Letter to the Editor

Successful management of solitary gastric plasmacytoma in a 22-year-old woman using a new protocol for combination therapy

Sir,

Extramedullary plasmacytomas (EMPs) of the gastrointestinal system are rare tumors, with no established consensus on treatment. We present the case of a 22-year-old woman with primary gastric plasmacytoma (GP) who was successfully managed with a three-pronged combination of endoscopic mucosal resection (EMR), radiotherapy, and chemotherapy. The patient presented with fever, epigastric pain, and gradual weight loss for 6 months. Medical history was unremarkable, and routine laboratory investigations were normal, except for anemia (9.0 g/dL). Total serum protein was normal (7.22 g/dL), but albumin/globulin ratio was inverted (0.7). Upper gastrointestinal endoscopy showed chronic friable erosions at lesser curvature of the stomach, and entire ulcerated area was sampled for biopsy.

Histopathology revealed focal mucosal erosions and dense plasma cell infiltration in submucosa with interspersed inflammatory cells. Immunohistochemistry showed plasmacytoid cells positive for CD 138, CD 45, and Kappa light chain; and immunonegative for CD 20, CD 3, CD 5, and lambda light chain. MIB-1 index was 5% [Figure 1].

Whole-body positron emission tomography (PET) scan showed fluorodeoxyglucose avid focal thickening in gastric corpus, focal colonic hypermetabolism, and diffuse marrow uptake. Bone marrow studies showed erythroid hyperplasia and 2% mature plasma cells. Repeat endoscopy revealed ulcerations in the body of the stomach, normal colon, and solitary rectal ulcer. Active proctitis and plasmacytoid infiltration in ulcerated gastric mucosa were diagnosed on endoscopic biopsy. Serum protein electrophoresis, immunofixation electrophoresis, and free light chain assay revealed serum monoclonal gammopathy, with gamma heavy chain and kappa light chain monoclonal protein. Beta-2-microglobulin level was 2.24 mg/L. Urinary Bence Jones protein was not detectable. A final clinical diagnosis of multiple myeloma with extramedullary GP was made.

Considering the younger age of the patient, localized tumor, no regional lymphadenopathy, and uninvolved bone marrow; EMR was deemed adequate, and further treatment was based on the combination of intravenous bortezomib, oral dexamethasone, and regional radiotherapy localized to the stomach and perigastric lymph nodes. A total dosage of 45 Gy fractionated over 25 doses was administered. Complete treatment schedule was tolerated well by the patient.

Biochemical and bone marrow studies were repeated after 8 and 16 months. There was loss of monoclonal protein, reduction in total immunoglobulin levels, and normal marrow plasma cells; and the patient remains symptom-free till date.

EMPs of the gastrointestinal system are rare plasma cell neoplasms, and GPs account for 2%–5% of all EMPs.[1] Gastrointestinal symptoms are most common presenting feature, and endoscopy can reveal gastric nodules, polyps, mucosal infiltration, or ulcers.[2] Endoscopic findings simulate adenocarcinoma, and sufficient number of biopsies is recommended for accurate diagnosis.[3] Computed tomography, magnetic resonance imaging, and PET scans can aid diagnosis.[4] Differential diagnoses include lymphoplasmacytic and mucosa-associated lymphoid tissue lymphomas, plasma cell granuloma, and Castleman's disease. Predominant population of CD 20 negative and CD 138 positive plasma cells with the absence of centrocytes, follicles, or lymphoepithelial lesions, is typical of EMPs.[5]

Meta-reviews identified <20 cases of primary gastric EMP, predominantly in elderly males. Most cases have mucosal tumors, and there is no established treatment guideline. Endoscopic mucosal/submucosal resection has achieved complete remission in three cases. Systemic chemotherapy with or without radiotherapy or surgery is usual approach.[1,2,6] Combination therapy of bortezomib, dexamethasone, and surgery has proven effective in advanced cases.[1] A causal relationship with *Helicobacter pylori* infection has been postulated.[2,5,6]

Despite no long-term follow-up studies of GPs, the prognosis is favorable with a 70% disease-free survival at 10 years. The rate of progression of EMP to multiple myeloma ranges from 11% to 30%, at 10 years.[5] A combination protocol of EMR, local radiotherapy, and bortezomib chemotherapy with dexamethasone, as in this case, can be considered a safe and effective approach in cases of GPs limited to mucosa; with follow-up and endoscopic monitoring as indicated.

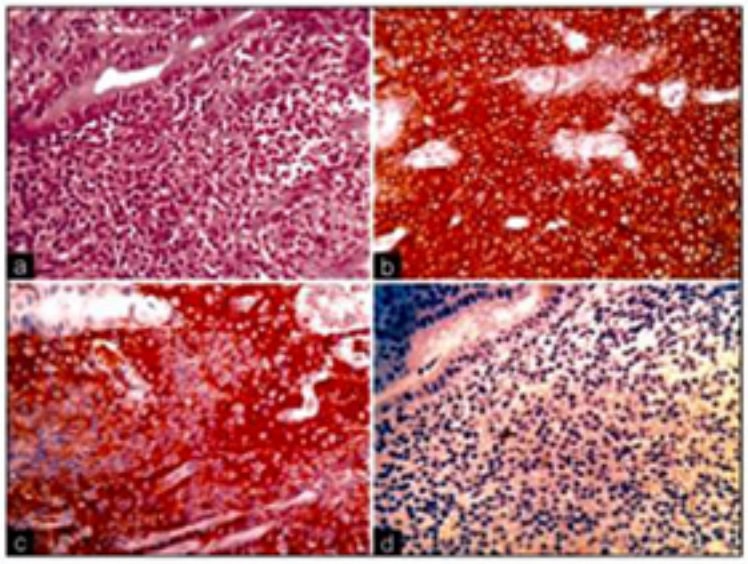

Figure 1: (a) Section of gastric endoscopic biopsy showing sheets of plasma cells in lamina propria (H and E, ×400). (b) Immunohistochemistry on gastric biopsy section showing membranous positivity of plasmacytoid cells for CD138 (Immunoperoxidase, ×400). (c) Immunohistochemistry on gastric biopsy section showing Kappa light chain positivity in tumor cells (Immunoperoxidase, ×400). (d) Immunohistochemistry on gastric biopsy section showing tumor cells negative for lambda light chains (Immuno[illegible]se, ×400)

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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