CASE REPORT

LANGERHAN CELL HISTIOCYTOSIS WITH MULTISYSTEM INVOLVEMENT IN AN ADULT
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ABSTRACT

Langerhan cell histiocytosis (LCH) is a rare disorder that primarily affects children. Its occurrence in adults is very rare. We report a case of 37-year-old male patient who presented with complaints of increased thirst, excessive passage of urine, shortness of breath and skin lesions. The diagnostic workup revealed endocrine involvement with diabetes insipidus, restrictive lung disease, skin biopsy consistent with LCH, bone and periodontal involvement. The skin lesions responded well to Psoralen- Ultraviolet Radiation A therapy (PUVA). He showed general improvement on systemic chemotherapy.

Key words: Langerhan cell histiocytosis, adult, diabetes insipidus, pulmonary fibrosis, cutaneous lesions.

INTRODUCTION

The histiocytoses are a group of diseases characterized by accumulation of reactive or neoplastic histiocytes in various tissues. Langerhan cell histiocytosis (synonyms). Histiocytosis X, Letterer-Siwe disease, Hand-Schuller-Christian syndrome, Hashimoto-Pritzker syndrome) is a rare disorder that primarily affects children but is also found in adults. One in 20,000 children are diagnosed each year and about 1 in 560,000 adults are affected. LCH is a reactive condition in which cells with a phenotype of Langerhan cells accumulate in various tissues and cause damage to tissues. The Langerhan cell is a histiocytic cell that represents a resident immigrant of bone marrow origin. The aetiology of LCH is unknown. Nearly any system of body can be involved. A patient may have single system or multisystem involvement. We report a case of a Pakistani adult patient suffering from LCH with multisystem involvement.

CASE REPORT

A 37 years old male patient presented with history of increased thirst and excessive passage of urine for five years, shortness of breath for four years and skin lesions for three years. On examination he was found to be anaemic with first degree clubbing of fingers. There was decreased chest movement and dull percussion note with decreased air entry in lower chest bilaterally. There was gingival swelling with loss of few teeth. Skin examination revealed diffuse erythematous patches covered with adherent greasy yellowish scales on scalp and retroauricular areas. Flexural skin rash showed adherent greasy yellowish scales with ulceration and purpuric pigmentation. Scattered yellow brown scaly purpuric papules were present on chest, abdomen and back. In the groin and gluteal region, pigmented scaly plaques were present with ulceration in perianal area. The patient was thoroughly investigated. His haemoglobin was 11.7 gm/dl and erythrocyte sedimentation rate was 39 mm in 1st hour. Random blood sugar, urea, creatinine, electrolytes and liver function test were within normal limits. Montoux test was negative. Urine detailed report showed a specific gravity of 1.002. Serum osmolality was 286 m osm/kg - (reference 278-305 m Osm/kg) and urine osmolality was 100 m Osm/kg - (reference 350-1010 m Osm/kg). Water deprivation test was positive. Patient responded to desmopressin indicating a cranial cause of diabetes insipidus. Specific gravity of urine became 1.025 after treatment with desmopressin. Venereal disease research laboratory test (VDRL), Treponema pallidum
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haemagglutination test (TPHA) and tests for HIV were negative. T3, T4, TSH, angiotensine converting enzyme, antinuclear antibodies and serum calcium were within normal limits. Hepatitis B surface antigen and anti-HCV were nonreactive. Ultrasound of abdomen and pelvis was normal. Skin swab and scraping were negative for bacterial and fungal microscopy and culture. Pulmonary function tests showed a restrictive lung disease with no reversibility. Electrocardiogram and echocardiography were normal. X ray chest and high resolution CT scan chest was suggestive of pulmonary fibrosis. X-ray skull and mandible was done and lytic lesions were seen in mandible. Bone scan showed photon deficient area in the body of left mandible and frontal region of skull indicating osteoclastic activity. Orthopantmogram (OPG) showed loss of multiple teeth and lucent lesions in body of mandible on left side. Magnetic Resonance Imaging (MRI) brain and pituitary fossa was normal. However there was an area of bone edema in frontal bone. Skin biopsy (scalp and axilla) was done. The histological appearance was consistent with the diagnosis of Langerhan cell histiocytosis. Also the lesional cells were found to be positive for S-100 protein. Patient was put on PUVA therapy. Skin lesions improved. As there was multisystem involvement, he was referred to oncology department for systemic chemotherapy. Presently he is taking systemic chemotherapy and condition has improved.

DISCUSSION

Langerhan cell histiocytosis in adults is a very rare disorder (1 in 560,000). Diabetes insipidus may be the presenting feature of LCH. Also there may be growth hormone deficiency and thyroid involvement. In adults LCH, skin and lung involvement is common. The most characteristic presentation is with scalp involvement as erythematous areas with greasy scales resembling seborrhoic dermatitis. On the trunk, the lesions are discrete yellow brown scaly papules with purpuric areas. Lung involvement is invariably associated with smoking. The symptoms include shortness of breath, chest pain, dry cough and in extreme cases lung collapse. Solitary bone involvement is common. The commonest sites are bones of calvarium but femur, scapula, rib, mandible and vertebrae are often affected. Periodontal involvement affecting particularly the lower molar areas may occur. Involvement of ear is common. External ear, middle ear and mastoid may be affected. Nail changes include paronychia, nail fold destruction, onycholysis and subungual expansion with nail plate loss. Lymph node involvement with chronic draining sinuses, hepatomegaly and splenomegaly may also be seen. An initial diagnosis of LCH is made on clinical and histological grounds. Diagnostic confidence increases if lesional cells are found to be positive for markers e.g. S-100 protein, peanut agglutinin or D-mannosidase. Definite diagnosis (according to criteria of Histiocytic Society) is made if lesional cells show positive staining with anti-CD1 antibodies or exhibit Birbeck granules on electron microscopy. Treatment in isolated bone disease includes curettage or intralesional steroid injection may be given in symptomatic weight bearing bones. If optic nerve or spinal cord is involved, low dose radiotherapy is advised. In single system skin disease, options include 20% nitrogen mustard, PUVA therapy and thalidomide. In multisystem LCH, systemic chemotherapy may be advised. Response rates of about 50-70% can be achieved using systemic chemotherapy.

Prognosis of LCH depends on age of patient, extent of disease and presence of vital organ failure.

REFERENCES


