

Background

- Plasma cell neoplasms are defined by a monoclonal proliferation of a single clone of immunoglobulin-producing plasma cells
- Extramedullary plasmacytomas are rare plasma cell dyscrasias that arise outside of the bone marrow, most commonly in the head or neck
- Extramedullary multiple myeloma involves the gastrointestinal (GI) tract in <5% of cases, often presenting with non-specific symptoms; here we present one such case of uncommon GI involvement of a plasma cell tumor

Case Description

62-year-old male with PMHx of active lambda light chain multiple myeloma presented with acute onset global fatigue, melena, and coffee ground emesis

Diagnosis: acute blood loss anemia in the setting of a GI bleed

Initial presentation:

Vitals and labs: blood pressure 90/62 mmHg, heart rate 95 bpm, hemoglobin (Hb) 3.9 g/dL

Exam: abdomen soft and nontender to palpation

Day 1 – Inpatient Admission:

Transfused with 3 units of packed red blood cells (pRBC) with appropriate Hb response to 6.5 g/dL

Endoscopy (EGD) revealed nonbleeding cratered duodenal ulcer with adherent clot (Figure 1), not amenable to endoscopic intervention

Day 2 – Continued Hospitalization:

Transfused with additional 1 unit of pRBC with appropriate Hb response to 8.1 g/dL

Day 3 – Discharged Home:

Hb up-trended without intervention to 8.6 g/dL prior to discharge

Hemodynamically stable, with no evidence of continued GI bleed

VIR embolization deferred

Discharged on PO Pantoprazole 40mg BID

Plan for follow up CBC in 1 week, repeat EGD in 1 month

Day 4 – Deceased:

Less than 24 hours following discharge, re-presented in cardiac arrest, with melanic stool and significant oropharyngeal blood

Resuscitation attempts were unsuccessful

Autopsy determined cause of death to be secondary to complications of extramedullary gastroduodenal disease from the patient's plasma cell myeloma

Plasma cell infiltration with resultant transmural duodenal ulceration involving the gastroduodenal artery had resulted in acute hemorrhage (Figures 2-6)

