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Case Report

A rare case report: Extramedullary Plasmacytoma of Duodenum without concomitant Multiple Myeloma, an unusual presentation

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ABSTRACT:
Extramedullary plasmacytoma (EMP) is a rare plasma cell neoplasm that arises outside the bone marrow, accounting for only 1% of plasma cell neoplasms. The gastrointestinal (GI) tract is an uncommon site for EMP, with duodenal involvement being extremely rare. We report the case of a 70-year-old female who presented with persistent abdominal pain, nausea, vomiting, and generalized weakness for three months. Upper gastrointestinal endoscopy revealed a bulky duodenal papilla, and biopsy was performed. Histopathological and immunohistochemical evaluation confirmed the diagnosis of extramedullary plasmacytoma. Extensive hematological and biochemical investigations ruled out concomitant multiple myeloma. Due to its rarity, duodenal EMP poses a diagnostic challenge and necessitates a multidisciplinary approach for optimal management. This case highlights the importance of histopathological and immunohistochemical evaluation in diagnosing duodenal plasmacytoma. Due to its rare presentation and nonspecific symptoms, early recognition and differentiation from multiple myeloma are crucial for appropriate treatment.

Keywords: Extramedullary Plasmacytoma; Plasma cell neoplasm; duodenum

INTRODUCTION:

Plasma cell neoplasms constitute mature B-cell malignancies exhibiting abnormal monoclonal proliferation of plasma cells, with multiple myeloma (MM) being the most common, characterised by diffuse bone marrow and other organ involvement; < 5% of patients present with solitary plasmacytomas (SPs), a single localised tumor of clonal plasma cells in either bone as solitary bone plasmacytomas (SBPs) or in soft tissues as extramedullary plasmacytomas (EMPs)(1,2) The incidence of EMP accounts for approximately 1% of plasma cell neoplasms (PCNs) and rarely involves the gastrointestinal tract (GI)(3,4)

CASE REPORT:

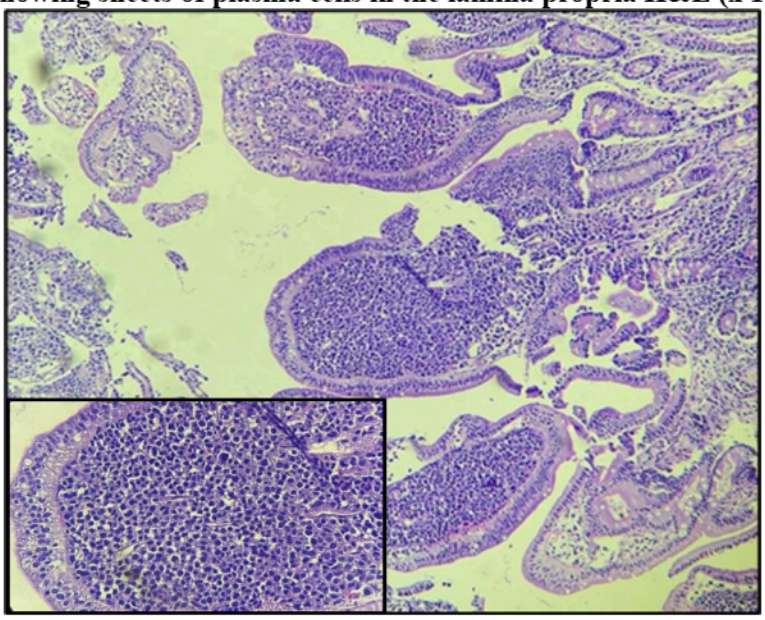
A 70-year-old female presented with the chief complaint of pain in the abdomen to our hospital. The pain was persistent and associated with nausea and vomiting. She had generalised weakness for 3 months. Her past medical history showed no family history of neoplastic, genetic, or metabolic disorders.

