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CASE NOTES

MULTIPLE MYELOMATOSIS WITH PROPTOSIS*

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MULTIPLE myelomata are uncommon endosteal tumours developing from the myeloblastic elements of the bone marrow. The ribs, sternum, clavicle, vertebral bodies, and pelvis are favourite sites; the skull is also frequently involved and the long bones may be affected, but never alone (Perkins, 1936). The disease has an insidious onset and is often only recognized late when multiple nodules appear or a spontaneous fracture of one of the long bones takes place. The orbital wall is sometimes an early site, the patient seeks medical advice for the ensuing proptosis.

Case Report

A female aged 50 was referred to me by Prof. Attiah in February, 1953, with right-sided proptosis of 2 months' duration. The condition developed very quickly, starting with pain above the right eye, followed in few days by failing vision, oedema, chemosis, and proptosis. The pain disappeared in 2 weeks but the other manifestations persisted, and the right vision was soon completely lost.

There was no fever, and no history of trauma, syphilis, or tuberculosis.

General Examination.—No swellings in the vault of the skull or other part of the body. No detectable abnormality in the heart or lungs. Nose and ears within normal limits. Pyorrhoea alveolaris present.

Ophthalmic Examination.—The right eye showed severe chemosis and oedema of the conjunctiva and lids, particularly the lower lid which was everted (Fig. 1). The globe was proptosed forwards downwards and inwards, and ocular movements were limited upwards and outwards. The cornea was hazy, the pupil was normal in size but did not react to light, and the fundus could not be seen. The eye was completely blind. The outer part of the supra-orbital margin was the seat of a visible and palpable bony hard swelling which extended to the right temporal fossa. This swelling was not tender on pressure.

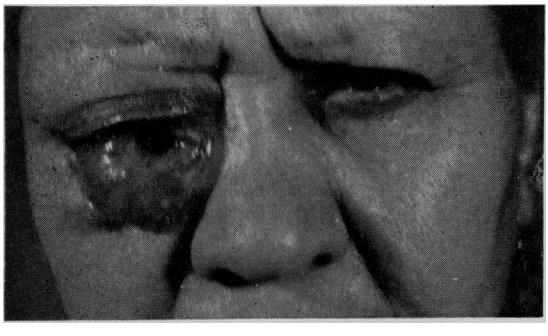

FIG. 1.—Right proptosis, secondary to multiple myelomatosis.

The left eye appeared to be normal; the vision was 6/36, and the fundus was normal.

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The exophthalmic measurements were 27 right, and 15 left.

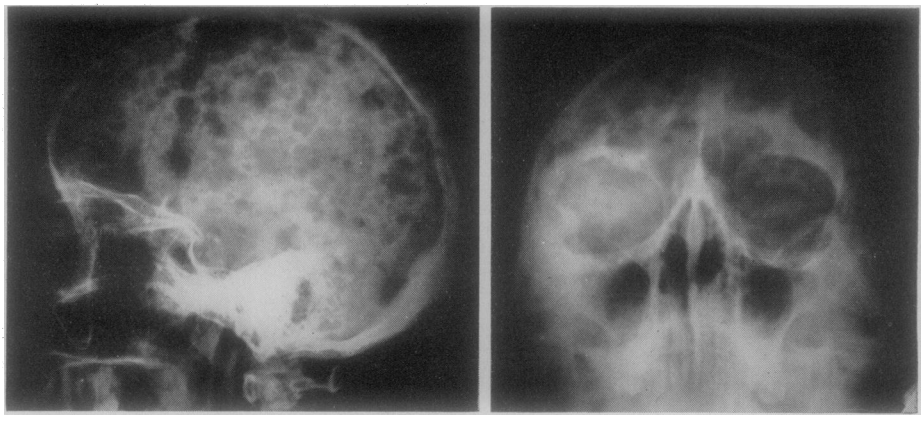
Laboratory Findings.—Wasserman reaction: negative. Red blood cells 4,920,000, haemoglobin 96 per cent., platelets 349,000 per c.mm., white blood cells 9,800 per c.mm., eosinophils 2 per cent., staff nucleated 5 per cent., segmented 60 per cent., lymphocytes 30 per cent., monocytes 3 per cent., myeloma cells, nil. Total serum protein 10·15 g., per cent., albumin 3·86 g., globulin 6·29 g., albumin/globulin ratio 0·59: 1, calcium 9·2 m. per cent., phosphorus 6·8 per cent., alkaline phosphatase 9·25 King and Armstrong units, formol gel reaction positive.