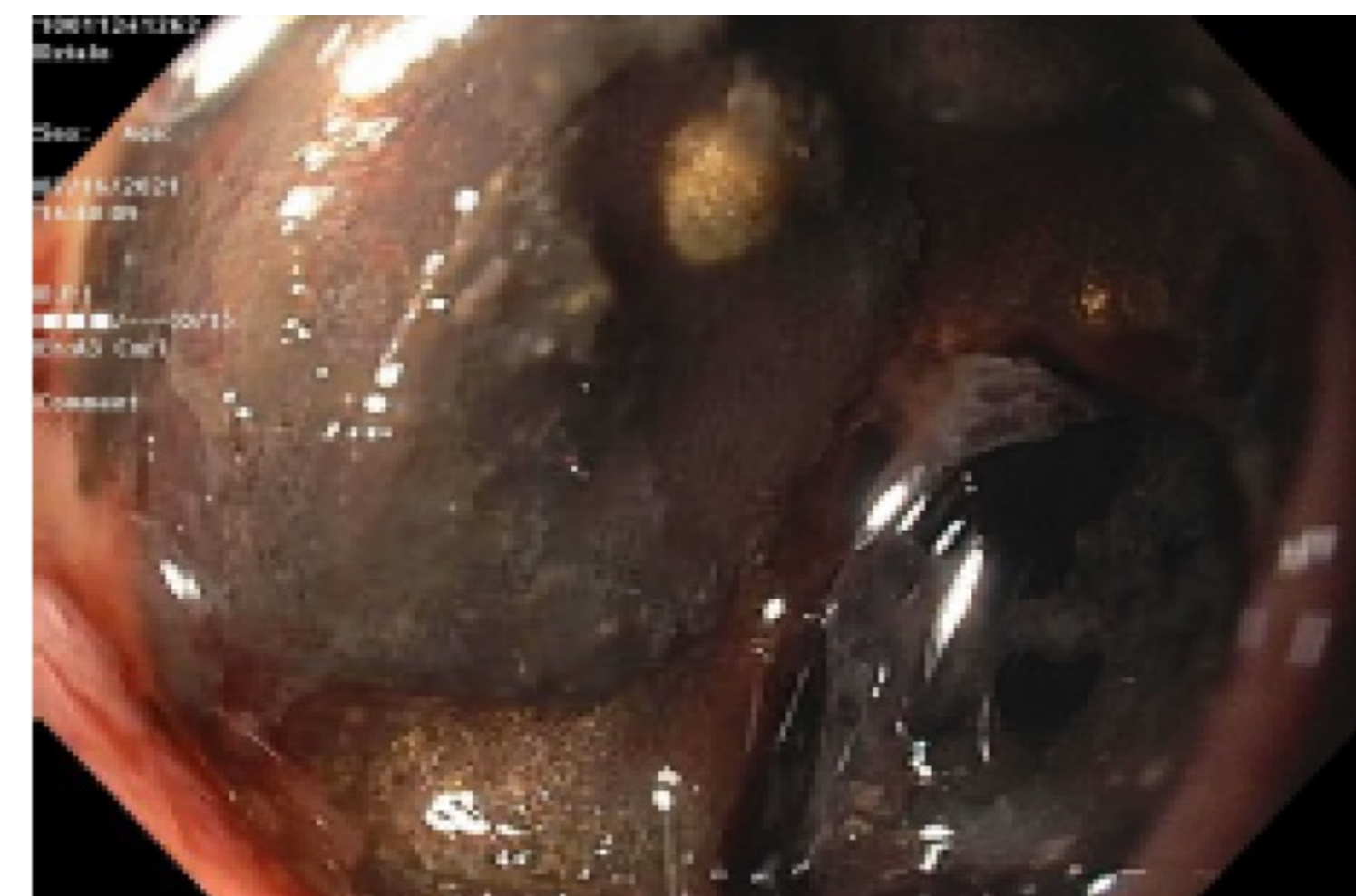


Figure 1: Ulcer at the duodenal bulb, completely obstructing duodenum

