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# Meningeal Involvement of Myeloma

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**Summary**: A rare case of IgG myeloma with meningeal involvement is described. By reviewing 13 reported cases of meningeal infiltration by systemic myeloma, IgD myeloma, leukemic transformation and pleural effusion were found to be the risk factors for the meningeal involvement in these cases.

**Key words**: myeloma—meningeal involvement—dementia—hyperviscosity—plasma cells in cerebrospinal fluid

## Introduction

Involving the nervous system in myeloma usually takes the following four forms; compression of the spinal cord and cauda equina, cranial nerve and intracranial involvement, compression of nerve roots, and peripheral neuropathy (Clark, 1954; Botterell and Fitzgerald, 1959; Hamre and Bruland, 1960; Kramer, 1963; Campbell and Halford, 1971; Davies-Jones and Esiri, 1971). Meningeal infiltration is rare in intracranial involvement.

This communication reports a rare case of myeloma with meningeal involvement in a patient who had an interesting mental symptom.

## Case Report

A 70-year-old man was admitted to this hospital on January 7, 1985, because of general fatigue. He was well until one week before coughs developed. There was no history of mental symptom, lumbago and headache.

On admission, his vital signs were temperature, 38.2℃; pulse rate 96; respiration 16; and blood pressure, 100/60 mmHg. He was alert and cooperative. Neither rash nor lymphadenopathy were found. A few persistent fine rales were audible at the base of both lungs. The chest x-ray revealed diffuse areas of reticular shadow at the base of the lungs without osteolytic lesion of the ribs.

The red blood cell count was $436\times10^4$ per c. mm without rouleaux formation. The white cell count was 15,100/mm$^3$ with a differential count (expressed in %), neutrocytes, 84; lymphocytes, 13; monocytes, 2; eosinocytes, 0.5; and basocytes, 0.5. The erythrocyte sedimentation rate was 40 mm per hour. Other laboratory findings were including urea nitrogen, 83.3 mg/dl; creatinine 1.9 mg; glucose 114 mg; calcium 9.5 mg; and sodium 149 mEq /l. He was treated with antibiotics from the day of admission.

On January 9, the patient arose from his bed and wandered around without any apparent reason, and became disoriented, inattentive and mute. For the following

few days, he was restless, constantly in motion in his bed and did not obey commands or speak. Psychiatric diagnosis on him was severe dementia.

On January 14, he was noted to have neck stiffness and Kernig's sign. A computed tomographic (CT) scan of the brain showed decreased attenuation of deep white matter of frontal lobes with narrowing anterior part of lateral ventricles (Fig. 1). Neither abnormal enhancement nor mass was identified.

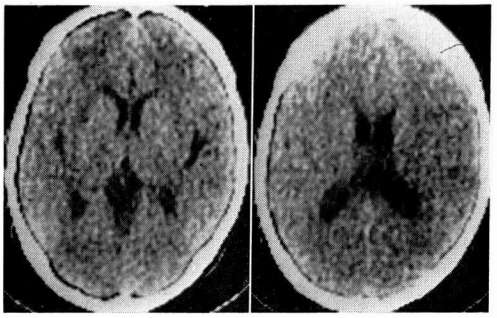

*Fig. 1.* Plain CT scan of the brain demonstrating decreased attenuation of deep white matter of frontal lobes and narrowing anterior part of the lateral ventricles.

Cerebrospinal fluid, which was obtained through a lumbar puncture, was clear and its pressure was 155 mm$H_2O$. It contained 207 white cells/3c. mm; glucose, 54; and protein, 33 mg/dl. Its culture was sterile. The differential count of the cerebrospinal fluid was (in %) monocytes, 48.0; plasma cells, 25.5; neutrocytes, 15.5; and lymphocytes; 11.0.

The plasma cells were in various size. Some irregular multiple nuclei and flame cells were also observed in them (Fig. 2, 3).

Total serum protein was 5.8 g/dl with 33.7 % albumin and 34.9 % $\gamma$-globulin. M protein was found in the gamma region of electrophoretical separation and an IgG monoclonal gammopathy was also seen by immunoelectrophoresis.

