

Answer

Extramedullary Plasmacytoma

Plasmacytomas are solitary masses of plasma cells, generally of bone marrow, accounting for 1% of all cancers and approximately 10% of hematologic malignant neoplasms.¹ Rarely, these plasma cell tumors arise entirely outside the bone marrow. The vast majority of cases occur in the head and neck, with 80% occurring in the nasal cavity, paranasal sinuses, and upper airways.² These cases generally present with symptoms related to mass effect, epistaxis, or airway obstruction. Microscopic disease of the spleen, liver, or lymph nodes has been found in up to 70% of cases at postmortem examination,³ and involvement of the entire gastrointestinal tract has been reported, although much less frequently. The incidence of pancreatic involvement based on autopsy studies has been reported at 2.3%,⁴ with reports of patients presenting with clinically significant disease such as obstructive jaundice being exceedingly rare.

Multiple myeloma, a disease of older individuals, has a reported mean age at diagnosis of 66 years. Extramedullary manifestations appear at a mean age of 50 years⁵ and are generally associated with a more aggressive form of the disease.⁶ Fewer than 25% of these patients are found to have monoclonal proteins in their serum or urine on laboratory examination.⁷

Treatment of extramedullary plasmacytoma is challenging. As most of these cases occur in the head and neck and these tumors are generally highly radiosensitive, curative radiation is the current treatment of choice, with reported local control rates ranging from 80% to 100%.⁷ For tumors arising in other sites, complete surgical resection should be considered.⁷ Adjuvant chemotherapy should be considered in patients with positive surgical margins, tumors larger than 5 cm, or high-grade disease.⁷

Our case of a 50-year-old previously healthy patient presenting with obstructive jaundice is an exceedingly rare diagnosis, with only a handful of case reports in a literature search. A pancreatic primary tumor, neuroendocrine tumor, or non-Hodgkin lymphoma must be strongly considered and ruled out. In this case, however, fine-needle aspiration biopsy clearly demonstrated monoclonal plasma cells (Figure 2). Immunohistochemistry staining was diffusely positive for λ light chain and CD138, confirming the diagnosis. Interestingly, some authors have suggested that smooth stenosis of the biliary

tree on endoscopic retrograde cholangiopancreatography should raise suspicion for plasmacytoma.⁸ The patient has received palliative pancreatic stenting and has shown good response to a chemotherapy regimen based on bortezomib and dexamethasone.

Accepted for Publication: January 25, 2012.

Correspondence: Alessandra Landmann, MD, Department of Surgery, University of Oklahoma Health Sciences Center, 818 16th St NW, Oklahoma City, OK 73106 (alessandra-landmann@ouhsc.edu).

Author Contributions: *Study concept and design:* Makipour, Morgan, Postier, and Pant. *Acquisition of data:* Landmann, Makipour, Morgan, Pant, Liu, and Anderson. *Analysis and interpretation of data:* Makipour, Postier, and Liu. *Drafting of the manuscript:* Landmann, Makipour, and Anderson. *Critical revision of the manuscript for important intellectual content:* Makipour, Morgan, Postier, Pant, and Liu. *Statistical analysis:* Landmann. *Obtained funding:* Postier. *Study supervision:* Makipour, Postier, and Pant.

Conflict of Interest Disclosures: None reported.

REFERENCES

1. Rajkumar VS. Clinical features, laboratory manifestations and diagnosis of multiple myeloma. *UpToDate*. 2011. <http://www.uptodate.com/contents/clinical-features-laboratory-manifestations-and-diagnosis-of-multiple-myeloma>. Accessed October 31, 2011.
2. Gupta P, Rice GD, Abraham K, Ghole V, Ketkar M. Extramedullary plasmacytoma of the pancreas and jejunum. *Clin Imaging*. 2009;33(3):240-243.
3. Jowitt SN, Jacobs A, Batman PA, Sapherson DA. Atypical progression of multiple myeloma with extensive extramedullary disease. *J Clin Pathol*. 1994;47(3):269-271.
4. Fischer A, Suhlrand MJ, Vogl SE. Myeloma of the head of the pancreas: a case report. *Cancer*. 1991;67(3):681-683.
5. Birjawi GA, Jalbout R, Musallam KM, Tawil AN, Taher AT, Khoury NJ. Abdominal manifestations of multiple myeloma: a retrospective radiologic overview. *Clin Lymphoma Myeloma*. 2008;8(6):348-351.
6. Hiller N, Goitein O, Ashkenazi YJ. Plasmacytoma of the pancreas. *Isr Med Assoc J*. 2004;6(11):704-705.
7. Soutar R, Lucraft H, Jackson G, et al; Guidelines Working Group of the UK Myeloma Forum; British Committee for Standards in Haematology; British Society for Haematology. Guidelines on the diagnosis and management of solitary plasmacytoma of bone and solitary extramedullary plasmacytoma. *Br J Haematol*. 2004;124(6):717-726.
8. Hirata S, Yamaguchi K, Bandai S, Izumo A, Chijiwa K, Tanaka M. Secondary extramedullary plasmacytoma involving the pancreas. *J Hepatobiliary Pancreat Surg*. 2002;9(1):111-115.