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Case Report

Plasmacytoma of the Ovary

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Abstract

Extramedullary plasmacytoma is a rare plasma cell neoplasm that involves the ovary very rarely. Here, I report a case of primary ovarian plasmacytoma in a 42-year-old patient. The patient presented with abdominal distention, and workup showed a left ovarian mass (6.5 cm × 8 cm) with marked ascites. The patient had a hysterectomy with bilateral salpingo-oophorectomy. Histopathologic, immunohistochemical, bone marrow examination, and other relevant examinations established the diagnosis of primary ovarian plasmacytoma with kappa light chain restriction. No evidence of malignancy in the omentum and ascitic fluid was detected. The patient did not need postoperative chemotherapy or radiotherapy, and 6-month follow-up was uneventful. Due to the scarcity of information on these extremely rare tumors with abdominal masses, the diagnosis should be considered if complete surgical resection is achieved and no evidence of multiple myeloma is found.

Keywords: Extramedullary plasmacytoma, extramedullary plasmacytoma, ovary, plasmacytoma, primary neoplasm

INTRODUCTION

Plasmacytomas are a group of neoplasms characterized by the proliferation of mature plasma cells, typically synthesizing monoclonal immunoglobulins.[1] They exist in three clinical forms: multiple myeloma (MM), medullary plasmacytoma (MP), and extra MP (EMP). Occasionally, plasmacytomas may present as solitary lesions in the bone marrow known as solitary intra MPs. In 5%–10% of reported solitary plasmacytomas, the lesions may be found outside the bone marrow and are named solitary EMPs.[2]

Voigt reported the first case of EMP in 1938,[3] but half of the reported cases have been published in the past 8 years [Table 1],[2-14] potentially indicating greater detection or incidence. EMP is a rare primary soft-tissue plasma cell tumor commonly found in the head and neck, with 90% occurring in the upper aerodigestive tract.[12,15] EMP typically affects middle-aged persons (55–60 years; male: female

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Table 1: Published cases of ovarian plasmacytoma (chronological order)

Author (reference)	Age (years)	Symptoms	Ovarian tumor size (cm)
Bambirra *et al*., 1984[4]	44	Abdominal pain bilateral	Right 14.3×5.3×4 Left 12.3×9 × 36
Hautzer, 1984[5]	56	Abdominal mass	Left 24×31×14.3
Talerman and Path, 1987[6]	35	Abdominal mass	Unilateral 15.3×12.3×9
Cook and Boylston, 1988[7]	63	Abdominal pain	Left 12.3×10×3.7
Andze *et al*., 1993[8]	12	Pelvic mass	Left 12.3×8 × 3.8
Emery *et al*., 1999[9]	54	Abdominal swelling	Left 15.3×13×3.8
Zhong *et al*., 2012[10]	54	Abdominal pain	Right cystic ovary 12×12×10
Shakuntala *et al*., 2012[11]	35	Abdominal mass, intermittent pain	Right 14×13.5×6
Mondal *et al*., 2015[12]	47	Abdominal pain	Left 10×9.5×6
Feldman *et al*., 2015[2]	84	Weight loss, constipation, dizziness, palpitation, loss of appetite	Right 10×11.4×8.5
Tomaselli *et al*., 2016[13]	46	Abdominal mass and pain	Right 15×14×10
Cárdenas-Perilla and Urrego-Meléndez, 2020[14]	48	Abdominal pain	Bilateral 10
Present case	42	Abdominal distention	Left 18×8 × 5

ratio of 3:1). Confirmed risk factors for EMP remain unknown, but prior radiation exposure has been suggested. These patients have a higher rate of progression to MM/disseminated disease, and they require close monitoring after appropriate treatment.[12] Ovarian plasmacytomas are unique and have an unusual presentation.[9,11,13]

CASE REPORT

In January 2016, a multiparous Libyan woman aged 42 years presented at the Department of Medicine, Tripoli Central Hospital, Tripoli, Libya, with abdominal distension of 7-day duration. Her workup included ultrasound examination, which revealed marked ascites and a left ovarian mass measuring 6.5 cm × 8 cm. Blood investigations revealed CA 125 of 1602 U/ml, mild iron deficiency anemia with Hgb 10.4 g/dl Vitamin D deficiency. Anti-HIV, anti-HCV, and Anti-HbsAg antibodies were negative.

