Short report

Cerebral compression by myeloma

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SUMMARY A patient presented with right hemiparesis, dysphasia and a large, palpable, left fronto-parietal mass arising from the calvarial diploe. A diagnosis of multiple myeloma was made on radiological and biochemical grounds, together with bone marrow examination. Few cases of cerebral compression by a massive plasmacytoma have been reported, but in some surgical management produced clinical deterioration. The present case was treated with radiotherapy and chemotherapy, with satisfactory results.

Symptomatic intracranial extension of multiple myeloma is uncommon. When it occurs, the deposits are usually at the skull base.¹⁻⁴ Despite the frequency of calvarial deposits, symptomatic cerebral compression is very rare. For example, Currie and Henson, in a review of 125 patients with multiple myeloma,¹ found 40 cases with spinal cord compression, 15 cases of "indirect" neurological complications, four patients with isolated cranial nerve palsies, and none with symptoms of cerebral compression. Other series are similar,⁵⁻⁸ with only those of Silverstein and Doniger⁷ and Carson *et al*⁵ containing single examples of possible cerebral compression in 227 and 90 cases of myeloma respectively. We present a case of cerebral compression by a large mass of myeloma tissue arising from the calvarium, treated successfully by radiotherapy and subsequent chemotherapy.

Case history

A 70 year old right handed woman (LH 875636) noticed some discomfort and swelling of the medial end of the left clavicle in August 1979. In January 1980 she first noticed weakness of the right limbs, which progressed over eight weeks. She became aware of a painless swelling on the left side of her head, which gradually enlarged. In February 1980 she noticed some difficulty with reading and word-finding. She had no sensory symptoms, or headaches. On admission in March 1980, the medial end

of the left clavicle was enlarged and a little tender. There was a soft swelling, 10 cm in diameter, in the left fronto-parietal region; it could not be separated from the surrounding skull, but the scalp could be moved over it. The swelling was fluctuant and transmitted a faint impulse on coughing. The patient was alert, but made occasional errors in naming simple objects, reading aloud and performing simple calculations. The fundi showed mild papilloedema. Stimuli in the periphery of the right visual field were sometimes neglected. The other cranial nerves were normal. There was a mild right hemiparesis. Joint position and vibration sense were slightly impaired on the right.

A skull radiograph (fig A) showed a large lucent area in the left fronto-parietal region with many smaller defects elsewhere. A computed tomographic scan (fig B, C) showed a large, high density mass, arising from the diploic space of the left parietal bone, displacing the brain medially and producing a shift of the ventricular system to the right. There was a homogeneous increase in radio-density of this mass after the injection of contrast medium. A chest radiograph showed erosion of the medial ends of both clavicles (especially the left) and erosion of the lateral end of the left clavicle. Haemoglobin was 10.5 gm/dl, and the erythrocyte sedimentation rate was 50 mm/hour. Blood urea was 11 mmol/l and calcium was 2.66 mmol/l. Serum protein electrophoresis pattern was normal, but serum IgM levels were slightly depressed (42 mg/dl). A specimen of urine was found to contain kappa-chain Bence-Jones protein. Bone marrow aspirate was of normal cellularity and contained a patchy increase in plasma cells, which were 35% of nucleated

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Figure Radiological findings (A) Skull radiograph on admission showing a large defect in the parietal bone and several smaller punched out lesions of bone. (B) Computed tomographic (CT) scan showing a large left parietal mass. There is a thin rim of bone on the external surface of the mass. At another level a similar rim of bone, arising from the inner table, lies over part of the inner aspect of the mass. The medial border of the mass is clearly demarcated. It was concluded that the mass was arising from within the bone and was limited medially by the dura mater. (C) Coronal CT scan after injection of contrast material. The left lateral ventricle and choroid are displaced medially under the falx. (D) CT scan six months after treatment shows the bony defect and no residual tumour.

cells in some areas. Some binucleate cells were seen, and Russell bodies were present in the cytoplasm of many cells. The trephine specimen contained clusters of plasma cells, some abnormal in form. These features, in conjunction with the characteristic radiographic findings and the presence of Bence-

Jones protein made the diagnosis of multiple myeloma inescapable. It was thought neither necessary nor advisable to biopsy the cranial mass.

Radiotherapy was started directly, a total dose of 3000 rads whole brain irradiation given in 10 fractions over 13 days. At the end of this course, melphalan 32

mg and prednisolone 160 mg were given in divided doses over four days. The patient's response to this treatment was dramatic. After completion of the course of chemotherapy, there was no detectable neurological deficit and within a month the mass had disappeared. Three similar courses of chemotherapy were given at six week intervals. Clinical review and CT scan (fig D) at six months after presentation revealed no evidence of recurrence of the cranial mass and no neurological deficit.

