CASE REPORT

A CASE REPORT OF HAND SCHULLER CHRISTIAN DISEASE

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HOW TO CITE THIS ARTICLE:

BC Sharath Kumar, Swetha Gowda, Adarsh Gowda, MG. Gopal, M Ramesh. "A case report of hand schuller christian disease". Journal of Evolution of Medical and Dental Sciences 2013; Vol. 2, Issue 43, October 28; Page: 8255-8258.

ABSTRACT: AIM: Hand-Schüller-Christian Disease is a rare proliferative disorder in which pathological Langerhans cells (LCs) accumulate in a variety of organs. It represents triad of diabetes insipidus, bone lesions and exophthalmos. Usually begins at the age of 2 and 6 years. 30%- develop skin lesions and 80%- develop bone lesions (cranium being preferentially involved). **MATERIAL & METHOD:** A 5-year-old boy was admitted to the Kempegowda Institute of medical sciences hospital, department of orthopaedics with pain in left hip since 2 weeks. The child was small for age, cachexic, mildly lethargic but occasionally irritable. He was referred to skin OPD for seborrheic dermatitis like picture over the scalp and multiple hypopigmented macules over the abdomen. Patient also had multiple boggy defects in the skull and exophthalmos and also history of long term continuous ear discharge and on and off fever. Cervical, axillary and inguinal lymphadenopathy were present. **RESULT:** Skin biopsy findings were consistent with histiocytosis. CD1a and S100 tumor markers were positive. Patient was put on chemotherapy-ETOPOSIDE+CYTOBLASTIN+BETAMETHASONE REGIMEN to which he responded successfully. **CONCLUSION:** Disorders like HAND-SCHÜLLER-CHRISTIAN DISEASE detected early can be effectively managed improving the overall survival, quality of life and disability free living.

KEYWORDS: Hand-Schuller-Christian Disease, Langerhans's Cells, Seborrheic Dermatitis, Exophthalmos.

INTRODUCTION: Langerhans cell histiocytosis is the name proposed by Risdall et al,(1)in 1983,for a group of disorders previously called histiocytosis X; where langerhans cells play a central role in the pathogenesis. Hand-schuller-christian disease is a rare entity comprising of exophthalmos, diabetes insipidus and geographic map skull. However, the first case was described by Hand (2) in 1893. Schuller (3)(1915) and Christian(4,5)(1920) had called in attention to this triad. The other types are Letterer siwe disease which is an acute, disseminated, multisystem, often fatal type of langerhans cell histiocytosis. Eosinophilic granuloma is chronic, benign and solitary and the mixed type(6).

CASE REPORT: A 5-year-old boy was admitted to the Kempegowda Institute of medical sciences hospital, department of orthopaedics with pain in left hip since 2 weeks. He was referred to skin OPD for seborrheic dermatitis like picture over the scalp (fig.1) and multiple hypopigmented macules over the abdomen (fig.2) Patient also had exophthalmos (fig.3) and also history of long term continuous ear discharge and on and off fever. At 2yrs 6months patient gives past history of right hip joint and was found to have an osteolytic lesion in the right ischium (fig.4) for which he was treated with aspiration and steroid infiltration. On examination, the child was small for age, cachexic, mildly lethargic but occasionally irritable. Cervical, axillary and inguinal lymphadenopathy were present. Following investigations were carried out:

- Complete blood reports were within normal limit except ESR-56mm/hr.

CASE REPORT

- LIVER FUNCTION TEST-normal except for serum alkaline phosphatase-197 IU/l.
- SERUM ELECTROLYTES-normal
- SERUM URIC ACID-normal
- SKIN BIOPSY-epidermis shows focal atrophy (fig.5). Papillary dermis shows sheets of cells comprising of predominantly histiocytes with abundant cytoplasm and vesicular nucleus. Several eosinophils were also noted (fig.6).
- X-Ray Skull: multiple boggy defects-geographic pattern (fig.7), (fig.8)
- X-Ray pelvis: osteolytic lesion in the left ilium (fig.9)
- CD1a and S100 tumor markers positive
- ULTRASOUND ABDOMEN AND PELVIS: Normal study
- BONE MARROW ASPIRATION: Normal

Clinical diagnosis of histiocytosis X was made and patient was started on chemotherapy: Etoposide + cytarabine+betamethasone regimen, to which he responded successfully. (fig.10), (fig.11), (fig.12), (fig.13).

