

中文題目：一位多發性骨髓瘤病人合併髓外漿細胞瘤

英文題目：Multiple myeloma with extramedullary plasmacytomas

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Introduction: Soft-tissue extramedullary plasmacytoma is uncommon in patients with multiple myeloma. Incidence of Extramedullary plasmacytoma is low. There are three types of extramedullary myeloma as local growth, hematogenous spread, or triggered by invasive surgical procedures. Local growth type involves the soft-tissue mass arising from focal bones as vertebrae, ribs and skull. Hematogenous spread can cause multiple nodules in the breast, liver or other organs.

Case report: A 63-year-old woman with refractory kappa light-chain multiple myeloma presented with intermittent epigastric pain and dark urine one year after receiving an autologous peripheral blood stem cell transplantation. Physical examination revealed icteric sclera and tenderness in the epigastric region. Laboratory testing revealed as follows with reference ranges: aspartate aminotransferase, 66 IU/L(reference range, <41 IU/L); alanine transaminase, 93 IU/L(reference range, <41 IU/L); direct bilirubin, 5.9 mg/dL(reference range, <0.2mg/dL); total bilirubin, 8.5 mg/dL(reference range, 0.3 to 1mg/dL); lipase, 1165 IU/L(reference range, 11 to 82IU/L). Computed tomography of the abdomen revealed swelling of the gallbladder, diffuse enlargement of the entire pancreas that caused dilatation of the pancreatic ducts (Figure 1a). Esophagogastroduodenoscopy revealed multiple submucosal tumors at the body and fundus of the stomach (Figure 1b). Biopsy from the stomach and pancreas showed diffuse infiltration of plasma cells. The CD138 stain reacted positively. And the CD3 and CD20 showed negative. The endoscopic findings, image discoveries, and the histological results were consistent with extramedullary involvement of multiple myeloma. A common bile duct stent was placed for the obstructive jaundice and it improved. Ixazomib, lenalidomide, and dexamethasone were prescribed for myeloma. However, *Enterococcus faecium* and *Pseudo. aeruginosa* caused biliary tract infection with *Pseudo. aeruginosa* bacteremia happened. Antibiotics use with Teicoplanin and Doripenem were given but the patient died one month after admission..

Discussion: There are many mechanisms about extramedullary myeloma spread have been reported. Adhesion molecules decrease as VLA-4, CD44, and P-selectin can

cause the dissemination of myeloma cells.^{3,4} CCR1, CCR2, and CXCR4 are chemokine receptors less active in extramedullary plasmacytoma.⁵ Increased angiogenesis factor as VEGF, MMP-9, angiopoetin-1, CD31, are related to severity of disease. All these could lead to the plasma cells from bone marrow microenvironment and become extramedullary plasmacytoma.

The extramedullary lesions can involve any tissue or organ, but the most common site is the submucosal lymphoid tissue of the upper respiratory tract.^{1,2} Gastrointestinal tract involvement is rare, accounting for only 10% of extramedullary plasmacytomas. The incidence of pancreatic involvement of extramedullary plasmacytomas was extremely rare with less than 70 cases reported until the present. Here we report is the rare case of extramedullary plasmacytomas of stomach and pancreas involvement diagnosed at the mean time.

No promising therapy has been established for extramedullary disease; thus, the prognosis of this disease is worse.² Kumar S et al proposed the therapies of combination with lenalidomide, cyclophosphamide, and dexamethasone may improve the outcome of extramedullary plasmacytoma. In this case we used the Ixazomib, lenalidomide, and dexamethasone for the patient but the patient still died of infection. Further research about mechanism of extramedullary plasmacytoma dissemination and spread could produce more specific therapies.

There were more and more cases about extramedullary plasmacytoma. If patients with multiple myeloma diagnosed, more intensive care about extramedullary plasmacytoma should be considered. And early diagnosis of extramedullary plasmacytoma with multiple myeloma will improve the prognosis of disease.

References:

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