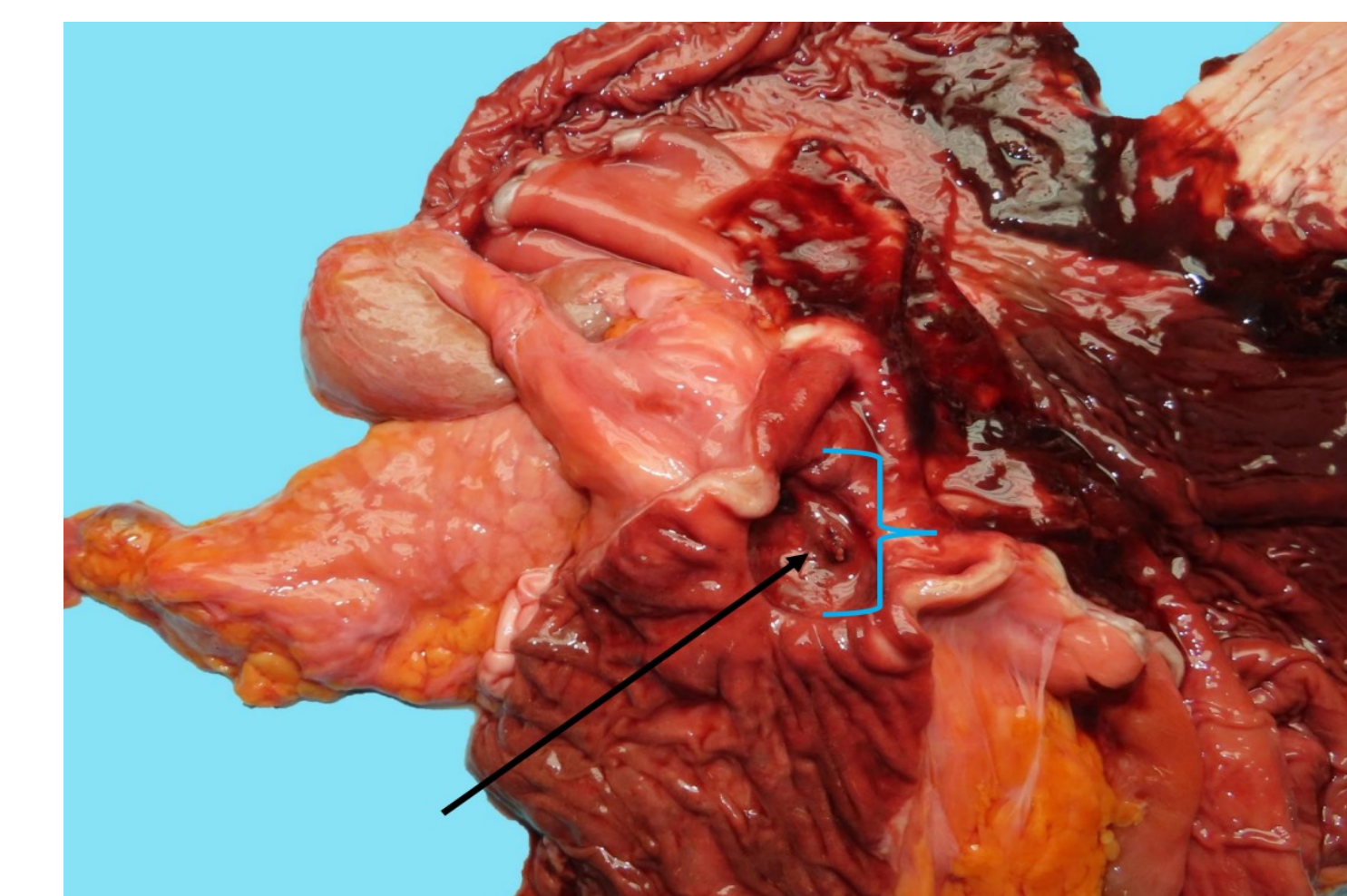


Figure 2: Gastroduodenal ulcer (brackets) with eroded gastroduodenal artery (arrow)

