Surgery 538 : 366 - 382 (1971).
5. LICHTENSTEIN, L : Histiocytosis X. Integration of eosinophilic granuloma of bone "Letterer-Siwe disease" as related manifestations of a single nosologic entity. Arch. Pathol. 56 : 84 (1973).
6. MERMANN, A.C. and DARGEON, H.W. : The management of certain non lipid reticuloendoiheliosis. Cancer 8 : 112 (1955).
7. OBERMAN, H.A. : Idiopathic histiocytosis X. A clinicopathologic study of 40 cases and review of the literature on eosinophilic granuloma of bone. Hand-Schüller-Christian disease and Letterer. Siwe disease. Pediatr. 28 : 307 (1961).
8. SIEGEL, J.S. and COLTMAN, C.A. : Histiocytosis X. Response to vinblastine sulfate. J. Amer. Ass. 1971 : 123 (1966).
9. STARLING, K.E.; DONALDSON, M.H.; HAGGARD, M.E.; VIETTI, T.J. and SUTOW, W.W. : Therapy of histiocytosis X with vincristine, vinblastine and cyclophosphamide. Amer. J. Dis. Child. 123 : 105 (1972).
10. SIMS, D.G. : Histiocytosis X. Arch. Dis. Child. 52 : 433 - 440 (1977).
11. WIDAGDO; SADELI, R.; WAHIDIYAT, I. : Hand-Schüller-Christian disease (Case Report). Pediatr. Indon. 14 : 218 - 223 (1974).

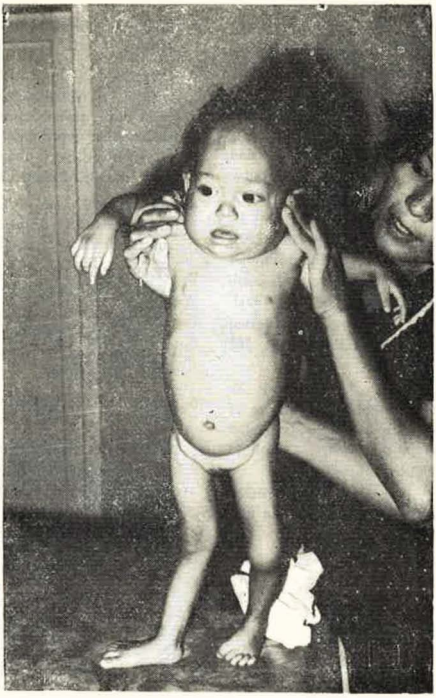

FIG. 1. *The girl with her mother. Note the swelling of the lymphnodes in the axilla and inguinal.*

