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# Multiple Myeloma with the Primary Gastric Manifestation

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## Abstract

Multiple myeloma is an immunoproliferative monoclonal disorder of plasma cells, characterized by an expansion of atypical cells, which produce monoclonal immunoglobulin. This neoplasm has several clinical presentations: solitary osseous plasmacytoma, plasmocytic leukemia or extramedullary plasmocytoma. Plasmocytoma are rare in the gastrointestinal tract. This case present a histologically confirmed case of primary gastric plasmocytoma.

## Introduction

Multiple myeloma (MM) is an immunoproliferative monoclonal disorder of plasma cells (plasmocytoma), characterized by an expansion of atypical cells, which produce monoclonal immunoglobulin (protein M). It presents as a discrete, solitary mass of neoplastic monoclonal plasma cells in either bone (most frequently) or more rarely, soft tissue MM constitutes about 1% of all malignant neoplasms with morbidity in Europe reported to be 4,5/100000/year and 5-years survival is 35% [1]. It affects more often males (M:F=2:1) and the age of presentation is ca. 62 for men and 61 for women.

Multiple myeloma has several clinical presentations: solitary osseous plasmacytoma (SOP; which is the most common), plasmocytic leukemia or extramedullary plasmocytoma (EMP) [2]. Primary EMPs (PEMPs) are rare, constituting fewer than 5% of all plasma cells neoplasms. Progression to disseminated plasma cell myeloma (PCM) is infrequent, occurs in approximately 15% of cases.

Extramedullary plasmocytoma could be a solitary tumor localized in bone itself (6%) or outside the bones (2%) [3] especially in the soft tissues of the head and neck. Soft tissue plasmacytomas of the head and neck tend to involve the nasal cavity or nasopharynx, rather than the oral cavity [4]. Other localizations of EMP are rare and not well characterized in the literature.

Primary gastric plasmocytoma is an extremely rare presentation of all extramedullary plasmocytoma. It constitutes 2-5% of EMCs. This type of tumor was described first time in 1928 year by Vasiliu i Popa [5], and since that time about 100 of new cases have been described [6]. This kind of tumor occurs in older patients with symptoms typical for GI tract disorders, such as weight loss, abdominal pain or upper gastrointestinal bleeding, which may delay diagnosis and treatment [5].

## Clinical Presentation

87 years old man was admitted to the hospital with symptoms of fever reaching 38.5°C, lasting for several weeks and not responding to treatment. He also complained of weakness, night sweats, loss of appetite and dry cough. No other symptoms of malignancy were reported. Co-morbidities at the time of presentation included: coronary heart disease, hypertension, atrial fibrillation, COPD.

In the laboratory results, mild normocytic anemia RBC 2.74 M/ul, Hgb 7.9 g/dl, HCT 23.6%, MCV 86.2 fL, WBC 7.2 K/ul, PLT 149.00 K/ul), low level of iron (2.18 umol/l, ferritin 520 ug/l) increased level of ferritin and inflammatory markers (CRP 120,9 mg/l, fibrinogen 9 g/L ) were observed. In routine urine analyses the proteinuria was present (0.3 g/l; while kidney function parameters such as creatinine, BUN were normal (creatinine 121.7 umol/l, BUN 9.3 mmol/l). In order to further address potential proteinuria, serum protein electrophoresis was performed but showed normal profile with no monoclonal protein present. Further diagnoses of fever of unknown origin included exclusion of all sites of inflammation were excluded (chest X-ray, USG of the abdomen, serial blood and urine cultures). Occult blood test in faeces was positive which led to subsequent gastroscopy and colonoscopy. The gastroscopy has indicated chronic inflammation of gastric mucosa (contain friable plaques in the whole stomach mucosa) with a small protrusion of mucosa (<5 mm) without ulceration. The urease test was negative. In the colonoscopy no abnormalities was found. Biopsies were taken of mucosal protrusion. In the histopathological analyses neoplastic infiltration of the gastric trunk was found, and after the immunological phenotyping multiple myeloma was diagnosed. The immunological phenotype was described: CD138+, LCA-, CK-, chromogranin -, CD68-, proteinogram S 100-, CD1a- (Figures 1 and 2).

In the subsequent detailed urine analysis monoclonal light chain

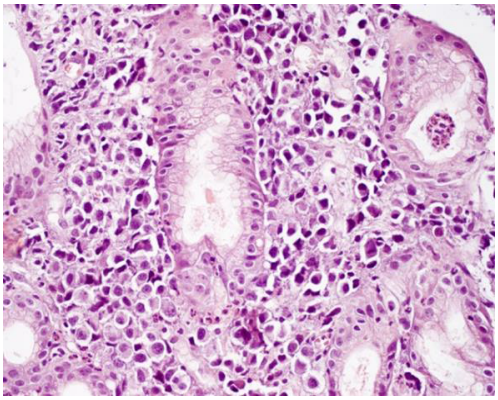

**Figure 1:** Neoplastic infiltration is present in the stroma between the glands, and also partially destroys mucous membrane. Tumor cells were small, with eosynophylic cytoplasme and polymorphic nucleuses, with visible nucleoluses. H&E.