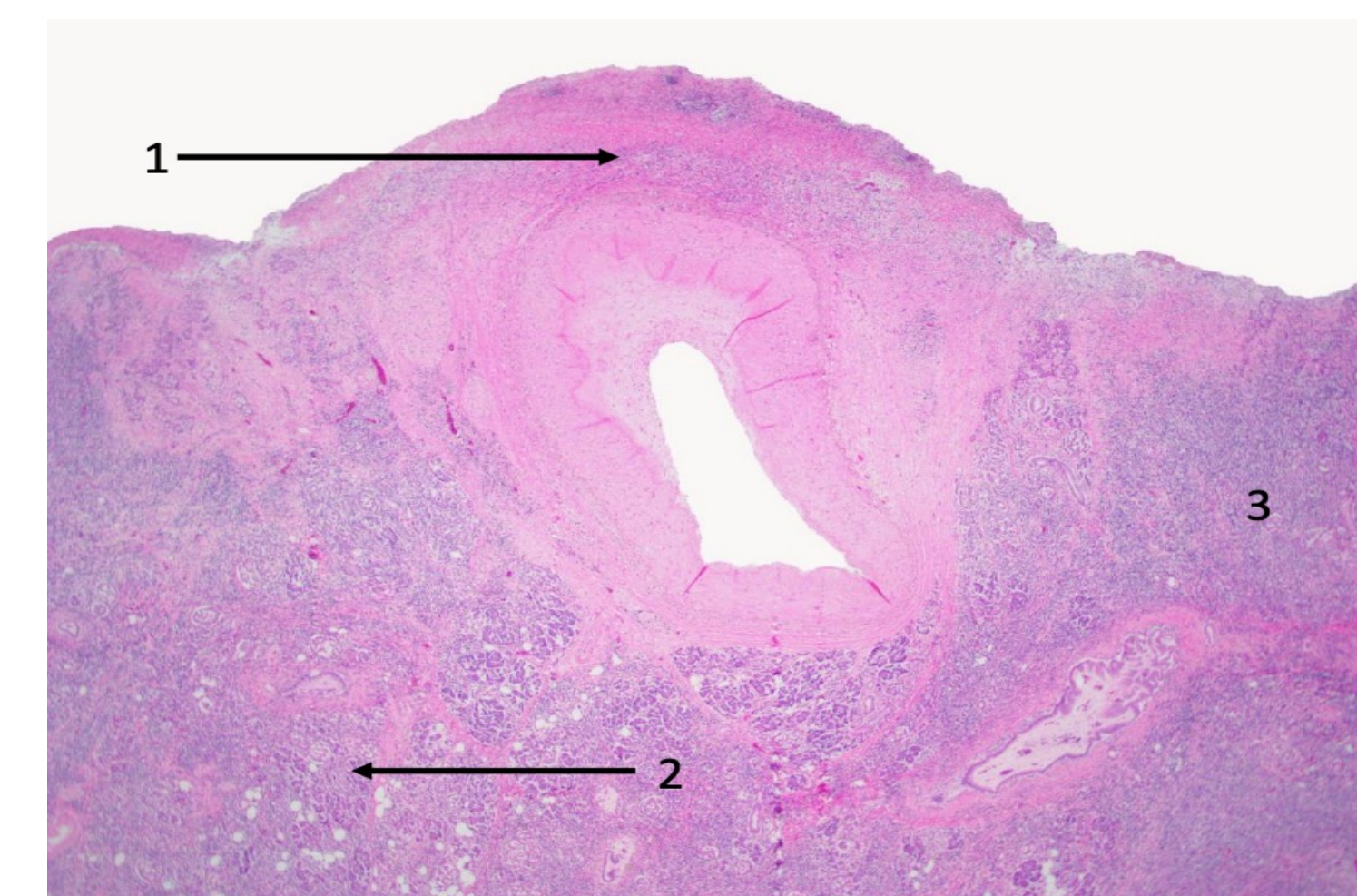


Figure 3: H&E, 2x: Artery with overlying fibrin cap (1), and surrounding pancreatic parenchyma (2) with plasma cell infiltrate (3)