No abnormal findings were observed in the preliminary laboratory tests performed which included biochemical and microbiological tests. Laboratory tests showed Hb 12.5 g/dL and calcium level of 10.5 mg/dL. Additionally, normal serum BUN/creatinine, sodium, potassium, albumin, and LDH levels were detected. No organism growth was observed on culture.

The patient was taken up for Upper GI Endoscopy which revealed bulky papilla in the duodenum and biopsy was taken. Diagnosis was reached through histopathological examination coupled with immunohistochemistry of the endoscopic biopsy which ruled out lymphoma, sarcoma, carcinoma or other malignancies.

The histopathological examination of the biopsy specimen showed duodenal papillae with unremarkable mucosal epithelium and distended lamina propria. The lamina propria showed sheets of monomorphic small round blue cells with few of them showing plasmacytoid morphology. (Figure 1)

Figure 1: Tumor biopsy showing sheets of plasma cells in the lamina propria H&E (x 100) and inset (400x)



Immunohistochemically, these neoplastic cells showed immunoreactivity for CD138, Kappa light chain and were negative for CK, CD20, and lambda light chains. (Figure 2 and Figure 3)

Figure 2: Immunohistochemistry displaying positivity for CD138 expression (100x)

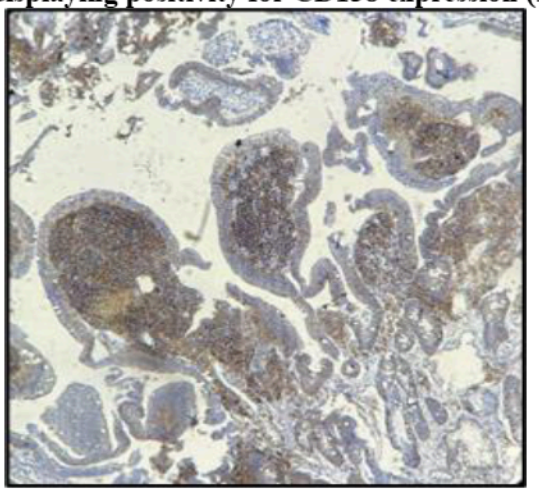
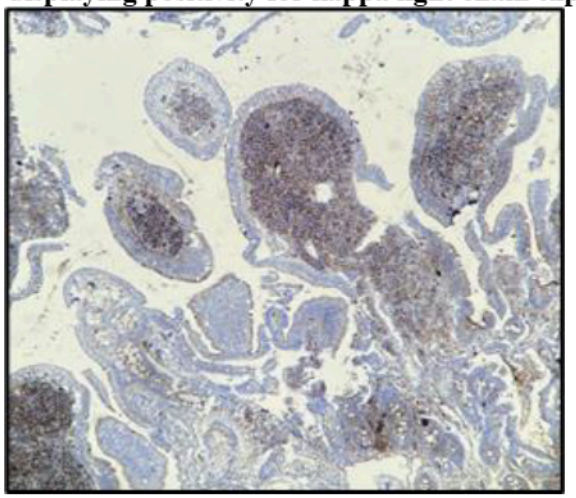


Figure 3: Immunohistochemistry displaying positivity for kappa light chain expression (100x)



These findings supported a diagnosis of plasmacytomas. Further laboratory work up showed liver and kidney function test results within the normal ranges.

The patient underwent a thorough hematologic evaluation, including bone marrow, bone imaging, and urine examination. Bone marrow aspirate and trephine biopsies showed no evidence of PCM. The urine analysis was negative for Bence Jones protein. Serum

and urine protein electrophoresis showed no monoclonal spike in the gamma-globulin region. This evaluation excluded multiple myeloma and confirmed that her condition was extramedullary plasmacytoma. She was taken up for surgical resection after 6 months of diagnosis following radiotherapy after referral to a higher centre.

