Case Report

Case of Gastric Adenocarcinoma Presenting as Microangiopathic Hemolytic Anemia (MAHA) and secondary Myelofibrosis with vertebral Metastasis

Pan K[1], Chakrabarti S[2], Choudhary R[3], Sarkar A[4], Panchadhyayee S[5]

[1]Koushik Pan*, [2]Subrata Chakrabarti*, [3]Rajdip Choudhury*, [4]Anup sarkar*, [5]Sujoy Panchadhyayee[x]

Post graduate trainee*, Department of Medicine, SSKM, IPGME & R, Kolkata, India. RMO[x], Department of Medicine, SSKM, IPGME & R, Kolkata, India.

Address of corresponding Author: Dr Koushik Pan, Email: panjoy86@gmail.com

Abstract

It is very rare for the primary presentation of a gastric malignancy to be with bone metastases. Also it is equally rare for MAHA to be a presenting feature of gastric adenocarcinoma as a paraneoplastic syndrome. This is a case report is 55years old female who presented with widespread vertebral deposits with diffuse bone marrow infiltration along with Coomb's negative haemolytic anemia with thrombocytopenia and schistocytes in peripheral blood typical of MAHA. The combination of MAHA and bone infiltration in gastric adenocarcinoma is a very rare entity.

Key words: MAHA, Myelofibrosis, Gastric adenocarcinoma, Metastasis

Case report

A 55 year female presented to outpatient Department with severe back pain, for last 3 months, along with weakness, fatigue, exertional dyspnoea for last 2 month. There was no history of trauma to trigger her back pain or any limb weakness.

History of contact with Tuberculosis was negative. She had one episode of hemetemesis 15 days back. Her past history was unremarkable. She never received any chemotherapy or radiation exposure. Physical examination revealed severe pallor, mild icterus, and bilateral pitting edema. No lymphadenopathy, clubbing or petechiae/purpura was noted. Chest examination was normal. Spleen was palpable 2cm below left costal margin, non tender.

Lab investigations Includes: Hb-45g/l, PCV-16.2%, TLC-8400/cu mm, N-56%, L-26% , PLT-32000/cu mm, ESR-93mm/1st hr. Peripheral Blood Smear examination showed mixture of Normocytic, Normochromic RBCs and Microcytic, Hypochromic RBCs, anisocytosis, polychromasia, few tear drop cells, schistocytes, nucleated RBCs(16/100WBCs). Reticulocyte count was 16.9%.

Renal function test, Electrolyte and Calcium/ Phospahte levels were within Normal limit. Presence of DIC was ruled out as PT, INR, APTT were normal.

Manuscript received: 13th Aug 2013
Reviewed: 26th Aug 2013
Author Corrected: 19th Sep 2013
Accepted for Publication: 30th Sep 2013

Total bilirubin-3.1mg/dl, unconjugated fraction-2.2 mg/dl, conjugated fraction-0.9 mg/dl, SGOT-37U/L, SGPT-54U/L, ALP-3636U/L, GGT-36U/L suggesting bone origin of the raised ALP.

Other investigations included LDH -699U/L, Uric acid-9.8 mg/dl, serum haptoglobin-0.08mg/dl. The blood picture established the presence of Microangiopathic Hemolytic Anemia.

Results of other investigation concluded following results. ECG-sinus tachycardia, CXR- Cardiomegaly & increased bronchovascular markings. USG - mild splenomegaly. Hb electrophoresis, serum ceruloplasmin was normal, direct and indirect Coomb's test was negative, ANA-negative. As bone marrow aspiration resulted dry tap, BM biopsy was done. Upper GI Endoscopy has shown presence of antral solitary gastric ulcer from where biopsy was taken for histopathology.

Bone trephine biopsy: Reactive erythroid and megakaryocytic hyperplasia in some places and secondary myelofibrosis in other places. A search for malignancy was made for which CT thorax and abdomen was ordered.

