# Acute diplopia and a solitary lung mass: A unique presentation of light-chain myeloma

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The patient described, a 51-yearold woman, had diplopia and cephalgia of two weeks' duration. On admission, the radiologic evaluation revealed a mass in the sphenoid sinus, multiple lesions in the calvarium and a solitary lung mass. Biopsy of the lung mass revealed an atypical plasmacytic infiltration. Laboratory findings confirmed the diagnosis of light-chain myeloma presenting with a pulmonary plasmacytoma and cranial nerve involvement.

(Key words: Diplopia, cephalgia, light-chain myeloma, lung mass, plasmacytoma)

Common thoracic manifestations of multiple myeloma include multiple lytic bony lesions, solitary plasmacytomas, and direct extension of a rib lesion. The latter presentation, a smooth homogeneous mass protruding from an osteolytic rib lesion, suggests myeloma as the etiology<sup>1</sup>; however, the differential diagnosis also includes osteomyelitis, rib metastases, primary bronchogenic carcinoma, and acute fungal infections. The majority of patients with myeloma are symptomatic at presentation<sup>2</sup> but rarely have isolated neurologic involvement. The case we describe herein is a unique presentation of myeloma involving a solitary pulmonary plasmacytoma found in conjunction with isolated sixth nerve palsy.

#### Report of case

During May 1993, a 51-year-old woman was seen in the Neurology Clinic with recent onset of cephalgia and diplopia. The cephalgia was located behind the right eye and had not been present previously. The patient denied any

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recent trauma, environmental exposures, or progressive neurologic disease. Her past medical history included paraplegia and a neurogenic bladder secondary to thoracic spinal cord trauma, renal lithiasis, and mild arthritis. She had no history of tobacco use, ethanolism, diabetes, or hypertension. She had been married for 34 years, had five children in good health, and did not work outside the home. Her medications included alprazolam, propoxiphene, and trimethoprim-sulfamethoxazole.

Her vital signs were normal. Neurologic evaluation revealed normal pupil function with visual acuities of 20/25 and 20/40 on the left and right, respectively. She had esotropia of the right eye, and the right eye would not abduct beyond midline. The remaining extraocular movements and cranial nerves were intact. She had paraparesis beginning at a mid-thoracic spinal level (T-4 through T-8) of long standing resulting from old trauma. No other neurologic abnormalities were noted. Her lungs were clear to auscultation; heart rate and rhythm were regular; and the findings of the abdominal examination were unremarkable.

A cerebral angiogram, done on admission, revealed no angiographic vascular abnormalities. A magnetic resonance imaging (MRI) study of the head revealed an increased signal in multiple bilateral lesions of the calvarium and of a mass partially occupying the clivus (*Figure 1*). A chest roentgenogram demonstrated a  $3 \times 5$ -cm mass overlying a solitary, lytic eighth rib lesion on the right (*Figure 2*).

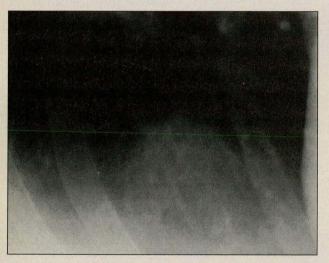
An initial dipstick test of the urine was 1+ for protein, whereas a simultaneous sulfosalicylic acid test was 4+ for protein—a discrepancy suggesting Bence Jones proteinuria.<sup>3</sup> The subsequent 24-hour urine collection contained 8198 mg of protein with 89.6%  $\gamma$ , 5.4% albumin, 2.7%  $\alpha_1$ , 1.1%  $\alpha_2$ , and 1.2%  $\beta$  fractions.

Laboratory studies revealed the following values: sodium, 139 mmol/L; potassium, 4.1 mmol/L; chloride, 100 mmol/L; bicarbonate, 27 mmol/L; blood urea nitrogen, 21 mg/dL; creatinine, 1.4 mg/dL; calcium, 10.2 mg/dL; phosphorus, 5.2 mg/dL; aspartate transaminase; 36 U/L; alanine transaminase, 39 U/L; lactic dehydrogenase, 523 U/L; creatinine phosphokinase, 163 U/L; alkaline phosphatase, 109 U/L; and  $\gamma$ -glutamyl transpeptidase, 123 U/L. Serum immunoelectrophoresis revealed no monoclonal spikes, low levels of IgA and IgM (22.1 mg/L and 15.2 mg/L, respectively), and a normal IgG level (812 mg/L).

Bone marrow and percutaneous right rib biopsies



**Figure 1.** Magnetic resonance imaging study of head shows increased signal in multiple bilateral lesions of the calvarium and of a mass partially occupying the clivus.



**Figure 2.** Chest roentgenogram showing  $3 \times 5$ -cm mass overlying solitary, lytic lesion on eighth right rib.

revealed atypical plasmacytic proliferation and infiltration. The clinical suspicion of high tumor burden was supported by bone scan findings of multiple osteolytic lesions in the ribs, thoracic spine, clavicles, and the calvarium. A diagnosis of stage IIIA light-chain multiple myeloma was made using the staging system of Durie and Salmon.<sup>4</sup>

#### Discussion

Multiple myeloma (or Kahler's disease) accounts for 10% of all hematologic malignancies and has a slight male predominance. Intrathoracic sites of origin are notably rare, with approximately 4% of all extramedullary plasma cell tumors involving the lung and bronchi. Thoracic manifestations of multiple myeloma may be primary (that is, solitary plasmacytoma of the respiratory tract or lung parenchyma) or secondary (that is, direct extension from a rib), with the latter being most common. Unusual thoracic presentations include medi-

astinal lymph node enlargement and pleural involvement. Specific neurologic manifestations include: spinal cord/nerve root compression ( $\sim 10\%$ ), distal sensorimotor polyneuropathy (< 1%), intracerebral invasion of the central nervous system (rare), and myelomatous meningitis, which occurs late in the course of aggressive disease.<sup>5</sup> In turn, light-chain disease represents about 20% of the annual incidence of myeloma (3 of 100,000 persons). These cases typically have a higher tumor burden and require aggressive chemotherapy.<sup>4</sup>

Biopsies of plasmacytoma typically show a fleshy, red-brown tissue composed primarily of plasma cells in abnormal aggregations. Light microscopy often reveals acidophilic, intracellular protein aggregates known as *Russell bodies*. Radiographic manifestations include punched-out lesions of the bone, vertebral compression fractures, and soft-tissue masses.

### Comment

Because myeloma is relatively common and has a multiplicity of presentations, unique presentations, such as the case presented here, will occur with regularity. In order to rapidly diagnose these unusual cases, the astute primary care physician should be aware of the diverse presentations and have a high index of suspicion for this disease.

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