DISCUSSION:

Plasma cell neoplasms entail several entities, such as solitary plasmacytoma of bone, EMP, and MM. Primary plasmacytoma is of two types. The medullary plasmacytoma arises from the bone marrow and the extramedullary plasmacytoma which usually arises in the soft tissues of the head and neck and upper respiratory tract and rarely gastrointestinal tract, liver, spleen, pancreas, breast, and skin.(5)(6)

Diagnosis can be established by presence of the tumor having monoclonal plasma cell pathology, confirming the absence of any other lesions, and ensuring the presence of no more than 10% clonal plasma cells in the bone marrow and absence of multiple myeloma-related tissue organ damage (Calcium > 11.5mg/dl, creatinine> 1.73mmol/L, anemia; normocyte normochrome and hemoglobin <10 g/dl or more than 2g/dl reduction in Hb, bone lesions; lytic lesions, severe osteopenia or pathological fractures)(7)

In this case scenario, we present a rare example of EMP without a concomitant diagnosis of multiple myeloma occurring in a 70-year-old female.

A study using immunohistochemical methods confirmed the monoclonal plasma cell nature of the cells with cell markers, e.g., CD 138, which indicates the necessity of performing an evaluation for a differential diagnosis from other cancers. The demonstration of light chains allows the phenotyping of whether the process is monoclonal, i.e., KAPPA or LAMBDA chain.

A negative bone marrow biopsy, the absence of bone lysis, and normal electrophoresis of blood immunoglobulins allows the possibility of a multiple myeloma to be excluded.(8)

To our knowledge, only few cases of plasmacytoma presented with a duodenal ulcer were reported in the literature which implies the extremely rare presentation. Patients with duodenal plasmacytoma may remain asymptomatic until there is significant tumor growth. This case report highlights the importance of histopathological examination and implication of IHC in cases of GI malignancies.

CONCLUSION:

This case implies that EMP of the small intestine causes a diagnostic dilemma because of its rare clinical presentation and nonspecific symptoms. Diagnosis of plasmacytoma necessitates further workup for multiple myeloma since treatment modalities are significantly different for both types of plasma cell neoplasms. Histopathological examination confirms the diagnosis and leads to an opportunity of a multidisciplinary approach for the treatment and thus a better prognosis.

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REFERENCES:

1. Fend F, Dogan A, Cook JR. Plasma cell neoplasms and related entities evolution in diagnosis and classification. Virchows Archiv. 2023 Jan 21;482(1):163–77.
2. Zhu X, Wang L, Zhu Y, Diao W, Li W, Gao Z, et al. Extramedullary Plasmacytoma: Long-Term Clinical Outcomes in a Single-Center in China and Literature Review. Ear Nose Throat J. 2021 May 17;100(4):227–32.
3. Alexiou C, Kau RJ, Dietzfelbinger H, Kremer M, Spiess JC, Schratzenstaller B, et al. Extramedullary plasmacytoma: tumor occurrence and therapeutic concepts. Cancer. 1999 Jun 1;85(11):2305–14.
4. LOPES DA SILVA R. Extramedullary plasmacytoma of the small intestine: Clinical features, diagnosis and treatment. J Dig Dis. 2012 Jan 21;13(1):10–8.
5. Lee SY, Kim JH, Shin JS, Shin C, In KH, Kang KH, et al. A case of extramedullary plasmacytoma arising from the posterior mediastinum. Korean J Intern Med. 2005 Jun;20(2):173–6.
6. Jizzini MN, Shah M, Yeung SCJ. Extramedullary Plasmacytoma Involving the Trachea: A Case Report and Literature Review. J Emerg Med. 2019 Sep;57(3):e65–7.
7. Mosebach J, Thierjung H, Schlemmer HP, Delorme S. Multiple Myeloma Guidelines and Their Recent Updates: Implications for Imaging. RöFo - Fortschritte auf dem Gebiet der Röntgenstrahlen und der bildgebenden Verfahren. 2019 Nov 28;191(11):998–1009.
8. Kamath S, Pratap D, Shetty KC, B. A rare diagnosis of nasal mass: extramedullary plasmacytoma. International Journal of Otorhinolaryngology and Head and Neck Surgery. 2018 Apr 26;4(3):853.