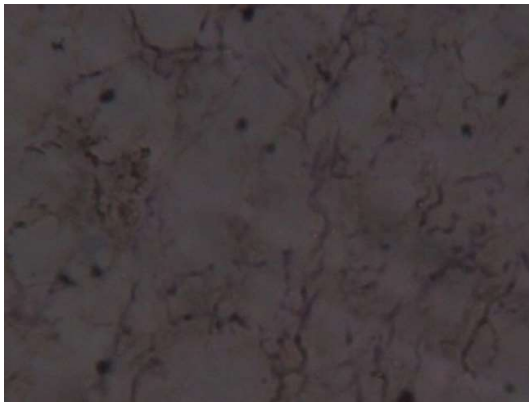
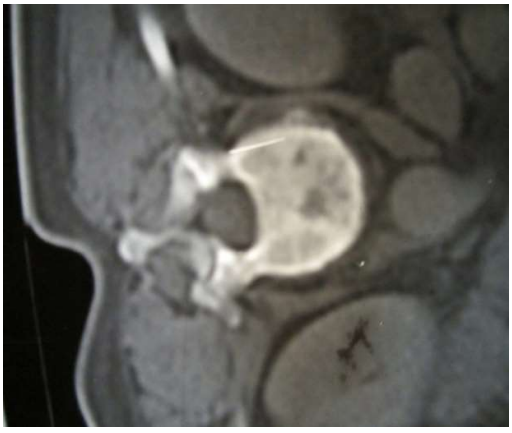


Fig 1-figure of bone marrow trephine biopsy showing myelofibrosis.



CT scan thorax and abdomen: has shown focal left pleural thickening, lytic destruction of D9 and D10 vertebrae with prevertebral mass. CT guided FNAC was suggestive of osteosclerosis

Fig 2- showing lytic destruction of dorsal vertebrae

MRI dorsal spine showed patchy areas of altered marrow signal intensity, showing heterogenous contrast, enhancement involving the bodies and posterior element of multiple dorsal vertebrae-suggestive of marrow infiltrative disorder.

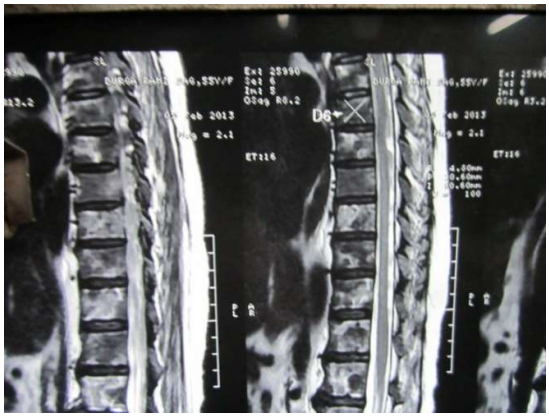


Figure 3: MRI spine showing diffuse vertebral destruction at multiple levels.

Patient continued to have a downhill course inspite of being administered supportive treatment including blood transfusions. Finally the gastric ulcer HPE report reached us which showed presence of poorly differentiated Adenocarcinoma. Patient expired within 2 days after another bout of massive hemetemesis.

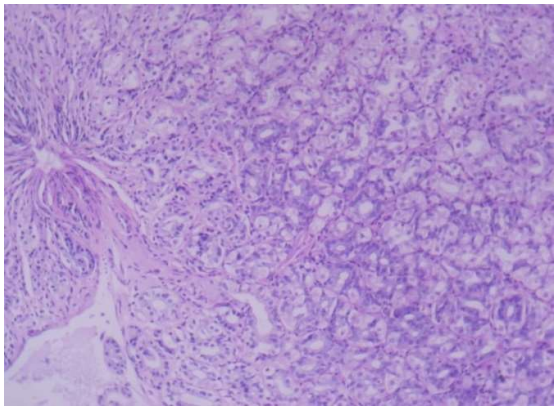


Fig 4: showing gastric adenocarcinoma in the HPE of gastric ulcer

Discussion

Microangiopathic hemolytic anemia can occur as a paraneoplastic syndrome in cancer patients, and it may present as the first manifestation. The most common tumors associated with MAHA are gastric, breast, and lung cancers[1-3]. A Korean study reported that 14 (25.5%) out of 55 MAHA patients had gastric cancer[3].