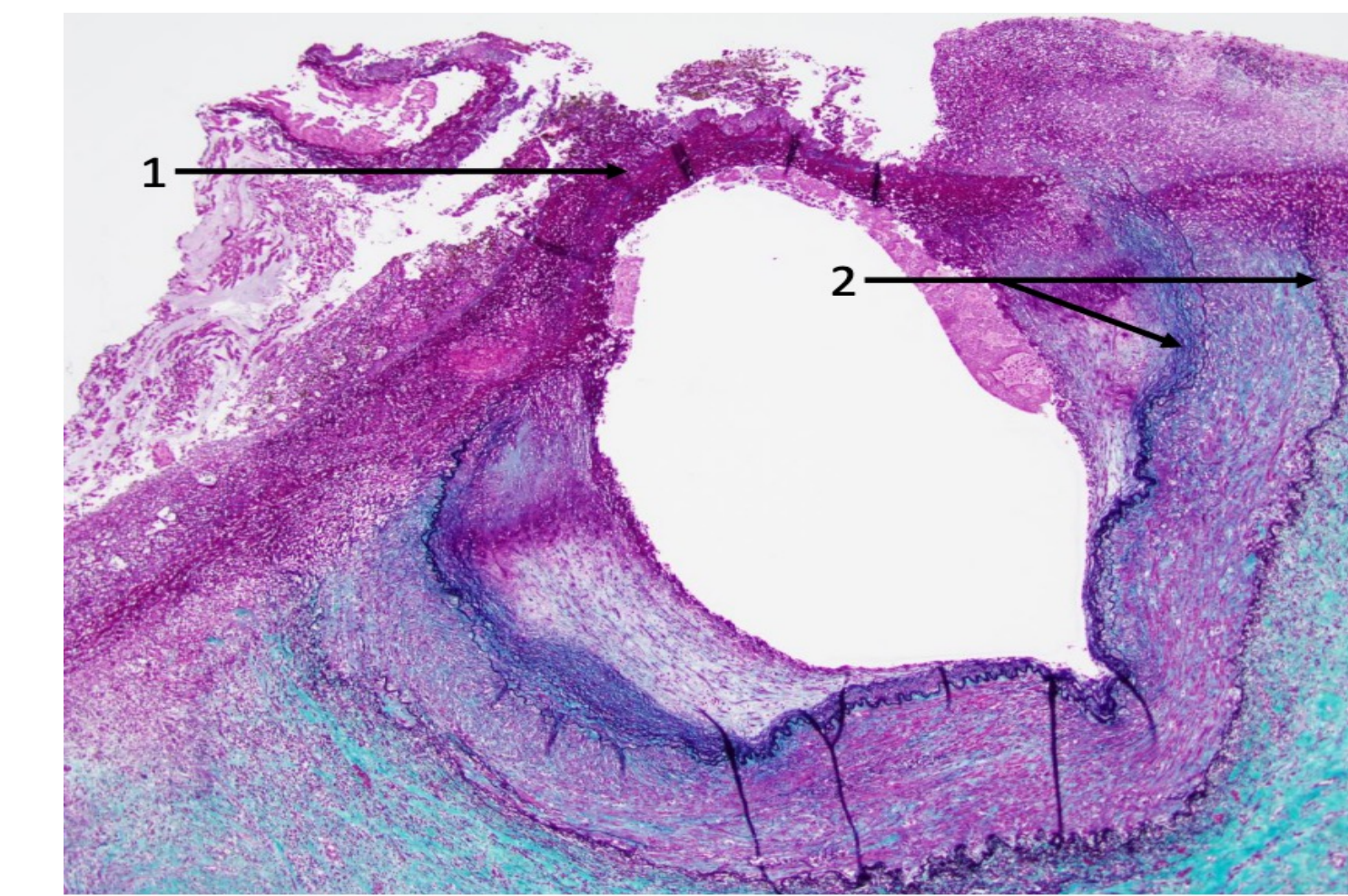


Figure 4: Elastic trichrome, 2x: Eroded/ruptured artery (1) with partial mural fibrinoid necrosis and disruption of the internal and external elastic layer (2)

