CASE NOTES

CHLOROMA OF THE ORBIT*

BY

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CHLOROMA is a rare type of leukaemia, and the process is nearly always acute. It is regarded as an atypical form of myeloblastic leukaemia and has a similar relation to myelogenous leukaemia to that between lympho-sarcoma and lymphatic leukaemia. The disease is commonly found in children and young adults, but it has occasionally been reported in the middle aged, chiefly in males. A congenital case has also been reported (Morrison, Samwick, and Rubinstein, 1939).

The lesion manifests itself as a localized mass, the sites of predilection being the bones of the skull and thorax. There may be chloroma masses in the kidney, muscle tissue, and skin, no tissue in the body being exempt except the brain and spinal medulla (Brannan, 1926). These tumours which are characteristically of a greenish colour are detected clinically under the periosteum, but the primary lesion is probably in the marrow cavity, the periosteal tumours being secondary. Some authors consider that the pigment, which fades on exposure to air, is of lipoid origin (Chiari, 1883; Malkin, 1925); others believe it to be haematogenous, and Goodman and Iverson (1946) say that it is chemically related to porphyrin. At autopsy it is found to be homogeneously distributed in the connective tissue, mucosal surfaces, and endothelial linings. Humble (1946) considers that it represents an intermediate stage in the breakdown of haemoglobin to bilirubin. The cells within the lesion are apparently identical with those in the circulating blood, and the symptoms are the same as in acute leukaemia with the addition of those produced by the tumour. The disease is always fatal within a few months.

Case Report

Case 1, a boy aged 11 years, was admitted to the Gandhi Eye Hospital, Aligarh, with proptosis of the left eye for one month and continuous fever for 6 days. After admission the right supra-orbital region began to swell. There was nothing significant in his past history or family history.

Examination.—The boy was poorly nourished with pallor of the skin. There was no bleeding from gums or nose. The temperature was 100°F. The spleen and liver were

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palpable one finger's breadth below the costal margin. The submandibular lymph glands were palpable. The glands in the axilla, neck, and inguinal regions were not enlarged.

Left Eye.—There was a swelling of the upper and lower lids extending up to the supra-orbital margin. The eyeball was proptosed down and out (Fig. 1). The swelling was firm and tender on pressure. The overlying skin was congested, of a bluish colour, and the veins prominent. Movements of the eyeball were limited in all directions. There was chemosis of the bulbar conjunctiva, and the cornea, exposed as a result of lagophthalmos, was becoming opalescent. The anterior chamber was normal, the pupil and reflexes were normal, and the lens and vitreous clear. The swelling made vision impossible. The optic disc was pale, and the veins prominent.

FIG. 1.—Case 1, showing marked proptosis of left eye, oedema of lids, and supra-orbital swelling. A small swelling can be seen on the right eye also.

Right Eye.—The upper and lower lids were swollen and congested, with tenderness in the upper and inner angle of the eye. The swelling was firm. Movements were normal in all directions. The cornea was normal. The visual acuity was 6/12.

The optic disc was slightly pale, but the patient was not co-operative, and details were not seen clearly.

The cardiovascular, respiratory, and nervous systems were all normal.

Peripheral Blood

Total white cell count	22,250 cells/c. mm.
Blast cells	18 per cent.
Promyelocytes	4 per cent.
Myelocytes	2 per cent.
Metamyelocytes	3 per cent.
Staff cells	3 per cent.
Polymorphs	26 per cent.
Lymphocytes	34 per cent.
Monocytes	10 per cent.

General Blood Picture

Marked anisocytosis, slight poikilocytosis, moderate polychromasia, apparently very few platelets.

Total red blood count	2.16 million per c.mm.
Haemoglobin	7.5 g. per cent.
Platelets	60,000 per c.mm.
Packed cell volume	20 per cent.
Mean corpuscular volume	92 c.μ.
Mean corpuscular haemoglobin	34 μμg.
Mean corpuscular haemoglobin concentration	37 per cent.

Urine.—Bence Jones protein absent. Albumin present in traces.

