# Extraosseous (Extramedullary) Plasmacytoma of the Adrenal Gland

*Craig G. Rogers, MD; Peter A. Pinto, MD; Edward G. Weir, MD*

**• Plasmacytomas are clonal proliferations of atypical plasma cells that manifest a localized osseous or extraosseous growth pattern. Although many represent solitary lesions of bone, fewer arise in extraosseous (extramedullary) tissues. We report a case of a primary extraosseous plasmacytoma of the adrenal gland. Magnetic resonance imaging (MRI) with contrast revealed a 3.5-cm, right adrenal mass with heterogeneous enhancement. Although the mass was small and nonfunctioning, concern for malignancy based on MRI findings prompted laparoscopic resection. Histologic and immunohistochemical findings were consistent with a plasmacytoma. This is the third reported case, to our knowledge, of a primary plasmacytoma of the adrenal gland. The present case is unique in that a contrast MRI was performed, which showed heterogeneous enhancement of the mass, providing further evidence beyond heterogeneous hyperintensity on T2-weighted images for a possible malignant process. Another unique feature is that a biopsy specimen of the lesion was taken, although it was nondiagnostic.**

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Plasmacytomas are clonal proliferations of atypical plasma cells that manifest a localized osseous or extraosseous growth pattern. Although many represent solitary lesions of bone, fewer arise in extraosseous (extramedullary) tissues. We report a case of primary extraosseous plasmacytoma of the adrenal gland and briefly review the literature.

## REPORT OF A CASE

A 75-year-old woman presented with a right adrenal mass, found incidentally on computed tomography for evaluation of microscopic hematuria. She reported intermittent right abdominal pain and fatigue. She had a remote history of breast cancer and had undergone a left radical mastectomy. Her family history was notable for carcinoma of the pancreas, lung, breast, and prostate. Physical examination results were unremarkable, and basic laboratory values were normal, including hematocrit, serum calcium levels, and renal function indices. Urinalysis was negative for glucose and protein. Urinary excretion of 17-ketosteroids, free cortisol, and catecholamines was within the normal range. Contrast magnetic resonance imaging (MRI) revealed a 3.5-cm, right adrenal mass with heterogeneous enhancement (Figure, A) and heterogenous hyperintensity on T2-weighted images. A fine-needle biopsy was performed to exclude metastatic breast carcinoma, but the biopsy specimen was nondiagnostic. A clinical diagnosis of a nonfunctioning adrenal tumor was made. Although the mass was small and nonfunctioning, we could not rule out the possibility of malignant potential given the heterogeneous enhancement of the mass on contrast MRI. The patient underwent an uncomplicated laparoscopic adrenalectomy. Pathologic examination of the resected specimen revealed a solitary plasmacytoma that was confined to the adrenal cortex.

Gross examination of the adrenal gland revealed a 3.2-cm, well-circumscribed, whitish tan cortical mass with no evidence of hemorrhage, necrosis, or cyst formation. Histologically, the mass was composed of a dense and diffuse infiltrate of atypical plasma cells and very few admixed lymphocytes. The plasma cells were unusually large and some were multinucleated (Figure, B). The results of immunohistochemical staining were positive for CD138 (Figure, C) and demonstrated λ immunoglobulin light chain restriction (Figure, D). The results were negative for CD20, cytokeratin, and both estrogen and progesterone receptors. The scattered lymphocytes in the mass represented a mixture of CD3-positive T cells and CD20-positive B cells. A staging bone marrow biopsy specimen and aspiration from the iliac crest showed trilineage elements of normal hematopoiesis and no evidence of a plasma cell dyscrasia. Plasma cells comprised approximately 5% of the total marrow cellularity, and they demonstrated a polyclonal pattern of light chain expression.

The patient's postoperative course was uneventful. Serum protein electrophoresis showed a broad-based γ region and no evidence of an M protein, and serum immunofixation electrophoresis showed no band of restricted electrophoretic mobility. Similarly, urine protein electrophoresis and immunofixation electrophoresis showed no evidence of a monoclonal protein and no evidence of restricted electrophoretic mobility. Skeletal survey showed a compression fracture of the lower thoracic spine but no other lesions. The patient was counseled to receive adjuvant radiotherapy and will be monitored with serum and urine protein electrophoresis and immunofixation studies.