Urine Bence-Jones proteose positive.

Marrow (sternum) total count 127,800, basophils 0·0 per cent., eosinophils 0·4 per cent., juveniles 0·8 per cent., segmented 1·2 per cent., lymphoctyes 1·6 per cent., normoblasts 1·2 per cent., myeloma cells 94·8 per cent.

Marrow (iliac crest) total count 49,600, basophils 2·0 per cent., eosinophils 2·0 per cent., myeloblasts 0·0 per cent., promyelocytes 1·2 per cent., myelocytes 2·8 per cent., juveniles 10·0 per cent., staff nucleated 4·2 per cent., segmented 11·2 per cent., lymphocytes 11·6 per cent., normoblasts 13·2 per cent., myeloma cells 41·8 per cent.

Radiological Examination.—The cranium as a whole is riddled with radiolucent areas, having a fairly well-defined outline; some have become confluent with each other, suggesting multiple myelomatosis (Figs 2 and 3).



FIGS 2 and 3.—Radiolucent areas in skull, characteristic of multiple myelomatosis.

The sternum, ribs, and pelvic bones, and the rest of the skeleton show no abnormality.

The lung fields show no abnormality.

Exploration of the swelling related to the orbit revealed a bone cavity full of reddish gelatinous material. A piece was removed and histologically examined by Prof. M. Hashem who made a diagnosis of plasma cell myeloma (Fig. 4).

Comment

The typical radiological manifestations in the skull bones and the positive laboratory tests, particularly the Bence-Jones reaction in the urine, strongly suggested the diagnosis of multiple myelomatosis. This was confirmed by histological study of the mass in the orbital rim.

The patient was of the usual age (50 yrs) at which these cases occur but was not

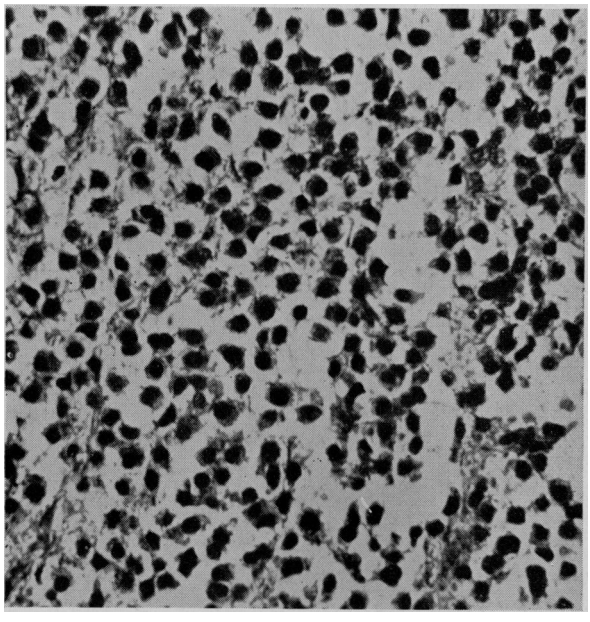


FIG. 4.—Photomicrograph of histological appearance of section showing abundance of plasma cells. ×400.

anaemic (haemoglobin 86 per cent.). The bone manifestations were limited to the skull and there were no pulmonary changes or gastro-intestinal trouble. This indicates that the case was seen at a comparatively early stage. Patients suffering from multiple myelomatosis are usually first seen with multiple bony tumours arising in different parts of the body and are much more anaemic than in this case.

The myeloma mass causing the proptosis developed in the upper and outer quadrant of the orbit, which is not the usual site. Duke-Elder (1952) states that such masses are most often seen in the upper and inner angle.

This disease is considered to be malignant, and according to Perkins (1936), all patients usually die within 2 years from cachexia or anaemia.

The right proptosis which caused this patient to seek advice was due to the myeloma mass developing in the wall of the orbit. This was painful to start with, but in 2 weeks the pain disappeared. The pain may have been due to an attack of haemorrhage in the developing mass.

I wish to thank Prof. Attiah, Professor of Ophthalmology, Cairo University, for referring the case, Prof. Hashem for the interest he has taken in the histological examination, Dr. Marey and Dr. Mahmoud El-Sayed for the radiological studies, and Dr. S. Dewi for the laboratory tests.

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