Cancer-associated MAHA (CA-MAHA) is a rare and fatal complication of malignant tumors. Most CA-MAHA patients die within a few weeks after the diagnosis, and the most common cause of death is infection[4]. Our patient died within 4 weeks. On average, the median time interval between the initial diagnosis of MAHA and the diagnosis of underlying malignancy is 6 days. Bone marrow examinations and bone scans are usually performed to detect CA-MAHA[5].

Our patient was diagnosed with CA-MAHA on the basis of the findings of bone marrow examination and histopathological examination of gastric ulcer although several studies have suggested that fibrinoid necrosis of the bone marrow and tumor cell emboli of the arteries, arterioles, and capillaries are the causes of CA-MAHA, its pathogenesis remains unclear[6].

Tumour-derived factors, procoagulants, decrease in von Willebrand factor (vWF)-cleaving protease ADAMTS13

play a role in CA-MAHA[6]. Bone metastases tend to occur in invasive cancers like Bormann types III and IV. Examination of the histology has shown that more than 80% of bone metastasis from gastric cancer was poorly differentiated adenocarcinoma.

Thoracic and lumbar vertebrae are the most frequent sites of bone metastasis[7] although there are reported cases of deposits in pelvis[8]. Patients with bone metastases have mean survival times of less than 5 months with the longest survival period reported being 3 years[6].

Bone pain and respiratory symptoms are observed more frequently in CA-MAHA than in non-CA-MAHA[7]. Our patient experienced bone pain and dyspnea.

There is no definitive treatment of choice for CA-MAHA. The low platelet count and hemoglobin level make red blood cell and platelet transfusion obligatory. Chemotherapy is the most reliable treatment of choice for the underlying cancer[1].

Funding: Nil

Conflict of interest: Nil

Permission from IRB: Yes

References

1. Elliott MA, Letendre L, Gastineau DA, Winters JL, Pruthi RK, Heit JA. Cancer-associated microangiopathic hemolytic anemia with thrombocytopenia: an important diagnostic consideration. Eur J Haematol 2010;85(1):43-50.

2. Kim YJ, Choi HM, Jung BG, Lee CK, Chung SY. Two cases of microangiopathic hemolytic anemia in patients with gastric cancer. Korean J Hematol 1990;25:575-9.

3. Youk DW, Choi MK, Kim KH, Kim HK, Park CW, Kim CC, Lee KS, Kim DJ, Kim BK. Microangiopathic hemolytic anemia in patients with gastrointestinal malignancy. Korean J Hematol 1983;18:207-14.

4. Arkenau HT, Müssig O, Buhr T, Jend HH, Porschen R. Microangiopathic hemolytic anemia (MAHA) as paraneoplastic syndrome in metastasized signet ring cell carcinomas: case reports and review of the literature. Z Gastroenterol 2005;43(8):719-22.

5. Hahn JS, Lee DH, Lee SJ, Min YH, Ko YW. A clinical study on microangiopathic hemolytic anemia. Korean J Hematol 1991;26:263-79.

6. Etoh T, Baba H, Taketomi A, Nakashima H, Kohnoe S, Seo Y, Fukuda T, Tomoda H. Diffuse bone metastasis with hematologic disorders from gastric cancer: clinicopathological features and prognosis. Oncol Rep 1999;6(3):601

7. Carstens SA, Resnick D. Diffuse sclerotic skeletal metastasisas an initial feature of gastric carcinoma. Arch Intern Med1980;140(12):1666-8

8. Chung YS, Choi TY, Ha CY, Kim HM, Lee KJ, Park CH, Fitzpatrick LA. An unusual case of osteoblastic metastasis from gastric carcinoma. Yonsei Med J2002;43(3):377–80

.....

How to cite this article?

Pan K, Chakrabarti S, Choudhary R, Sarkar A, Panchadhyayee S. Case of Gastric Adenocarcinoma Presenting as Microangiopathic Hemolytic Anemia (MAHA) and secondary Myelofibrosis with vertebral Metastasis. *Int J Med Res Rev* 2013;1(4):212-215. doi:10.17511/ijmrr.2013.i04.10

.....