Abdominal and pelvis with intravenous contrast [Figure 1] revealed a left ovarian mass measuring 9 cm × 8.7 cm, with a cystic part and a contrast-enhanced solid part, with marked ascites in the abdomen and pelvis. She underwent a transabdominal hysterectomy and bilateral salpingo-oophorectomy. The patient did not require chemotherapy or radiotherapy postoperatively, and 6-month follow-up was uneventful.

The ovarian mass had a smooth outer surface and measured 18 cm × 8 cm × 5 cm. Serial sections of the mass showed a grayish-brown solid surface with some cystic changes. Histopathological examination shows diffuse sheets of plasma cell infiltration replacing all ovarian stroma [Figure 2]. The plasma cells are characterized by eccentrically located nuclei and bright eosinophilic cytoplasm with high mitotic figures. Many multinucleated cell forms are seen. Histopathologic examination showed no evidence of malignancy in the omentum or ascitic fluid.

Immunohistochemical analysis showed that the plasma cells were positive for CD138 and leukocyte common antigen, with kappa light chain restriction and faint focal positivity for CD79a. The results were negative for CD20, CD3, myeloperoxidase (MPO), placental alkaline phosphatase (PLAP), inhibin, CK, and CD68. The proliferative index for Ki67 was high (40%). The workup for MM was negative.

The bone marrow biopsy shows bone trabeculae surrounding normocellular marrow spaces containing trilineage hematopoietic elements admixed with scattered single plasma cells and small clusters of plasma cell (<10% of the whole cellularity). Immunohistochemical examination showed that the plasma cells were positive for CD138 with no restriction to kappa or lambda light chains (positive for both kappa and lambda) and reactive marrow with no evidence of monoclonal plasma cell infiltration.

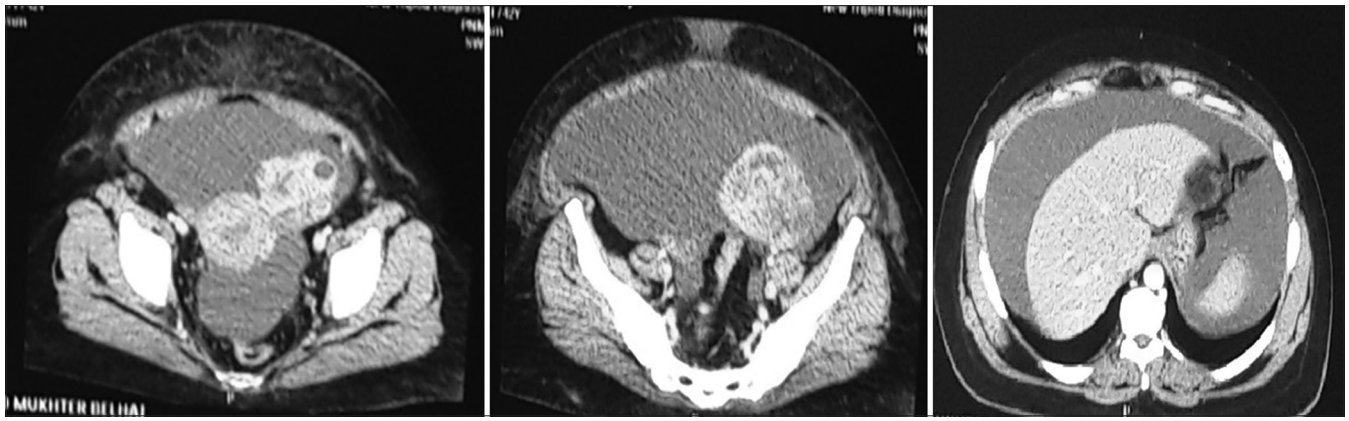

Figure 1: Computed tomography scan abdomen and pelvis. Evidence of sizeable mixed density mass measuring about 9 cm × 8.9 cm, arising from the left ovary, showing cystic par and contrast enhancement solid part. Marked ascites fluid is seen in the abdominal and pelvic cavity

