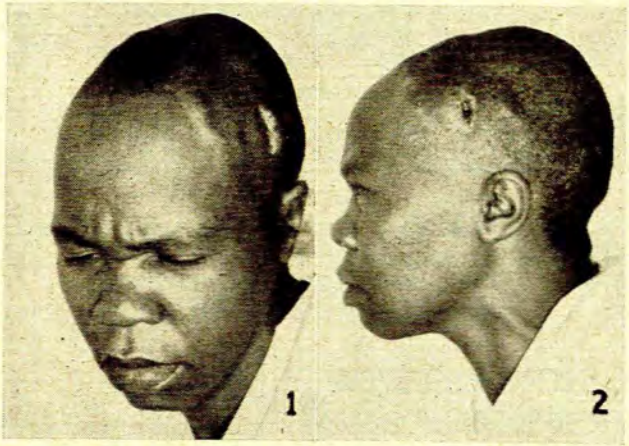
A CASE OF MULTIPLE MYELOMA PRESENTING CLINICALLY AS A SOLITARY TUMOUR OF THE SKULL

J. M. HOFFMAN, F.R.C.S. (EDIN.), *Surgeon* and H. VAN Z. DE KLERK, F.R.C.S. (EDIN.), D.OBSTET., R.C.O.G., *Surgical Registrar*
Livingstone Hospital, Port Elizabeth

This case of multiple myeloma (plasmacytoma) in a Bantu woman, presenting clinically as a solitary tumour of the skull, is interesting on clinical, pathological and radiological grounds.

Plasmacytoma occurs mainly in bone but occasionally in soft tissue, when it is confined to the submucosa of the air-passages and gastro-intestinal tract. Solitary plasmacytoma is rare, most 'solitary' cases proving to be multiple on follow-up.



The patient is a Bantu woman of 49 with a history of a gradually increasing swelling over the left parieto-occipital region for 6 months (Figs. 1 and 2). On examination a swelling is apparent over the left parieto-occipital area, 4 × 6 inches in size, irregularly lobulated and of the consistency of soft rubber mainly, but variable in parts. A plaque of bone is palpable in the centre, and defects in the skull margin at the edges.

X-ray Examination

A large bony defect is seen over the left parieto-occipital region (Figs. 3 and 4). There is fair definition of the edges, with scalloping. A plaque of the outer table has also lifted up. Generally small areas of sclerotic bone are surrounded by areas of diminished density.

X-rays of the vertebrae and pelvis show small osteolytic deposits.

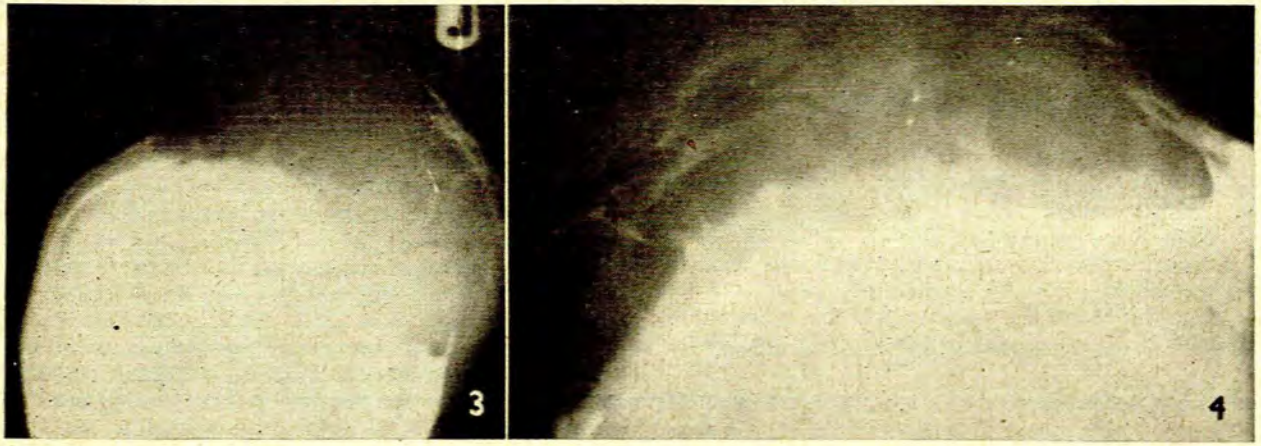
Cerebral arteriograms show no evidence of excessive vascularization or intracranial shift.