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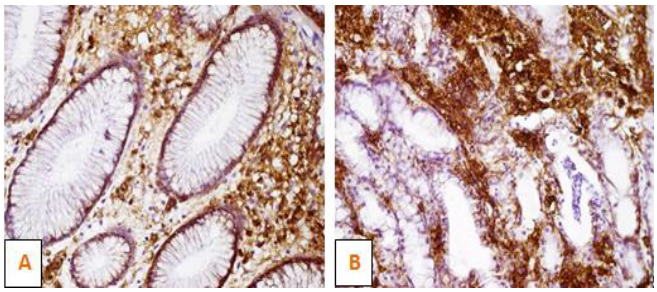


**Figure 2:** Tumor cell immunophenotype: positive CD138 (Figure 2a) and kappa chains (Figure 2b), negative pan-cytokeratynine (pan-CK), common leukocytic antigens (LCA), CD68 i CD1a.

immunoglobulin zone was found and in the blood high levels of free kappa chain 1310 mg/l were present (normal range: 3.3-19.4). The level of free chains lambda was normal 10.2 mg/l (n: 5.71–26.3). The ratio of free kappa to lambda chains was therefore highly increased to 128 (n: 0.26–1.65). In most cases of plasmocytoma the plasmocytes synthesize complete M protein (pertained to IgG or IgA class), but in 10% of cases these cells produce light chains or their portions only. These are then secreted in the urine as a Bence-Jones protein. In such situation subtype of plasmocytoma: light chain disease is diagnosed (as it was in this case). Chest X-ray of flat bones revealed no osteolytic changes. Calcium levels: both total (2.17 mmol/l) and ionized (1.05 mmol/l) were normal. In bone marrow aspiration infiltration of plasmocytic cells was present in about 60–70%. In the past (about 10 years ago) patient had trepanobiopsy (because of pancytopenia) – no pathologic changes were found at that time.

On the basis of these results and the clinical presentation the multiple myeloma subtype - light chain disease was diagnosed with the stomach involvment. Because of poor general condition and co-morbidities (when diagnosis was made patient had severe congestive heart failure with hypotension) patient did not qualify for hematopoietic cell transplantation, but for therapy with systemic glycocortycosteroids. Apart from this, the patient was treated with antibiotics, anti-fungal drug and immunoglobulin. Despite of treatment patient died.

## Discussion

Usually primary gastric EMP is diagnosed in elderly patients (like in this case), but there are no typical symptoms. Sometimes patients complain of pain or epigastric discomfort, weight loss or gastrointestinal bleeding. This patient's main problem was fever and weakness. There was no obvious symptom of gastrointestinal bleeding, but occult blood test was positive. The differential diagnosis includes stomach carcinoma and other non-Hodgkin's lymphomas like: lymphoplasmacytic lymphoma, follicular lymphoma, monocytoid B-cell lymphoma and mucosa-associated lymphoid tissue lymphomas (MALT).

In the literature there are some similar cases described. Esfandyari [7] presents a case of patient who was the same as in our patient a lesion in the stomach and involvement of the bone marrow. Their patient also was in advanced age. There were also changes in the stomach mucosa like friable plaques, but in our patient the tumor was also present. Gaur describes a patient with primary solitary bone plasmocytoma who had a gastric relapse of this tumor. The recurrence of the tumor took place after radiotherapy. What important at the beginning there was only tumor in vertebral column, but recurrence was present in bone marrow and stomach (but symptoms become from gastrointestinal tract – the same as in our case) [8]. On the basis of results we can't arbitrate between the primary gastric plasmocytoma with progression to typical multiple myeloma or there was primary multiple myeloma with the same time infiltration of the stomach by plasmatic cells.

In published data surgical treatment [9], inclouding endoscopic submucosal dissection [10] or irradiation sometimes with chemotherapy is recommended. Surgical excision is recommended in all cases of extramedullary plasmocytomas, with one exception – extramedullary plasmocytomas with the head and neck localization [9]. In such situation radiotherapy alone is suggested [11]. If the tumor is excised without adequate surgical margin some authors recommend radiotherapy after the primary surgery [9,12]. Till now, there is no evidence that adjuvant chemotherapy is beneficial. There are some evidences that are good results of chemotherapy alone. In the literature there are different treatment plans: bortezomib alone, or bortezomib+dexamethasone [13]. But it seems that *Helicobacter Pylori* infection can have influence on the prognosis [14]. Eradication of this infection can cause regression of neoplastic changes localized in stomach [14] but it is not a role [15]. That patient had negative test for *H. Pylori* [16].

