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LETTER TO THE EDITOR

A large polyp in the rectum: not always an epithelial lesion

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Dear Editor:

Solitary extramedullary plasmocytoma (SEP) is an exceedingly rare entity accounting for less than 5 % of all plasma cell dyscrasias, such as monoclonal gammopathy of undetermined significance (MGUS), multiple myeloma (MM), solitary plasmocytoma of the bone, and monoclonal immunoglobulin deposition diseases. SEP is usually found in the upper respiratory tract and secondary in the gastrointestinal tract, with only 6.2 % arising in the large bowel [1]. Only fifteen case of ano-rectal SEP were reported in the English literature.

The diagnosis of SEP requires demonstration of a monoclonal plasma cell infiltration in organs other than the bone marrow without evidence of systemic involvement [2].

SEP patients mainly manifest with local masses and relevant symptoms, which are nonspecific and depend on the site and spread of the tumor. In the low GI tract, obstruction, rectal bleeding, and abdominal pain were more commonly described.

Among plasma cell neoplasia, SEP shows the best prognosis with 10-year overall survival rate of 70 %. Although a transformation into MM has been described in a small percentage of case (~15 %), patients with SEP that progressed to MM had a 100 % 5-year survival rate [3].

We reported a case of rectal SEP in a 60-years-old man presented with a 1 month of history of painless hematochezia and diarrhea. Past medical and surgical history of ischemic heart disease and femur fracture was reported.

At physical examination, no palpable abdominal or rectal mass were detected.

Colonoscopy revealed one small polyp in the cecum and three polyps in the rectum, two small and one of large size. Endoscopic mucosal resection (EMR) was performed for the largest.

At macroscopic examination, the large rectal polyp, of 4×3×2.5 cm of size, showed a smooth surface superficially ulcerated. The surgical margin was inked in black. At section, the specimen showed a fleshy and multicystic appearance and was involved in toto. All sections were processed for histological analysis.

The three small size polyps were tubulo adenoma with low grade dysplasia.

Histological analysis of the largest polyp showed massive infiltration by medium-sized cells, characterized by abundant pale to eosinophilic cytoplasm and eccentric nuclei with a cartwheel arrangement of the chromatin. Neither mitosis nor apoptotic figures were found. No residual rectal glands were found. Congo red staining showed no amyloid deposits.

At immunohistochemistry, tumor cells revealed diffuse reactivity for CD138 and for immunoglobulin kappa light chains. Immunostaining for lambda light chains, CD20, CD45 were negative on tumoral cells. Ki67 showed a low proliferation rate. Immunostaining for CD3 revealed some accompanying T cells CD3+.

A diagnosis of rectal plasmocytoma was retained.

As rectal plasmocytomas are extremely rare, reactive plasmacytosis, plasma cell granuloma, and lymphoma with plasma cell differentiation (mucosa-associated lymphoid tissue (MALT), lymphoblastic, and immunoblastic) have to be excluded in order to avoid misdiagnosis.

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The most challenging differential diagnosis is between a plasmocytoma and a MALT lymphoma with marked plasma cell differentiation, as they may have a similar histological appearance.

In MALT lymphoma, the neoplastic infiltrate is composed of B lymphocytes and plasma cells. In our case, the absence of a small B lymphocyte infiltrate and the absence of expression of CD20 by the plasmocytic cell lead us to rule out the diagnosis of a an indolent B lymphoma with marked plasma cell differentiation.

A reactive plasmacytosis and a plasma cell granuloma have been excluded as they originated from polyclonal plasma cells and in our case a restriction for the immunoglobulin kappa light chains was observed.

We also excluded more aggressive lymphoma showing differentiation into immature plasma cells (lymphoblastic and immunoblastic) as no significant atypia or high proliferation index were observed in our case.

Further investigations were made to determine whether the tumor was solitary or a localization of a multiple myeloma. MRI showed neither bone lesions suspicious for medullary replacement nor lytic lesions. Laboratory tests showed no monoclonal gammaglobulin peak, nor LDH and β2-microglobulin elevation. Renal function and calcemia were normal. No proteinuria was found. The κ/λ ratio was normal.

At 8 months of follow-up, the patient is asymptomatic and colonoscopy revealed no recurrence.

SEP shows an indolent clinical behavior. The 10-year overall survival rate is 70 % in SEP. Approximately 65 % of the patients had no recurrence and did not progress to MM [4] Only 15 % of the SEP seems to progress to MM compared to solitary bone plasmocytoma (SBP) with a significantly higher risk of progression to MM. Moreover, patients with SEP evolved into MM had a 100 % 5-year survival rate as compared to 33 % for SBP.

Due to the rarity and the scattered reports of SEP, determination of the prognostic factors can be difficult. Large tumor size (>5 cm) and elevated β2 microglobulin (>3.5 mg/L) seems to be adverse factors, affecting survival. Radiotherapy and serum β2 microglobulin <3.5 mg/L were favorable prognostic factors for local control, multiple myeloma-free survival, and progression-free survival in patients with SEP [3].

In our case, the tumor size was less than 5 cm and the β2 microglobulin was not elevated (<3.5 mg/L). Radiotherapy was not performed as a complete surgical resection was achieved.

For SEP in GI tract treated with primary surgery, RT will only be required in case of inadequate surgical margins. Combined therapy is also recommended if complete surgical tumor resection is unfeasible or impossible and/or lymph node areas are affected and/or the patient is high risk to progress to MM. No beneficial effect of adjuvant chemotherapy on disease control or prevention of progression to multiple myeloma has been reported by several authors [3].

Despite this tumor presents a good prognosis, a long-term follow-up is recommended as in a minority of cases, SEP may progress into the more aggressive MM.

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