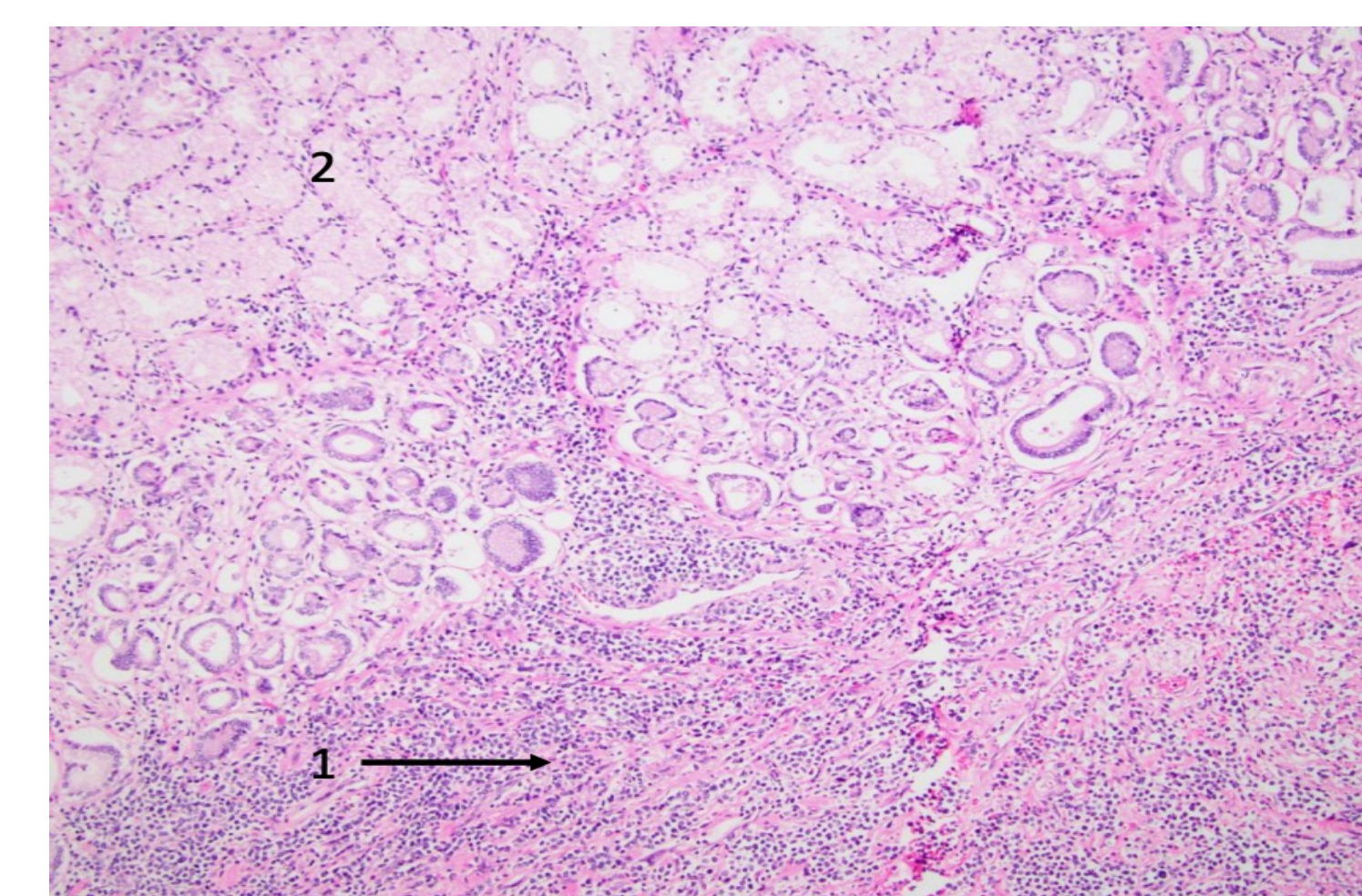


Figure 5: H&E, 10x: Plasma cells (1) infiltrating Brunner's glands (2) of the deep duodenal mucosa