## COMMENT

Plasma cell neoplasms are characterized by monoclonal proliferations of plasma cells that are associated with a single monotypic immunoglobulin product. Plasma cell neoplasms most commonly arise in the bone marrow,

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From the Department of Urology, Brady Urological Institute (Drs Rogers and Pinto) and Department of Pathology (Dr Weir), Johns Hopkins Hospital, Baltimore, Md. Dr Pinto is currently with Urologic Oncology Section, Surgery Branch, National Cancer Center, Bethesda, Md.

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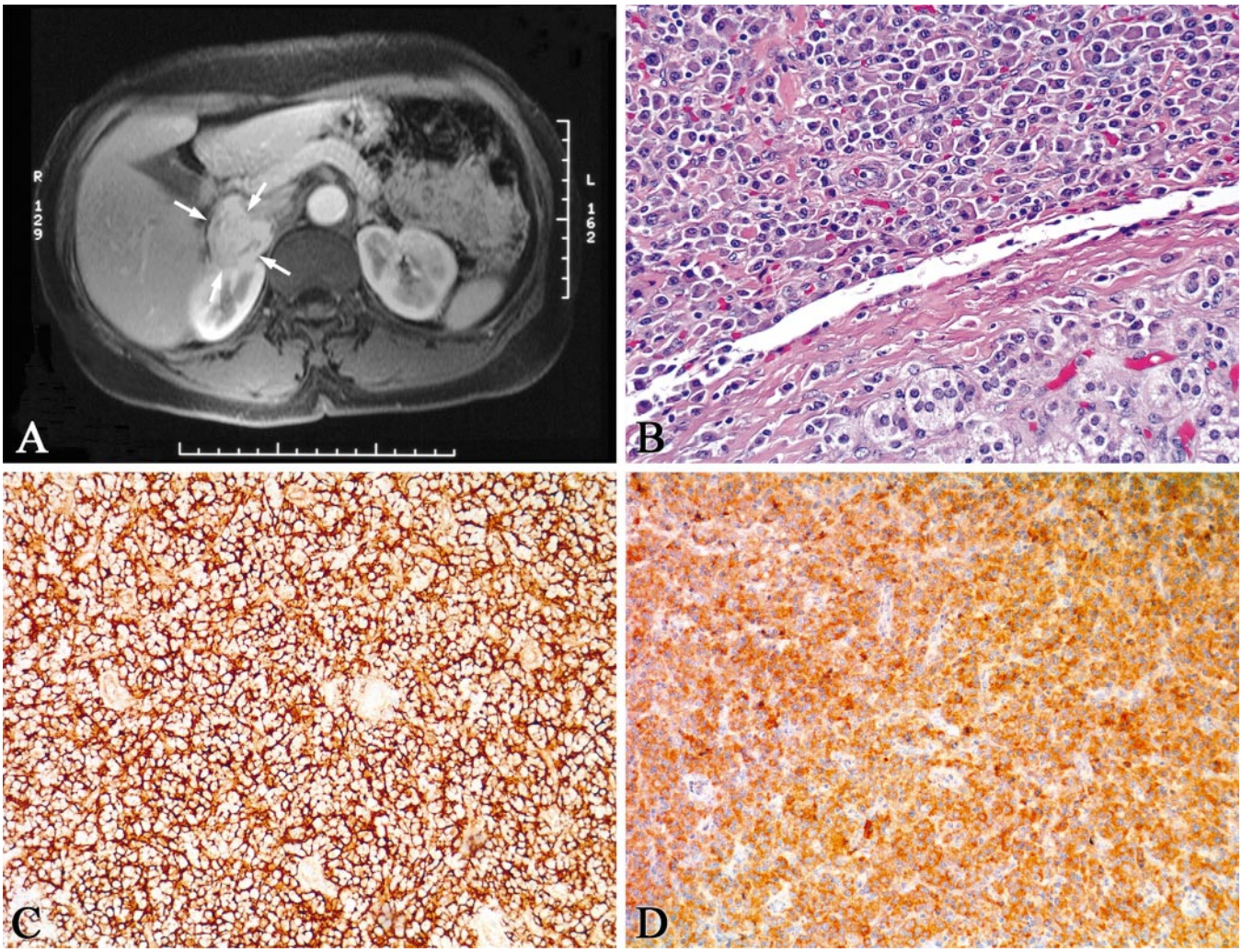
Corresponding author: Craig G. Rogers, MD, Department of Urology, Brady Urological Institute, Marburg 1, Johns Hopkins Hospital, 600 N Wolfe St, Baltimore, MD 21287 (e-mail: crogers6@jhmi.edu).