Discussion

Myeloma tissue compressing the brain may arise from the calvarium,^{2 9-15} from the dura,^{2 16-19} or within the cerebral substance, according to some authors.^{20 21} Thirteen previously reported cases of cerebral compression by myeloma are summarised in the table; two cases apparently arising intracerebrally and one case of cerebellar compression by a mass arising from the posterior fossa convexity are

Table Reported cases of cerebral compression by myeloma

Author	Solit or mult	Site of origin	Region	Systemic ICP dis	Ep	Focal	Pal-pable mass	Operation		XRT	Chemo	Response to treatment
								Exc biopsy	Deterioration			
Kalischer 1928 ⁹ (Case 1)	Mult	Bone	Parietal	—	—	Hemiparesis (tender)	0	0	—	0	0	Died
Balan & Ballif 1932 ¹⁰	Mult	Bone	Fronto-par	?	—	Hemiparesis	—	0	—	0	0	Died
James & Turner 1952 ¹¹	Mult	Bone	Occipital	—	—	Hemiplegia	—	0	—	—	Stilbamide	Died
Stark & Henson (present case)	Mult	Bone	Fronto-par	—	—	Hemiparesis	—	0	—	—	Melphalan + pred	Excellent at six months
Villata 1928 ¹²	Solit	Bone	Occipital	?	0	Hemiparesis	—	Ex	No	0	0	Recurred in two years "Cured"
Kaufman 1945 ¹³	Solit	Bone	Occipital	0	0	0	—	Ex	No	—	0	
Clarke 1954 ² (Case 2)	Solit	Bone	Fronto-par (L)	?	—	Hemiparesis (L)	—	B	No	—	0	Recurred two years later "Satisfactory"
Chang & Jing 1970 ¹⁴	Solit	Bone	Parietal	0	—	0	0	Ex	No	—	0	
Kutcher 1974 ¹⁵ (Case 1)	Solit	Bone	Parietal	0	0	0	—	Ex	No	—	0	"Satisfactory"
(Case 2)	Solit	Bone	Parietal	0	0	Hemiparesis	0	Ex	Yes	—	0	Permanent mild hemiparesis
Medoc <i>et al</i> 1961 ²²	Mult	Dura	Frontal	0	—	0	0	Ex	No	0	0	Operative difficulties. Full recovery
Clarke 1954 ² (Case 3)	Solit	Dura	Tentorium (sup aspect)	0	—	Hemiparesis	0	Ex	Yes	0	0	Died 36 hours post op
Weiner <i>et al</i> 1966 ¹⁷	Solit	Dura	Temporal	0	—	Hemiparesis	0	Ex	Yes	—	0	Hemiparesis worse
Moossy & Wilson 1967 ¹⁸	Solit	Dura	Frontal	0	—	Frontal syndrome	0	Ex	Trans-ient	—	0	Post op haematoma drained— asymptomatic
Someren 1971 ¹⁹	Solit	Dura	Post-fossa convexity	0	—	Ataxia	0	Ex	Yes	—	0	Permanent ataxia
Kramer 1963 ²⁰	Mult	Intra CBL	Temporal	—	—	Hemiparesis	0	0	—	0	Cortisone	Died
Labauge 1963 ²¹	Solit	Intra CBL	Frontal	0	—	Hemiparesis	0	Ex	Yes	0	0	Died post op

Mult: multiple myeloma, Solit: Solitary plasmacytoma, Systemic disease: + = anaemia, paraproteinaemia, Bence Jones proteinuria or abnormal bone marrow, — = none of these, ? = insufficient data, ICP: evidence of raised intracranial pressure, Ep: epileptic seizures, Operation: Ex = excision attempted, B = biopsy only, O = no operation, XRT: radiotherapy, Chemo: chemotherapy, Pred: Prednisolone.

appended for comparison, especially with regard to surgical results, but are not further analysed below. Only four of these cases had disseminated myeloma, the other nine suffered from solitary plasmacytomas of the calvarium (six cases) or dura (three cases). Features of raised intracranial pressure were present in nine cases, focal neurological signs in nine, and seizures in only one. Metabolic disturbances produce seizures in patients with myeloma more often than cerebral compression.^{2 23} A mass was palpable in six of the nine tumours arising from the calvarium, and in one such case there were no neurological signs, despite demonstrable cerebral compression.¹³ Operative excision was attempted in 10 cases. Difficulties during the procedure were recorded in two patients; in one, haemorrhage and cerebral oedema required prompt transfusion and intravenous mannitol,²² while, in the other, the surgeon abandoned the procedure after obtaining a biopsy.² There was one post-operative death² and in three cases,^{15 17 18} there was neurological deterioration, permanent in two.^{15 17}

It should be noted that basal infiltration by myeloma may mimic direct cerebral compression. For example, Mahoudeau *et al*²⁴ reported a case presenting with raised intracranial pressure and subsequently developing a right hemiparesis (without aphasia) and seizures. Ventriculography showed no displacement and exploration revealed basal arachnoidal infiltration with tumour.

Diagnosis of cerebral compression by myeloma may be aided by angiography^{14 15} or computed tomography. Only in cases of solitary plasmacytoma or tumours not arising from bone will biopsy be necessary. Our opinion, based on a review of the relevant literature, is that surgery should be avoided, or limited to biopsy, and that treatment should be with radiotherapy and, in the case of generalised myeloma, chemotherapy. This course of action produced satisfactory results in the case presented here.

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