Sternal Bone Marrow Aspiration, Fragments Hyperplastic

Blast cells	30
Promyelocytes	6
Myelocytes	10
Metamyelocytes	5
Staff cells	6
Polymorphs	20
Lymphocytes	9
Monocytes	2
Undifferentiated cells	4
Plasma cells	2
Early normoblast	Nil
Intermediate normoblast	2
Late normoblast	4
Megakaryocytes	Nil
Leuco-erythroblastic ratio	15:6:1

Case 2, a boy aged 6 years, was admitted to the Gandhi Eye Hospital, Aligarh, with fever, extreme weakness, pallor, and proptosis of both eyes. The symptoms were of one month's duration. There was no loss of vision or any history of epistaxis, haemoptysis, or bleeding from the gums.

Examination.—The boy was poorly nourished with marked pallor of the skin and mucous membranes. The temperature was 102°F. The submandibular and preauricular lymph glands were enlarged, but the lymph glands in the region of neck, axilla, and groin were not enlarged.

Small subconjunctival haemorrhages were present. Accommodation was normal. Proptosis and lagophthalmos were present but more marked in the right eye (Fig. 2).

FIG. 2.—Case 2, showing marked proptosis of right eye.

The fundus examination revealed tortuous vessels with multiple haemorrhages near the disc which had a pale centre. The blood could be seen moving in one or two blood vessels, spurting past a constriction.

The spleen and liver were palpable one finger's breadth below the costal margin. The cardiovascular, respiratory, and central nervous systems revealed nothing important.

Peripheral Blood

Haemoglobin	3.0 g. per cent.
Erythrocytes	0.8 million per c.mm.
Packed cell volume	9 per cent.
Mean corpuscular volume	112 c.μ.
Mean corpuscular haemoglobin concentration	33 per cent.
Total leucocytes	88,000 per c.mm.

Differential Leucocytes

Myeloblasts	42% per cent.
Promyelocytes	6% per cent.
Neutrophils	12% per cent.
N. myelocytes	10% per cent.
Lymphocytes	30% per cent.

The peroxidase test was positive in the blast cells. Thrombocytes were markedly diminished. Few nucleated red blood cells were also seen. Bone marrow biopsy could not be done.

A skiagram of the orbit was normal.

Urine.—Bence Jones protein absent.

Kahn Test.—Negative.

Progress.—The patient was given cortisone and a blood transfusion, but the parents removed him from hospital after a few days against medical advice, and he died at his home after a week.

Discussion

The blood picture in chloroma may be myeloid or lymphoid in type (Brannan, 1926; Washburn, 1930), but other workers are of the opinion that all cases of chloroma are myeloblastic, and that the lymphoblasts occasionally described would on further examination be found to be myeloblasts. Other workers (Roehm, Riker, and Olsen, 1937) consider the blast cell to be a primitive cell, while Pasca (1949) and Durie, Lemberg, and Sear (1950) consider it to be a haemocytoblast which may differentiate as a lymphocyte or a myelocyte. Gump, Hester, and Lohr (1936) described a case of monocytic chloroma in a male aged 55 years.

In this paper investigations could be completed in Case 1, but no sternal or tibial aspiration was possible in Case 2.

Case 1.—Both the peripheral and sternal puncture smears showed that the nucleus of the blast cells was either oval or reniform, containing two to three nucleoli (Fig. 3).

FIG. 3.—Case 1, sternal puncture smear. $\times 500$.

The cytoplasm in many cells contained auer bodies and azure granules (Figs 4 and 5, opposite). The blast cells were peroxidase-positive. As is evident from the myelogram, the more mature cells were mostly of the granular series (Fig. 6, overleaf). The haematological picture was consistent with that of acute paramyeloblastic leukaemia (Naegeli type).

FIG. 4.—Case 1, sternal puncture smear, showing blast cells of a monocytoïd character. $\times 1125$.

FIG. 5.—Case 1, sternal puncture smear, showing Auer bodies in the cytoplasm. $\times 1125$.

Case 2.—The peripheral blood showed typical myeloblasts with two to four nuclei. They were strongly peroxidase positive, and a diagnosis of acute myeloblastic chloroma was made.

The general condition of Case 1 was very poor, though his blood picture was not so acute as that of Case 2, who looked clinically in a better general condition.

FIG. 6.—Bone biopsy from supra-orbital swelling showing immature white-cell aggregation under periosteum. Haematoxylin and eosin. $\times 300$.

Summary

The literature and pathology pertaining to certain aspects of chloroma are briefly reviewed.

Two cases of chloroma of the orbit are reported, one presenting an acute para-myeloblastic (Naegeli) blood picture and the other an acute myeloblastic blood picture.

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