Primary pulmonary plasmacytoma with immunoglobulin G/ λ light chain monoclonal gammopathy

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Pprimary pulmonary plasmacytoma (PPP) is a very uncommon form of extramedullary plasmacytoma. Serum M-proteins presented in a minority of patients with PPP.¹⁻³

In addition, most serum M-proteins were usually immunoglobulin (Ig) G/ κ chains.^{1,2} We report a case of PPP with IgG/ λ monoclonal gammopathy.

Clinical Summary

A 72-year-old man, a heavy smoker for more than 50 years, had a productive cough with whitish sputum for many years. The chest x-ray film showed a nodule in the right lung. The chest computed tomographic (CT) scan revealed one spiculated nodule, 1.5 cm in size, at the right lower lobe. Head and abdominal CT scans and the whole-body bone scan all showed no evidence of distant metastasis. Bronchoscopic examination disclosed no endobronchial lesion and the washing cytologic results were negative for acid-fast bacilli and malignancy. A CT-guided biopsy revealed that all the tissue fragments were predominantly replaced by plasma cells (Figure 1, A), which were strongly immunoreactive to MUM-1 (Figure 1, B), CD138, and VS38C, but negative for CD3, CD20, cytokeratin, and thyroid transcription factor-1 protein, indicating a plasmacytoma. A bone marrow biopsy disclosed no evidence of myeloma. The bone survey revealed a suspicious focal radiolucency with sclerotic margin at the right inferior pubic ramus. The pelvic magnetic resonance image (MRI) showed three focal bone lesions in the left iliac bone without periosteal reaction, suggesting benign lesions. Serum immunoelectrophoresis showed a thin band of IgG/ λ light chain monoclonal gammopathy superimposing on a polyclonal background (Figure 2). Urine Bence Jones protein was negative. A wedge resection of the right lower lobe via video-assisted thoracoscopic surgery was performed. Grossly, a gray-white round nodule measuring $1.6 \times 1.5 \times 1.5$ cm was noted. The pathologic report indicated a plasmacytoma that was similar to the previous biopsy specimen. The section margin was free. Postoperatively, a CT-guided needle aspiration for the left iliac bone lesions was performed but failed because the cortical bone was too hard to be passed through by biopsy needles. Six months later, the

Figure 1. A, Histologic finding of the tumor displaying sheets of plasma cells with extracellular hyaline globules. B, The tumor cells demonstrated strong nuclear staining of MUM-1, which is specific to plasmacytomas.

follow-up pelvic MRI showed no interval change. Serum and urine immunoelectrophoresis revealed a very faint band of monoclonal λ chain superimposing on a background of polyclonal gammopathy. Ten months after the diagnosis, the patient is well and without evidence of disease recurrence.

Discussion

Extramedullary plasmacytomas are plasma cell tumors that often originate in the nasal cavity or the upper respiratory tract. PPPs are rare. So far, fewer than 30 cases have been reported and verified by immunohistochemistry in the English literature.¹⁻⁴ The latest review article² of 19 cases of PPP with immunohistochemical support for diagnosis showed most patients with PPP are of middle age or older (range, 14-79 years) with a mean and median ages of 57 and 55 years. In the 19 patients, the male/female ratio was 1.4:1. The overall 2- and 5-year survivals were 66% and 40%, respectively.

The diagnosis of PPP is difficult. The findings on CT or MRI are nonspecific. The diagnosis is made only by histologic examination of the specimen, including immunohistochemical study, but

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Figure 2. Preoperative serum immunofixation electrophoresis showing a thin band of IgG/λ monoclonal gammopathy superimposing on a polyclonal background. *ELP*, Electrophoresis; *G*, IgG; *A*, IgA; *M*, IgM; *K*, κ (kappa); *L*, λ (lambda).

material obtained by fine needle aspiration or bronchoscopic biopsy is usually insufficient to make a definite diagnosis. Adequate tissue samples must be obtained by tumor resection via either video-assisted thoracoscopic surgery or thoracotomy. After the histologic diagnosis of pulmonary plasmacytoma has been made, a systemic staging with skeletal survey, bone marrow aspiration, and immunoelectrophoresis of serum and urine are required to exclude multiple myeloma.

We report a case of PPP with a rare presentation of IgG/λ monoclonal gammopathy. To our knowledge, only 2 other cases with IgG/λ chains secreted by PPP have been described.^{3,5} One patient had a similar presentation of PPP with IgG/λ monoclonal gammopathy. The other one presented with developing urine λ chain and serum IgG 6 months after the operation when skull lesions occurred, and multiple myeloma developed later. The M-protein level is directly related to tumor burden and clinical course. Three cases in which the M-protein level returned to normal after tumor resection have been reported.¹⁻³ In a review literature of PPP,⁴ multiple myeloma ultimately developed in 9 (40%) of 22 patients. Therefore, serial quantitation of M-protein after treatment of PPP is required. Persistence or increase in M-protein after treatment of extramedullary plasmacytomas suggests systemic disease.

In conclusion, this case is unique in the presentation of PPP with IgG/λ monoclonal gammopathy. The possibility of development of multiple myeloma in this patient should be kept in mind due to the persistence of M-protein after surgery.

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