On January 23, a lumbar puncture was performed for an immunofluorescent study of plasma cells in the cerebrospinal fluid.

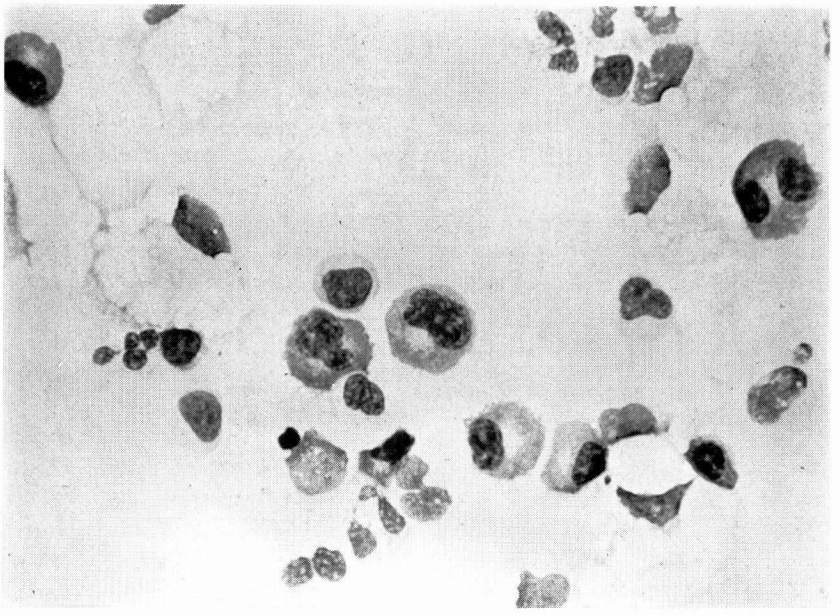

*Fig. 2.* CSF cytologic examination showing large and small plasma cells with either irregular or multiple nuclei. May-Giemsa stain. ×200

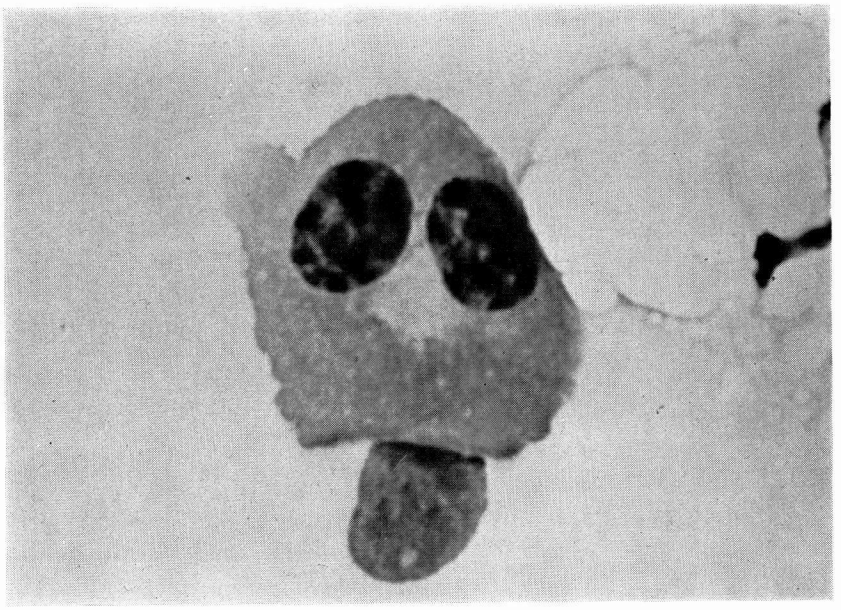

*Fig. 3.* CSF cytology demonstrating so called "flame cell". May-Giemsa stain. ×500

The cytoplasm fluoresced with antisera against lambda chains but not against kappa chains.

The patient died from respiratory failure on this day. Both the skeletal survey and bone marrow puncture were not performed. Permission for autopsy examination was not obtained.

