BILATERAL EXOPHTHALMOS AND LYMPHOBLASTIC ALEUKAEMIC LEUKAEMIA*†

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AMONG sixteen cases of acute leukaemia with orbital leukaemic infiltration in children, there were six myeloblastic, six monoblastic, two lymphoblastic (Mortada 1962, 1963, 1964a), one undifferentiated stem-cell, and one aleukaemic leukaemia.

The following case of lymphoblastic aleukaemic leukaemia is described because bilateral proptosis in this condition has been only rarely reported, and because in cases of malignant lymphoma a sternal bone marrow puncture is necessary to exclude aleukaemic leukaemia.

Case Report

An 18-months-old male infant had bilateral proptosis of 3 weeks' duration. There was no relevant family history. No drugs such as pyrimidon or antibiotics had been given nor had any form of irradiation been applied.

X-ray films of the skull and orbits, and the urine, faeces, and blood pressure were normal. The Wassermann reaction was negative. There were no septic foci in the body.

The child was pale, with body temperature 38°C. and pulse 100/min. The gums bled easily. The liver was not enlarged, but the spleen was enlarged one finger's breadth below the costal margin, and the cervical lymph glands were also enlarged.

After one week the proptosis in both eyes had progressed to 24 mm. (Figure), with limitation of ocular movements. Both fundi showed dilated tortuous retinal vessels, and orbital masses were felt above each globe.

FIGURE.—Bilateral proptosis with lymphoblastic aleukaemic leukaemia in a male infant aged 18 months.

Blood Count.—Haemoglobin 40 per cent.; red blood corpuscles 2,500,000 per c.mm.; white blood corpuscles 7,000 per c.mm.; basophils 0 per cent., eosinophils 0 per cent., staff nucleated 2 per cent., segmented 20 per cent. (total polymorphs 22 per cent.); lymphocytes 75 per cent., monocytes 3 per cent., platelets 16,000.

Biopsy.—Tissue from the left orbital mass fixed in formalin and stained with haematoxylin and eosin showed large cells of varied shapes held in a fine reticular stroma. The nuclei were large, of various shapes, sizes, and staining properties. The histopathological picture resembled that of a reticulo-sarcoma.

Bone Marrow.—As the blood count showed no leukaemia but only lymphocytosis, a sternal bone marrow count was performed. Basophils 0 per cent., eosinophils 3 per cent., blast cells 28 per cent., promyelocytes 1 per cent., myelocytes 4 per cent., juveniles 2 per cent., staff nucleated

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4 per cent., segmented 13 per cent., lymphocytes 32 per cent., monocytes 0 per cent., megaloblasts 0 per cent., macroblasts 2 per cent., normoblasts 10 per cent., reticulum cells 1 per cent. Peroxidase staining did not show oxidase granules in the 28 per cent. blast cells.

As the preponderating cell in both blood and bone marrow was the lymphocyte and as the blast cells in bone marrow did not show oxidase granules after peroxidase staining, the blast cells were interpreted as lymphoblasts and the case was diagnosed as one of lymphoblastic aleukaemic leukaemia.

Treatment.—The orbital tumours were irradiated and supportive blood transfusions, vitamins, and corticosteroids were given, but the child died one month later.

Discussion

In aleukaemic leukaemia, although the total cell count is relatively normal, the differential count usually shows some abnormal cells or preponderance of a particular cell which gives a clue to the diagnosis (Britton, 1963). In this case the preponderance of lymphocytes in blood and bone marrow favoured the diagnosis of lymphoblastic aleukaemic leukaemia. Most writers have stressed the fact that most cases of aleukaemic leukaemia are of the lymphatic type (Cappell, 1958; Best and Taylor, 1961; Falkenstein and Fowler, 1943).

In sections fixed in formalin and stained with haematoxylin and eosin, it is difficult to differentiate lymphoblastic, monoblastic, myeloblastic, or reticulosarcoma infiltrations. All differentiation is easier with Giemsa stain (Mortada, 1964b).

Summary

(1) This case of bilateral exophthalmos with lymphoblastic aleukaemic leukaemia is the first to be described from Egypt.

(2) In cases of malignant lymphoma of the orbit, lacrimal gland, conjunctiva, and lids, a sternal bone marrow puncture must be performed to exclude the possibility of aleukaemic leukaemia.

(3) The orbit contained many dormant reticulum cells, and is commonly affected in acute leukaemia.

(4) Most cases of aleukaemic leukaemia are of the lymphoblastic type.

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