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Case Report

Solitary Ovarian Plasmocytoma: A Rare localization of Extramedullary Plasmocytoma. A Case Report

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Abstract:

Background: Solitary plasmacytoma is a rare malignant neoplasm of plasma cells accounting 5-10% of all plasma cell dyscrasias with extramedullary plasmocytoma in 3- 5%. Their localization in the female genital tract is quite rare, either as solitary plasmacytomas or as partof a disseminated MM. Solitary ovarian plasmocytoma is extremely rare. *Case:* A 52-year-oldwoman, presented presented postmenopausal recurring episodes of metrorrhagia with left ovarian mass. She was diagnosed with solitary ovarian plasmocytoma without systemic disease.The patient underwent complete surgery resection and a full work up to rule out multiple myeloma that objectived a small serum monoclonal protein that had resolved postoperatively. At 17 months of follow-up, the patient is still alive and doing well with no signs of recurrenceor progression to mutliple myeloma. Although rare, solitary plasmacytoma of the ovary can occur without any overt symptoms or laboratory abnormalities tests and require prompt and adequate treatment and rigorous monitoring due to their ability to relapse or progress to MM. Complete surgical resection followed by active surveillance is appropriate.

Keywords: solitary plasmocytoma; extramedullary; ovary; multiple myeloma

Introduction

Solitary plasmacytoma is a rare malignant neoplasm of plasma cells accounting 5-10% of all plasma cell dyscrasias [1, 2]. The diagnosis of solitary plasmacytoma requires a single lesion composed of monoclonal plasma cells without evidence of systemic disease, such as bone marrow involvement, evidence of systemic amyloidosis, hypercalcemia, and organ damage (ie, renal insufficiency), or multiple sites of involvement on comprehensive imaging [2]. Solitary plasmacytoma of bone (SPB) and extramedullary plasmacytoma (EMP) belong to the broader diagnosis of solitary plasmacytomas, but have distinct characteristics at presentation [3, 4]. EMP comprises 3-5% of all plasma cell tumors [5]. They're roughly one-third as common as SPB and about 80% present in the upper aerodigestive tract, with nasal cavity, paranasal sinuses, and nasopharynx being the most common sites of involvement [1, 6]. The 5-year risk of advancing to multiple myeloma (MM) is 30-50% for patients presenting with SPB but only 10-30% when found in the extramedullary soft tissue with a 10-year overall survival rate of 70% [7, 8]. EMP in the female genital tract are quite rare, either as solitary plasmacytomas or as part of a disseminated MM. There are few cases described in the literature [9-12]. We report here a case of solitary ovarian plasmocytoma.

Case and Methods

A 52-year-old multiparous woman and mother of 3 children, with past medical history of type 2 diabetes under oral treatment for 7 years, and spontaneous menopause at 49 years without hormonal therapy, presented recurring episodes of metrorrhagia for 3 years. Physical examination showed stable patient with no sign of hemodynamic instability. On vaginal and rectal examination, the uterus and cervix felt normal. A filling of bilateral vaginal sac was identified. There was tenderness during examination. Bilateral parametria were supple and rectal mucosa was free. Pelvic ultrasound revealed enlarged uterus with a heterogeneous thickning of endometrium and a heterogeneous left latero-uterine mass. Biological exams showed, Hb level at 14 g/dl, with normal blood cell count. Serum cancer antigen (CA-125) < 4 U/ml and lactic dehydrogenase

(LDH) was high at 489 U/l. Beta human chorionic gonadotrophin (β-hcg), carcinoembryonic antigen (CEA), and alpha fetoprotein levels (AFP) were normal. Serum anti-HIV, and anti-HbSAg levels were also normal. Before surgery, diagnosis of ovarian granulosa tumor was suspected. The exploratory laparotomy revealed an enlarged and dilated left ovary measuring 4.5*3.5*3.0 cm and joined by the uterine cervix (Figure. 1).

Figure 1: *Macroscopic examination showed enlarged and dilated left ovary measuring 4.5*3.5*3.0 cm*.