Light chain disease – plasmocytoma can have different prognosis: in about 15% of cases the progression to the typical tumor [16], in 13% become disseminated extramedullary plasmocytoma, in 7% of cases local lymph nodes became affected. Most patients with an extramedullary plasmocytomas die because of unrelated causes [17].

## Clinical Practice Points

Primary gastric EMP can have different clinical presentation. Not necessarily symptoms include gastrointestinal bleeding; sometimes fever could be the only symptom. What is important the size of the primary tumour is not a good predictor of increased risk of progression to the typical tumour and dissemination? In this case, despite of small size of tumour in the stomach the medullary involvement is present.

When we have patient with FUO (fever of unknown origin) we have to think about neoplasm (including plasmocytoma even if the protein electrophoresis is unchanged).

## Conclusion

We have reported here a case of elderly patients in whom multiple myeloma initially localized to gastric mucosa rapidly progressed to systemic disease leading to patient's death. It is therefore important to include plasmocytoma in the differential diagnosis of atypical presentation of gastric tumours even in the absence of other symptoms of multiple myeloma.

### References

1. Anderson KC, Alsina M, Bensinger W, Biermann JS, Chanan Khan A (2007) National Comprehensive Cancer Network (NCCN) Multiple myeloma. Clinical practice guidelines in oncology J Natl Compr Canc Netw 5: 118-147.
2. Bolek TW, Marcus RB Jr, Mendenhall NP (1996) Solitary plasmacytoma of bone and soft tissue. Int J Radiat Oncol Biol Phys 36: 329-333.
3. Mendenhall WM, Mendenhall CM, Mendenhall NP (2003) Solitary plasmacytoma of bone and soft tissues. Am J Otolaryngol 24: 395-399.
4. Liebross RH, Ha CS, Cox JD, Weber D, Delasalle K, et al. (1999) Clinical course of solitary extramedullary plasmacytoma. Radiother Oncol 52: 245-249.
5. Krishnamoorthy N, Bal MM, Ramadwar M, Deodhar K, Mohandas KM (2010) A rare case of primary gastric plasmacytoma: an unforeseen surprise. J Cancer Res Ther 6: 549-551.
6. Nolan KD, Mone MC, Nelson EW (2005) Plasma cell neoplasms. Review of disease progression and report of a new variant. Surg Oncol 14: 85-90.
7. Esfandyari T, Abraham SC, Arora AS (2007) Gastrointestinal plasmacytoma that

caused anemia in a patient with multiple myeloma. Nat Clin Pract Gastroenterol Hepatol 4: 111-115.

8. Gaur S, Oo TH, Aish LS, Mansoor S (2005) Gastric relapse of solitary bone plasmacytoma. Am J Clin Oncol 28: 325-326.
9. Han YJ, Park SJ, Park MI, Moon W, Kim SE, et al. (2014) Solitary extramedullary plasmacytoma in the gastrointestinal tract: report of two cases and review of literature. Korean J Gastroenterol 63: 316-320.
10. Park CH, Lee SM, Kim TO, Kim DU, Jung WJ, et al. (2009) Treatment of solitary extramedullary plasmacytoma of the stomach with endoscopic submucosal dissection. Gut Liver 3: 334-337.
11. Tan J, Lade S, Harrison S, Opat S, Mac Manus MP (2012) Complete remission of localised gastric plasmacytomas following definitive radiotherapy. J Med Imaging Radiat Oncol 56: 328-331.
12. Galieni P, Cavo M, Pulsoni A, Avvisati G, Bigazzi C, et al. (2000) Clinical outcome of extramedullary plasmacytoma. Haematologica 85: 47-51.
13. Katodritou E, Kartsios C, Gastari V, Verrou E, Mihou D, et al. (2008) Successful treatment of extramedullary gastric plasmacytoma with the combination of bortezomib and dexamethasone: first reported case. Leuk Res 32: 339-341.
14. Lu HS, Xu YF, Gan MF (2010) Primary gastric plasmacytoma associated with Helicobacter pylori infection: a report of two cases with different prognosis. Int J Hematol 92: 174-178.
15. Kawasaki N, Nakada K, Kawasaki Y, Suzuki Y, Masuda K, et al. (2009) Primary gastric plasmacytoma refractory to Helicobacter pylori eradication therapy. Dig Endosc 21: 37-39.
16. Dimopoulos MA, Kiamouris C, Moulopoulos LA (1999) Solitary plasmacytoma of bone and extramedullary plasmacytoma. Hematol Oncol Clin North Am 13: 1249-1257.
17. Maskin LP, Díaz MF, Hlavnicka A, Rubio H, de la Cámara A, et al. (2008) Gastrointestinal bleeding secondary to multiple gastric plasmacytoma. Am J Clin Oncol 31: 100-101.