thoracic-full abdominal computed tomography (CT) scan was performed in order to detect malignancies. On abdominal CT, 1 to 2 lymphadenopathies of 15 × 12 mm in the peripancreatic and perigastric area and pathological wall thickening (2.5 cm) at the level of the gastric corpus were detected. Gastroduodenoscopy revealed an edematous, partly ulcerated lesion protruding from the mucosa that extended to the angularis from the gastric cardia. Gastric tissue biopsy report indicated poorly differentiated adenocarcinoma (signet-ring cell predominant). The case was accepted as MAHA secondary to gastric carcinoma (ADAMTS-13 activity tested earlier was within normal limits at 84%). While waiting for the results of the biopsy and the other tests, the patient underwent 14 sessions of TPE in total. Following TPE, platelet count increased from 25×10^9/L up to 162×10^9/L, fragmented erythrocyte rate in peripheral smear decreased more than 75% and other laboratory findings of hemolysis (LDH, bilirubin, etc.) significantly decreased. The patient was transferred to the medical oncology clinic for the chemotherapeutic treatment of the primary gastric carcinoma.

Conclusion: Malignancy-associated MAHA is generally linked to a poor prognosis and the optimal treatment is not known. However, there is evidence for the importance of promptly initiating an effective antineoplastic regimen and it is also noteworthy that administering therapeutic plasma exchange (TPE) therapy for the purpose of immunocomplex removal could be beneficial in patients with symptoms of bleeding and thrombosis.

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A signet ring cell carcinoma presented as refractory acquired thrombotic thrombocytopenic purpura

U. Malkan[1,*], M. Albayrak[1], H. Ozturk[1], M. Reis Aras[1], B. Saglam[1], K. Dogan[2]

[1] *University of Health Sciences, Diskapi Yildirim Beyazit Training and Research Hospital, Department of Hematology, Ankara, Turkey*
[2] *University of Health Sciences, Diskapi Yildirim Beyazit Training and Research Hospital, Department of Pathology, Ankara, Turkey*

Objective: Microangiopathic hemolytic anemia (MAHA) can be observed as a paraneoplastic syndrome (PS) in certain tumors. MAHA related signet ring cell carcinoma (SRCC) of an unknown origin is very infrequent. Herein we present a SRCC case presented with refractory acquired thrombotic thrombocytopenic purpura (TTP).

Case report: 35 years old men applied to emergency service with fatigue and headache on January 2020. In his anamnesis he had a history of alcoholic pancreatitis. His physical examination was normal except the neurological symptoms which are temporary loss of consciousness and disorientation. His laboratory tests resulted as white blood cell 9020/μL, hemoglobin 3.5 g/dL, platelet 18,000/μL, MCV 110.7 fl, urea 58 mg/dL, creatinine 0.84 mg/dL, AST 68 u/L, ALT 33 u/L, indirect bilirubin 1.88 mg/dL, LDH 2257 u/L, reticulocyte 0.1, haptoglobulin <8 mg/dL, INR 1.42, Prothrombin time 13.2, fibrinogen 184 mg/dL, coombs negative. He had consulted to our clinic with bicytopenia and hemolysis. Schistocytes, micro-spherocytes and thrombocytopenia were observed in his blood smear. Microangiopathic hemolytic anemia was present and he was considered as thrombotic thrombocytopenic purpura. Plasma exchange treatment was initiated however he was refractory to this treatment. He had epistaxis and blurred vision during the follow-up. Superficial hemorrhages on the edges of the optic disc and roth spots were detected. Pain had emerged in his right arm. Doppler ultrasonography revealed the occlusion of cephalic vein with non-recanalized thrombus in the subacute process from the antecubital level at the forearm level. Thorax and abdomen computerized tomography (CT) resulted as liver 220 cm, spleen 14 cm, minimal pleural effusion, thickening of minor curvature in stomach corpus with hepatogastric and paraceliac lymphadenopathy. As a result of CT endoscopic examination was planned. Bone marrow investigation by our clinic resulted as the metastasis of adenocarcinoma. Ulcerations and necrosis was observed by gastric endoscopy procedure. Biopsy was taken during endoscopic intervention which resulted as signet ring cell carcinoma. He was transferred to oncology clinic for his treatment. Unfortunately he died in one month after his transfer.

Conclusion: Only 40% of TTP cases have the complete pentad and in 75% of the cases there is a triad of microangiopathic hemolytic anemia, thrombocytopenia, and neurological findings. In our case there was no acute kidney failure, however all the other features favored TTP, and diagnosis was made without the kidney failure. MAHA may be seen as a PS in some tumors, especially gastric cancers. Tumor related MAHA is generally accompanied by bone marrow (BM) metastases. As a result, BM investigation may be used as the main diagnostic method to find the underlying cancer. Total plasma exchange is usually performed in the treatment of cancer-associated TTP, however fewer than 20% of the cases respond to plasma exchange. Likely, our case did not respond to plasma exchange treatment either. The clinical course of cases with tumor related MAHA is usually poor, and these cases are usually refractory to plasma exchange treatment. In conclusion, physicians should suspect a malignancy and BM involvement when faced with a case of refractory TTP.

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