Endometrium was polypoid with hemorrhagic stigma and an interstitiel myoma measuring 0.1*0.8 cm in favor of uterine leiomyoma. Histological study of left ovary showed a well circumscribed, encapsulated and white tumor measuring 4.0*3.2 cm. Pathological examination revealed tumor proliferation with insular and trabecular architecture with monotonous mature plasma cells with minimal cytonuclear atypia effacing the ovary (Figure. 2).

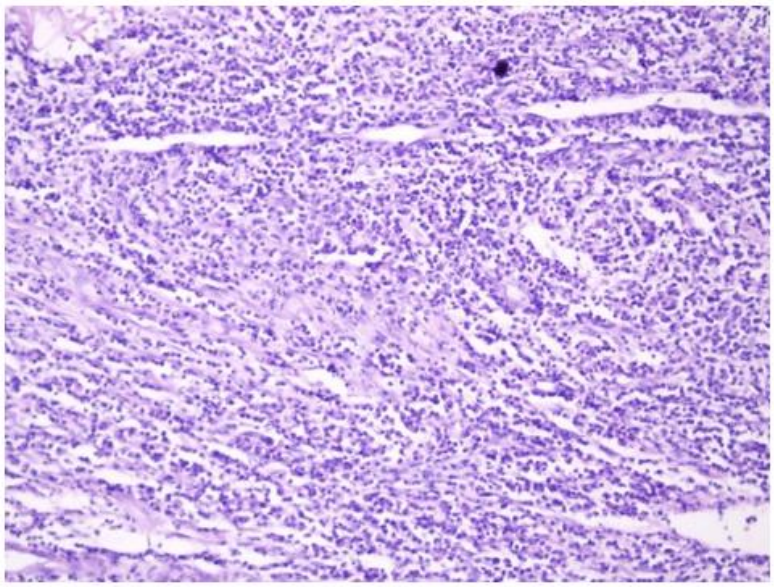

Figure 2: *Low power view is showing plasma cells arranged in sheets (H & E)*

Immunohistochemical staining performed on the ovarian specimen showed positive expression of epithelial membrane antigen (EMA) (Fig. 3a), CD138 (Fig. 3b), with a lambda light chain restriction (Fig. 3c). Tumor cells were negative to cytokeratin, anti-smooth muscle, inhibine and H-Caldesmone. Antigen Ki-67 level was at 10%. Postoperatively, the patient underwent a full work-up to rule out MM, serum protein electrophoresis revealed a small monoclonal protein of 0.4 g/dl present in the gamma region. Urine immunoprotein tests were negative. Kidney function tests and calcemia were normal. Beta-2 microglobulin level was at 3.11 mg/l. Bone marrow examinations revealed 3% of plasma cells. X-

ray examinations of the skeleton were normal. The diagnosis of solitary ovarian plasmocytoma was retained and the patient didn't receive adjuvant treatment because surgical resection was complete. The patient is alive and doing well but the clinical and biological monitoring is irregular given the COVID pandemic and lack of access to specialized care establishment for tests and consultation. The current follow-up is 17 months.