DISCUSSION: Histiocytosis X is a clonal proliferation disease of langerhan's cells that express an immunophenotype positive for S100 and CD1a(7) Hand-schuller-christian disease is a well described entity which has appeared sporadically till 1935.the cases have been reported from the western literature but authentic case reports from India are few(8,9). It classically refers to the clinical triad of skull lesions (80%), exophthalmos and diabetes insipidus and the classical triad is only found in 10% cases. In this case the patient had skin involvement in the form of hypopigmented macules which is seen in only 30% of the patients. He also presented with chronic otitis media of both the ears which is another feature. According to the present concept, the syndrome of hand schuller Christian disease, letterer siwe disease and eosinophilic granuloma are considered a clinical variety of a common pathological process (10, 11). Our case has a favourable prognosis due to non-involvement of the lungs and liver. (12) Chemotherapy coupled with radiation to localized bony lesions and supportive measures are useful in treatment, there being no unified therapeutic measures.(13)

REFERENCES:

1. Risdall AJ, Dehner LP, Duray Pet al. Histiocytosis X (LC histiocytosis) Prognostic role of histopathology. Arch Pathol Lab Med 1983; 107-109.
2. Hand, A.: General tuberculosis. Trans. Path. Soc. Philadelphia. 16: 282, 1893 and Polyuria and tuberculosis. Arch. Paediat., 10: 673, 1893.
3. Schuller, A.: Ueber eigenartige schadeldefekte in Jugendalter. Fortschr. a. d. Geb. d. Roentgenstrahlen, 23: 12, 1915-1916.
4. Christian, H. A.: Defects in membranous bones, exophthalmos and diabetes insipidus. An unusual syndrome of dyspituitarism-a clinical study. Contrib. Med. & Biol. Res., 1: 390, 1919 (Hoeber, New York)
5. Christian, H. A.: Defects in membraneous bones, exophthalmos, and diabetes insipidus-An unusual syndrome of dyspituitarism. Med. Clin. North Amer., 3: 849, 1919-1920. (As quoted by Liberman et al 1969)

CASE REPORT

6. Berry DH, Becton DL. Natural history of Histiocytosis X. Hematol Oncol Clin North Am 1987;1 :23
7. William CL, Busque L, Griffith BB et al. Langerhans cell histiocytosis-a clonal proliferative disease. N Engl J Med. 1994; 331:154-60
8. Dahat, H. B., Ingle, V. N. and Patil, S. D.: Histiocytosis "-X". Ind. J. Paediat., 41: 149-152, 1974.
9. Thomas, P. A.: Hand-Schuller-Christian disease (Histiocytosis). Ind. J. Surg., 26: 264-269, 1964
10. Avery, M. E., McAfee, J. G. and Guild, H. G.: The course and prognosis of reticulo-endotheliosis (Eosinophilic granuloma, Letterer-Siwe disease and Hand Schuller-Christian disease)-A study of 40 cases. Amer. J. Med., 22: 636-652, 1957.
11. Henderson, J. I.: Histiocytic reticulosis - A review of 20 patients in paediatric age group. Med. J. Austr., 2: 485-488, 1969
12. Hamilton, J. B., Barner, J. L., Kennedy, P. C. and McCort, J. J.: Osseous manifestations of eosinophilic granuloma; report of cases. Radiology, 47: 445-456, 1946
13. McClelland J, Pritchard J, Chu AC. Current controversies. Hematol Oncol. Clin North Am 1987;1:147

FIG.1: Seborrheic dermatitis of the scalp

Fig.2: Hypopigmented macules

Fig.3: Exophthalmos

Fig.4: X-ray showing osteolytic lesion in right ischium

Fig. 5: low power

Fig. 6: high power

Fig. 5 and fig.6: Histopathology showing epidermal atrophy. Papillary dermis shows sheets of cells comprising of predominantly histiocytes with abundant cytoplasm and vesicular nucleus. Several eosinophils were also noted.

CASE REPORT

Fig. 7 & fig. 8: X-ray showing geographic skull

Fig. 9: X-ray pelvis showing new osteolytic lesion in the left ilium

Fig.10: post-chemotherapy status

Fig.11: seborrheic dermatitis of the scalp healed

Fig.12:post-chemotherapy X-ray skull

Fig.13: post chemotherapy X-ray pelvis

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Date of Submission: 11/10/2013.

Date of Peer Review: 12/10/2013.

Date of Acceptance: 19/10/2013.

Date of Publishing: 22/10/2013