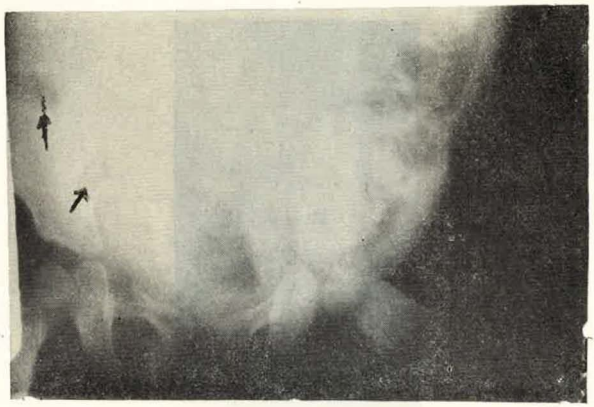

FIG. 2. *Several osteolytic lesions in the pelvic bones*

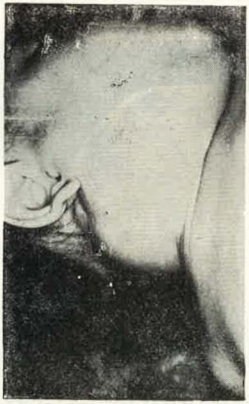

FIG. 3. *Lymphnodes swellings in the neck, the greatest diameter was 6 cm.*

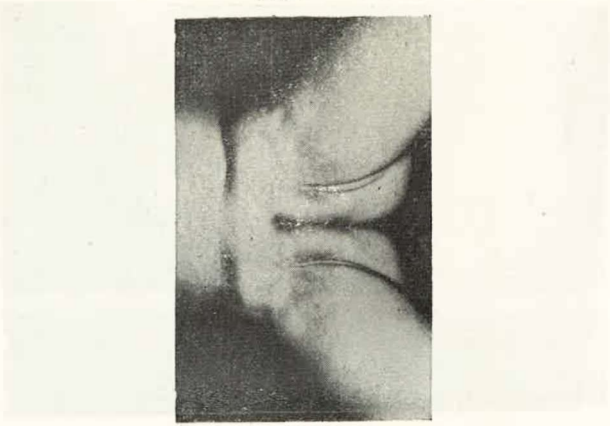

FIG. 4. *Several swellings of the lymphnodes in the inguinal area, diameter between 2 cm to 6 cm.*

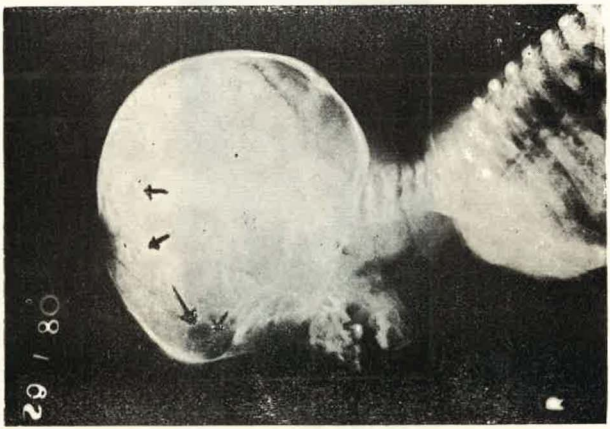

FIG. 5 : *Several osteolytic lesions in the parietal bone, diameter 1 cm - 3 cm.*

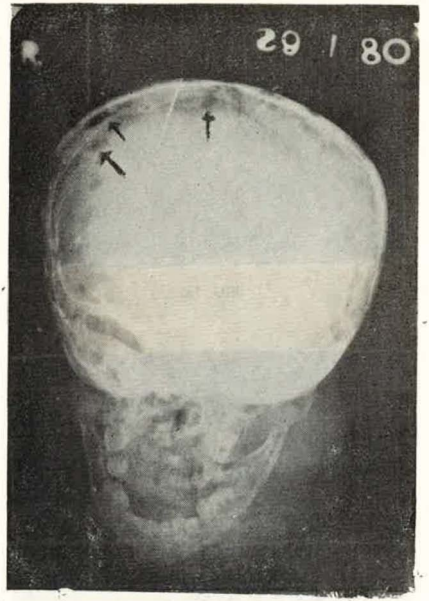

FIG. 6. *Several osteolytic lesions in the frontal bone, diameter 1 cm - 3 cm.*

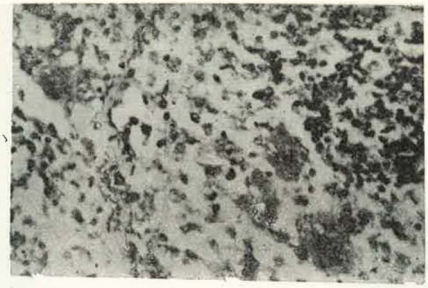

FIG. 7A.

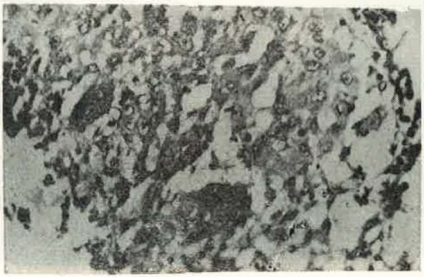

FIG. 7B.

Fig. 7A and 7B Photomicrography of the lymphnode tissue showing proliferation of the histiocytes.