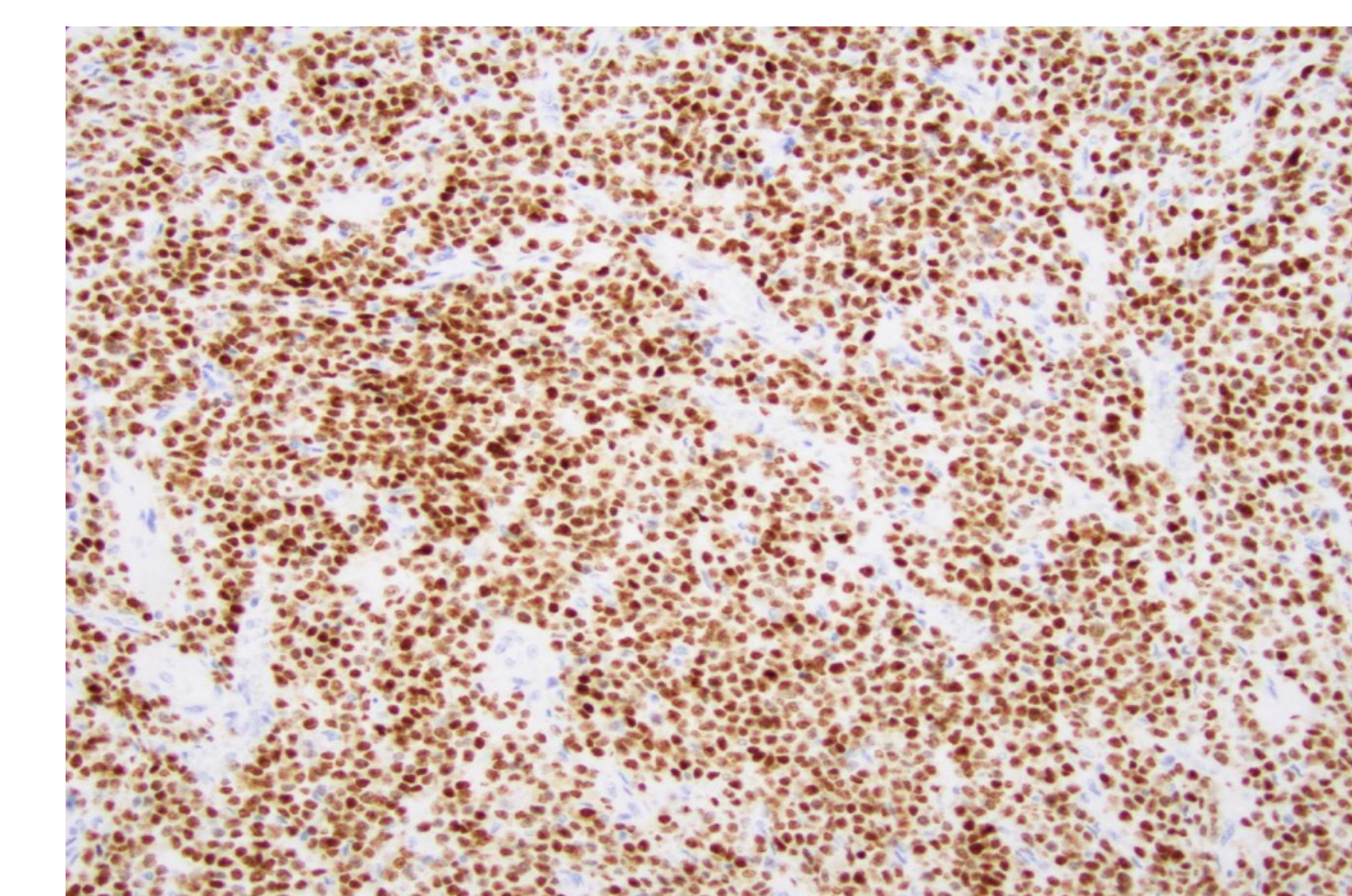


Figure 6: MUM1, 20x: Plasma cell infiltrate

Discussion

- Though typically limited to the bone marrow, extramedullary plasma cell myeloma can present as plasmacytoma in up to 20% of cases
- GI involvement of such plasmacytomas is incredibly rare; within the GI tract, involvement of the small bowel is least likely
- GI extramedullary disease often presents with abdominal pain in the setting of small bowel obstruction and acute or chronic GI bleeding anywhere along the alimentary canal
- While rare, plasmacytomas should be considered in patients with known plasma cell neoplasm who present with an upper GI bleed, especially in cases where the identified lesion is not amenable to EGD therapy, as disease onset is swift and mortality rates are high

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