Other Examinations

Blood picture: Hb. 62%. White blood cells 17,400 per c.mm. (neutrophils 84%, monocytes 2%, lymphocytes 13%, eosinophils 1%). PCV 26%.
ESR 30 mm. Westergren.
Serum albumen 3·3 g.%, globulin 6·7 g.%, A/G 0·5 : 1.
W.R. negative.
Bence Jones protein negative (3 occasions).
Plasma electrophoresis—β globulin fraction increased.
Bone marrow—no abnormal cells detected.
Serum alkaline phosphatase 19 K.A. units. Serum calcium 8·8 mg.%. Serum phosphorus 1·9 mg.%.

Histology

On biopsy, the mass was found to be plasmacytoma, with clusters of plasma cells situated in a stroma of small round cells.



DISCUSSION

Disregarding the histology, a brief discussion on the differential diagnosis is not out of place. The differential diagnosis is that of large skull defects associated with large soft-tissue swellings, as follows:

Bone Tumours Proper

(*a*) Osteogenic sarcoma of the osteolytic type: Some new bone formation but mainly destruction. May also be radiating spicules and periosteal elevation. Not so much destruction as in this case.

(*b*) Solitary osteoblastoma—not described in the skull.

(*c*) Fibrosarcoma: Soft-tissue swelling with erosion of outer table.

(*d*) Reticulum-cell sarcoma—not in skull.

(*e*) Plasmacytoma: Nearly always multiple and osteolytic. Some bone sclerosis in between. We find no previous case of a single large skull deposit occurring so early in the disease. It is possible that deposits may coalesce if the patient survives long enough.

(*f*) Metastatic carcinoma: Irregular 'moth-eaten' osteolytic deposit related to a meningeal artery. Erode inner table more than outer.

Congenital—Cholesteatoma:

Arise in diploë. Erode inner table more than outer. Areas of lesser density with sclerotic margins leading to an appearance of scalloped margins. With very large lesions islands of bone (usually outer table) may remain intact and may be lifted out of position. This condition is very similar in its X-ray appearance, to the case reported here, especially in view of the scalloped edges and the elevation of plaques of outer table.

Endocrinopathies—Hyperparathyroidism:

Generalized osteoporosis and small cysts (areas of decalcification).

Deficiency—Osteomalacia and osteoporosis:

Generalized.

Rare Diseases

(*a*) Disorders of marrow constituents

(i) Lipoid granulomas and Hand-Sculler-Christian disease: X-ray appearance, large geographical skull defects with smooth edges and no reaction at edges. May be associated with exophthalmos, diabetes insipidus, and involvement of other membrane.

(ii) Eosinophilic granuloma: quite small clear-cut areas.

(iii) Letterer-Siwe disease (aleukaemic reticulosis): Special areas of bone loss. Occurs in infants.

(iv) Multiple myeloma—see above.

(*b*) Mesenchymal defects

Paget's disease and osteoporosis circumscripta: Much smaller lesions and with fluffy areas of mixed spongy bone and dense bone.

(*c*) Disorders of endochondral ossification

Fibrous dysplasia—may be monostotic or polystotic and associated with pigmentation and sexual precocity. Radiologically—diffuse osteolytic areas with fluffy cottonwool appearance of the tables.

Meningioma

Hyperostosis of diploë. Stippling if very vascular. May be irregular decalcification. If sarcomatous change occurs an appearance of lytic areas with scalloped margins may be seen. Besides plasmacytoma, only in this condition and in cholesteatomas is scalloping described.

SUMMARY

A case of multiple myeloma presenting clinically as a large solitary skull tumour is described. The differential diagnosis is discussed. The radiological picture is bizarre in that there is a large irregular osteolytic area with some osteosclerotic areas; complete erosion of the inner table and the lifting up of plaques of outer table; and scalloped edges. It will be noted that in only two skull conditions, viz. cholesteatoma and meningioma, are scalloped edges met with any frequency.

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