*A, Abdominal magnetic resonance image showing a 3.5-cm, right adrenal mass with heterogeneous enhancement consistent with malignancy (postcontrast, T1-weighted image with fat suppression). B, Microscopic view of surgically resected adrenal mass showing extraosseous (extramedullary) adrenal plasmacytoma. Well-circumscribed plasma cell tumor adjacent to benign adrenal cortical tissue. The plasma cells are unusually large and are cytologically atypical (hematoxylin-eosin, original magnification ×20). C, Immunohistochemical staining of resected adrenal plasmacytoma specimen for CD138 highlights the cytoplasmic membranes of the atypical plasma cells (original magnification ×10). D, Plasma cells from the resected adrenal plasmacytoma specimen show a monoclonal pattern of λ immunoglobulin light chain expression (immunohistochemical staining, original magnification ×10).*

where they are classified as malignant disorders (plasma cell myeloma) or benign conditions (monoclonal gammopathy of undetermined significance). Plasma cell myeloma is either multifocal or widely disseminated in the marrow cavity and is often complicated by osteolytic lesions, pathologic fractures, hypercalcemia, anemia, and an aggressive clinical course. Plasmacytomas are plasma cell neoplasms that are cytologically identical to plasma cell myeloma but present as solitary osseous or extraosseous (extramedullary) lesions.[1] Extraosseous plasmacytomas are rare, comprising less than 3% of all plasma cell neoplasms. Approximately 80% of extraosseous plasmacytomas occur in the head and neck, particularly the tissues of the upper respiratory tract. Other sites of involvement include the gastrointestinal tract, breast, thyroid, testis, bladder, retroperitoneum, and lymph nodes.[2]

To our knowledge, only 2 cases of adrenal plasmacytoma have been reported in the literature. Fujikata et al[3] reported on a large adrenal plasmacytoma with functional abnormalities that required open excision. Kahara et al[4] reported on a small, nonfunctioning, asymptomatic adrenal plasmacytoma with heterogeneous hyperintensity on T2-weighted MRIs that was resected laparoscopically. The present case is unique in that a contrast MRI was performed showing heterogeneous enhancement of the mass, providing further evidence beyond heterogeneous hyperintensity on T2-weighted images for a possible malignant process. Another unique feature is that a biopsy of the lesion was performed, although the biopsy specimen was nondiagnostic. A nondiagnostic rate of needle biopsy as high as 28% has been reported, which is likely due to sampling error. The ability to identify the adrenal lesion accurately in diagnostic aspirates is greater than 95%.[5]

The definitive diagnosis of plasmacytoma is based on the pathologic confirmation of a solitary plasma cell tumor, with or without a monoclonal gammopathy, and the absence of plasma cell myeloma on the bone marrow biopsy specimen. Moreover, patients with plasmacytomas

show no evidence of hypercalcemia, anemia, and renal insufficiency.

Treatment options for plasmacytoma include surgical resection and radiation therapy, given the relative sensitivity of plasma cell neoplasms to radiation. Although seldom indicated, chemotherapy has a much greater effect on solitary plasmacytomas than multiple myeloma.[6] The prognosis of extraosseous plasmacytoma is favorable, but local recurrence occurs in 30% of cases and dissemination in 40%. Progression to multiple myeloma occurs in 17% to 33% of cases, in contrast to the more aggressive solitary osseous plasmacytoma, which transforms to myeloma in greater than 50% of cases.[7]

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