## Discussion

Although neither skeletal survey nor bone marrow study were conducted, IgG lambda myeloma was established by both the hematological and immunological examinations.

Plasma cells are never present in normal cerebrospinal fluid. Their presence in the fluid have been described in cases of viral meningitis, multiple sclerosis and chronic infections such as sarcoidosis, tuberculosis, syphilis and subacute sclerosing panencephalitis (Schaltenbrand, 1954; Péter, 1967; Kölmel, 1977). In those conditions, population of plasma cells in cerebrospinal fluid was a few percent or less of the total.

However, many plasma cells which resembled abnormal cells of multiple myeloma were found from this patient. They were identified as monoclonal myeloma cells from the immunofluorescent study.

Involving leptomeninges with systemic myeloma is an unusual phenomenon. The first such case was reported in 1968 by Ben-Bassat et al. Since then, there have been described at least 12 cases (Maldonado et al. 1970; Spar and Argyrakis, 1972; Afifi, 1974; Woodruff et al. 1978; Hughes and Votaw, 1979; Slager et al. 1979; Davies-Jones et al. 1980; Schulman et al. 1980; Spier et al. 1980; Johnston et al. 1982; Yamagata et al. 1983). By reviewing those and the present case, neurological manifestations were consisted as follows; impairment of consciousness in seven patients (Maldonado et al. 1970; Spar and Argyrakis, 1972; Hughes and Votaw, 1979; Slager et al. 1979; Davies-Jones et al.

1980; Spier et al. 1980; Yamagata et al. 1983), meningeal irritation in five (Maldonado et al. 1970; Slager et al. 1979; Spier et al. 1980; Yamagata et al. 1983; Present case) and cranial nerves involvement in three (Maldonado et al. 1970; Woodruff et al. 1978; Schulman et al. 1980). Two patients were neurologically asymptomatic meningeal involvement (Ben-Bassat et al. 1968; Johnston et al. 1982). There were three immunologic types as follows, three cases in each; IgG (Maldonado et al. 1970; Davies-Jones et al. 1980; Present case), IgA (Spar and Argyrakis, 1972; Hughes and Votaw, 1979; Spier et al. 1980) and IgD (Ben-Bassat et al. 1968; Schulman et al. 1980; Johnston et al. 1982). Two patients had Bence Jones protein (Woodruff et al. 1978; Yamagataet al. 1983). Although the IgD myeloma were thought to be a rare type of multiple myeloma, Schulman et al. (1980) suggested that meningeal involvement in this type might be more common than previously thought. Since leukemic transformation and plasma cell leukemia were seen in six patients (Ben-Bassat et al. 1968; Maldonado et al. 1970; Woodruff et al. 1978; Davies-Jones et al. 1980; Spier et al. 1980; Johnston et al. 1982), hematogeneous spread was the most probable mechanism for meningeal involvement. On the other hand, Maldonado et al. (1970) described a case of meningeal myeloma suggesting the possibility of contiguous involvement from the dural myelomatous lesion.

Four patients had pleural myelomatous effusion which was uncommon in myeloma (Maldonado et al. 1970; Hughes and Votaw, 1979; Schulman et al. 1980; Johnston et al. 1982). Because of this high incidence, Johnston et al. (1982) proposed giving a diagnostic lumbar puncture to a patient with myelomatous pleural effusion.

Among seven cases which were performed postmortem examination (Ben-Bassat et al. 1968; Maldonado et al. 1970; Hughes and Votaw, 1979; Slager et al. 1979; Davies-Jones et al. 1980; Spier et al. 1980; Johnston et al. 1982), two showed perivascular cuffs of cerebral vessels (Ben-Bassat et al. 1968; Davies-Jones et al. 1980). Under CT study, the abnormality observed in this case could be interpreted that the cerebral perivascular cuffs and parenchymatous invasion of myeloma cells were existed.

It seems probable that the patient's mental state was attributed by hyperviscosity-induced dementia like the case reported by Mueller et al. (1983).

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