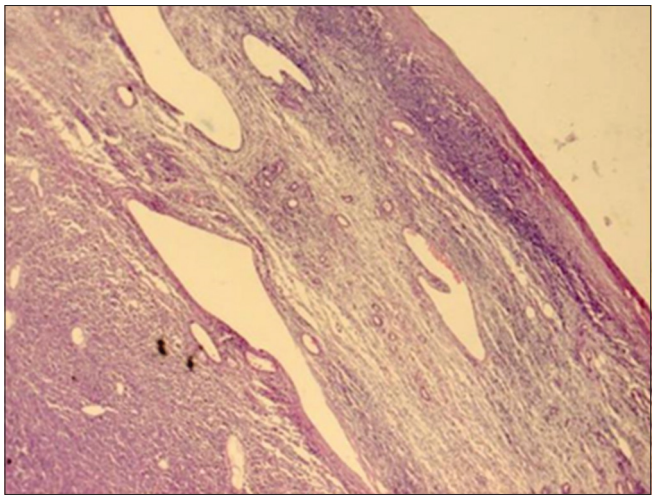

Figure 2: Histopathologic photo of varian plasmacytoma: Diffuse sheet of plasma cells infiltration replacing all ovarian stroma, the plasma cells characterized by eccentrically located nuclei, and bright eosinophilic cytoplasm. Many multinucleated cell forms are seen

In March 2016, the patient underwent a whole-body positron emission tomography/computed tomography (PET/CT) scan, which revealed no pathological F18-fluorodeoxyglucose uptake (neither focal nor diffuse), suggesting active malignancy. Postoperatively, CA 125 was 132.0 U/ml (February 10, 2016), and in October 2016, it was reduced to 20.8 U/ml (within normal limits).

DISCUSSION

Plasmacytoma develops from clonal proliferation of plasma cells identical to plasma cells of MM both cytologically and immunophenotypically. EMP is a rare primary soft-tissue plasmacytoma, representing <5% of all plasma cell tumors. These tumors generally remain localized and are more responsive to therapy.[11]

Diagnosis of a solitary EMP requires the exclusion of MM. This distinction is important since more than 60% of patients treated for a solitary plasmacytoma are cured with only local therapies. In comparison, the 5-year survival of patients with MM is about 35%. The criteria for diagnosing solitary EMP include (a) histologically confirmed solitary plasma cell lesion, (b) <10% plasma cells with no dyscrasia in bone marrow biopsy of a distant site, (c) absence of end-organ damage such as those found in MM, and (d) a full-body skeletal survey to exclude intramedullary disease. In solitary EMP, aside from the possible occurrence of monoclonal gammopathies, all laboratory values are expected to be normal. These include beta 2-immunoglobulin, blood counts, electrolytes, serum-free light chains, and serum protein electrophoresis.[2,12] Unlike MM, EMP usually does not exhibit serum M protein or Bence Jones light chains in urine. Nevertheless, EMP may show M protein in up to 25% of cases. In our case, neither M protein nor Bence Jones protein was present.[12]

Review of the literature review shows that most of the women present with a mass and pain in the abdomen [Table 1]. Ovarian plasmacytoma is usually large at the time of presentation and more likely to be on the left side.[9,12] Shakuntala *et al*. reported an ovarian plasmacytoma presented as a large (15 cm × 15 cm) right adnexal mass.[11] Cardenas-Perilla *et al*. reported the detection of bilateral ovarian plasmacytoma by PET/CT 2 years after her treatment for EMP in the left femur.[14] All reported cases were either solid

cystic or solid mass, except for the case reported by Zhong *et al*., who reported a case with intraperitoneal hemorrhage (ovarian rupture).[10] Ovarian EMP has been observed in elderly persons but only one child (12 years).[8]

CONCLUSIONS

The ovarian plasmacytoma is a rare tumor with information known about it. It is possible to be present in women with abdominal masses with or without ascites. Adjuvant therapy for ovarian plasmacytomas has not been established. If complete surgical resection is achieved and there is no evidence of MM, a strategy of observation only should be considered.

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Declaration of patient consent

The authors certify that they obtained all appropriate patient consent forms. In the form, the patient has given her consent for images and other clinical information to be reported in the journal. The patient understands that names and initials will not be published and due efforts will be made to conceal patient identity, but anonymity cannot be guaranteed.

Authors' contributions

All authors were involved in the investigation of the reported case and drafting and revising the case report.

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Nil.

Conflicts of interest

There are no conflicts of interest.

Compliance with ethical principles

No prior ethical approval is required for single case reports and small case series provided patient consent is obtained and all data are presented anonymously.

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