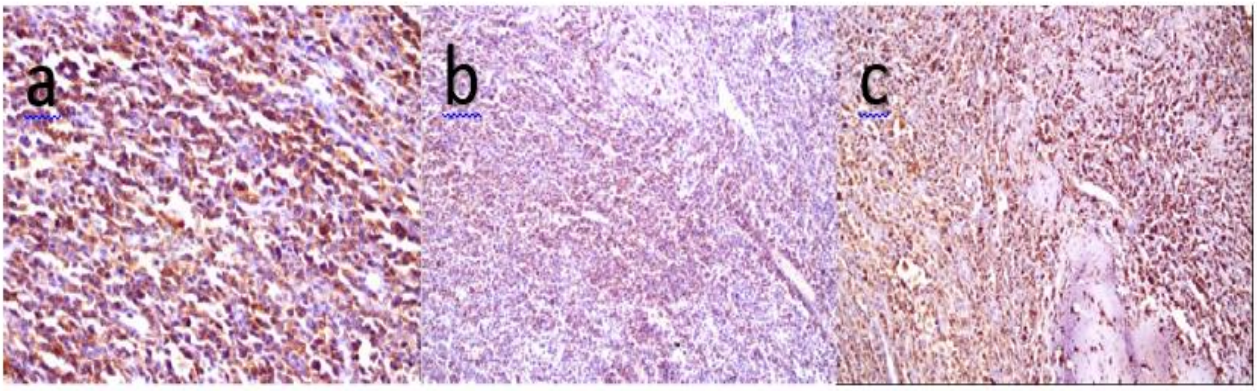

Figure 3: *Immunohistochemical staining showed (a) : Epithelial Membran Antigen (EMA) positive expression. (b) : CD138 positive. (c) : Lambda light chain positive.*

Discussion

EMP is a rare entity that comprise 3-5% of all plasma cells neoplasms [5] and result from the proliferation of monoclonal plasma cells with < 10% in bone marrow examinations with no evidence of systemic disease and with normal skeletal survey [2], aside from possibly monoclonal gammopathy [6]. EMP are present in 80% in head and neck [1,6], and typically affects the middle-aged persons (median 55- 60 years) with a predominance of female sex. Risk factors for EMP remain unknown; however, prior radiation exposure has been suggested. These patients have a higher rate of progression to MM, and they require close monitoring after appropriate treatment [13]. EMP in the female genital tract are extremely rare, there are few cases described [9- 12] and solitary ovarian plasmocytoma remains exceptional, Voegt et al. reported the first case in 1938 [14], and since then rare cases have been published as single observation or small series [12]. These tumors are usually large at the time of presentation and their size is greater than 12 cm with abdominal pain and/or mass [11]. Our patient presented dilated ovary mesearing 45*35*30 mm with small tumor measuring 40*32 mm, and without symptoms clinically. Metrorrhagia could be explained by the leiomyoma rather than the ovarian mass. Preoperatively, ovarian granulosa tumor was suspected in our patient because of the frequency, age, postmenopause and localization, therefore macroscopic examination and immunohistochemical staining remain essential to confirm or exclude diagnosis. Ovarian plasmocytomas are more likely involving the left ovary and usually without evidence of disseminated disease [11, 12]. In this case, the patient presented left ovarian plasmocytoma with positive CD138, and lambda light chain restriction. The work up to rule out MM showed solitary plasmocytoma with a small monoclonal protein at 0.4 g/dl. Adjuvant radiotherapy is suggested where full resection of the lesion is impossible or cannot be confirmed [6]. Our patient didn't receive adjuvant radiotherapy due to the complete surgical resection. For solitary EMP, active surveillance after complete surgical resection is recommended. The rate of progression and survival data are very sparse and are collected from case reports. Median survival of patients with EMP is 4- 10 years. Local recurrence and progression to MM may occur, justifying prolonged and rigorous follow up [11, 15]. At follow-up, only 9% of patients progress with resolved serum monoclonal protein versus 71% of progression for patients with persistent serum monoclonal protein after treatment [16]. Our patient had normalized serum monoclonal protein after surgery and she's doing well after 17 months of follow up.

Conclusion:

Although rare, solitary plasmacytoma of the ovary can occur without any overt symptoms or laboratory abnormalities tests. These represent an entity of plasma cell dyscrasias and thus require prompt and adequate treatment and rigorous monitoring due to their ability to relapse or progress to MM. Complete surgical resection followed by active surveillance is appropriate. Due to the disparity in management and prognosis, it is important to distinguish solitary ovarian plasmocytoma from ovarian involvement in the context of MM.

Declaration of Competing Interest

None

Author contributions

SF conceived and designed the manuscript. SF wrote the manuscript. SF and KS reviewed and edited the manuscript and provided patient management. AM and AT contributed to patient management and review of the manuscript. All authors read and approved the final manuscript.

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Patient Consent

Written, informed consent for publication of this report has been obtained from the patient. All identifying information has been removed to preserve confidentiality. We would like to thank the patient and his family for their contribution to this article.

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