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 years[5] 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.

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