

Case Report

Solitary ovarian plasmacytoma. A case report and review of literature

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1. Background

Plasma cell dyscrasias are a group of neoplasms characterized by the proliferation of mature plasma cells typically synthesizing monoclonal immunoglobulins. These neoplasms generally present with many centers of proliferation, as in multiple myeloma (MM). Occasionally, they may present as solitary lesions. These are commonly formed in the bone marrow and are known as solitary intramedullary plasmacytomas. In 5–10% of reported solitary plasmacytomas (Susnerwala et al., 1997; Dagan et al., 2009), these lesions may be found outside the bone marrow and are named solitary extramedullary plasmacytomas (EMP).

Solitary extramedullary plasmacytomas (EMP) most frequently arise in the upper aerodigestive tract (Gerry and Lentsch, 2013). Other common sites include gastrointestinal and urogenital tracts, lymph nodes, thyroid, testes, and breast (Wiltshaw, 1976; Alexiou et al., 1999). We report just the second published case of solitary ovarian extramedullary plasmacytoma together with a review of the literature (Voegt, 1938; Bambirra et al., 1982; Hautzer, 1984; Cook and Boylston, 1988; Emery et al., 1999; Shakuntala et al., 2013) with regard to presentation, diagnostic work up and management.

2. Case report

An 84 year old Hispanic woman G3 P3 presented to the emergency department (ED) with a chief complaint of dizziness, palpitations, constipation, decreased appetite, and early satiety as well as an unexplained 10–15 lb weight loss over the previous several months. A large abdominal mass was palpated on physical exam and ultrasound done in the ED revealed an 11 × 7 cm nonvascular well circumscribed mass in the right adnexa. CT scan showed a large heterogeneous partially calcified right adnexal mass measuring 10 × 11.4 × 8.5 cm. There was no pelvic or retroperitoneal lymphadenopathy on imaging.

Carcinoembryonic antigen (CEA) was 0.8 ng/mL and serum cancer antigen (CA-125) was <4 U/mL. LDH at the time of diagnosis was slightly elevated at 227 IU/L.

After obtaining informed consent, the patient underwent a bilateral salpingo-oophorectomy with right pelvic and para-aortic lymphadenectomy. Intraoperatively an enlarged right adnexa measuring 12 cm was found adherent to the abdominal wall peritoneum laterally. The mass also extended into the retroperitoneal space, adhering to the external iliac artery and vein. There were adhesions to the cecum and rectosigmoid mesentery as well. The uterus and left adnexa appeared normal. A single slightly enlarged obturator fossa lymph node, as well as several enlarged nodes along the high common iliac vessel were dissected during surgery.

The gross anatomy of the resected right ovary is shown in Fig. 1. Pathological examination of the right adnexa revealed sheets of monotonous mature appearing plasma cells effacing the ovary [Fig. 2a–b]. Specimens submitted for peritoneal cytology were negative. The enlarged lymph nodes removed during surgery were shown to have an exuberant plasmacytic infiltrate. However, in-situ hybridization for kappa and lambda highlighted them as polyclonal plasma cells. Therefore, the immune architecture and morphology of the lymph nodes did not support an overt plasma cell or B-lymphoproliferative disorder and was possibly of a reactive nature. Immunohistochemical staining performed on the ovarian specimen demonstrated that the tumor cells were positive for CD138 with in-situ hybridization (ISH) showing a kappa light chain restriction. There were negative for cytokeratin, CD20, CD3, CD5, BCL-1, and PAX-5 [Fig. 3a–c].

Postoperatively, the patient underwent a full work-up to rule out multiple myeloma. This included beta-2 microglobulin, LDH (159 IU/L), and iron studies that were all within normal limits. Serum protein electrophoresis revealed IgG, IgM, and IgA all within normal limits (WNL), with a small IgG kappa monoclonal protein of 0.3 g/dL present

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Fig. 1. Macroscopic examination shows a 14 cm, 597 g ovary with smooth capsule that is focally calcified; cut surface shows multiple cysts with myxoid, rubbery and hemorrhagic septa and contents.

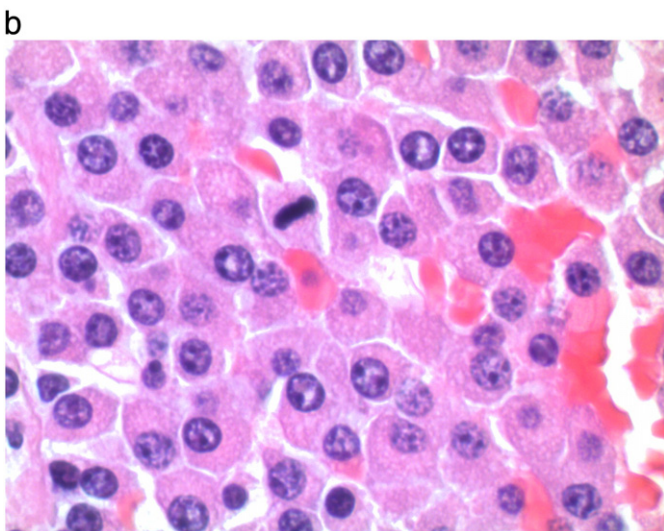
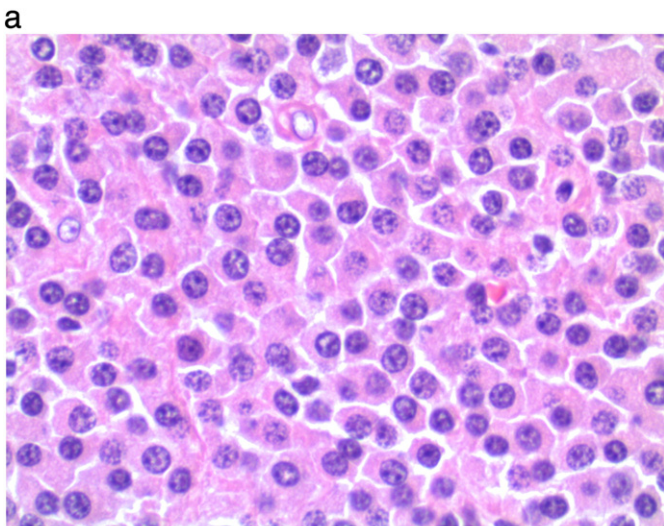
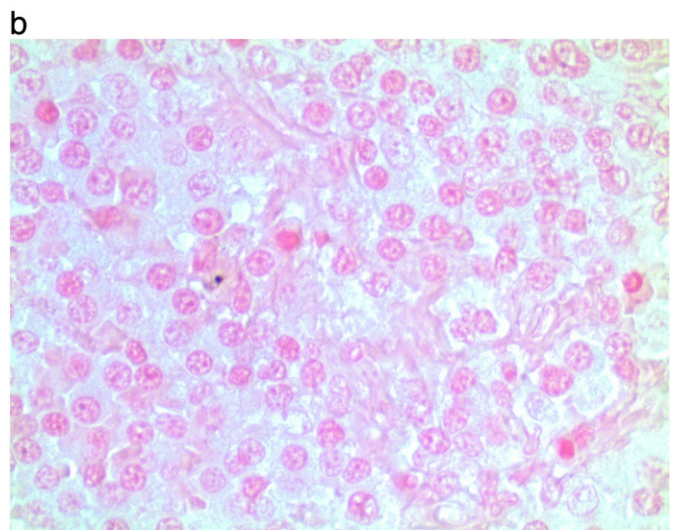
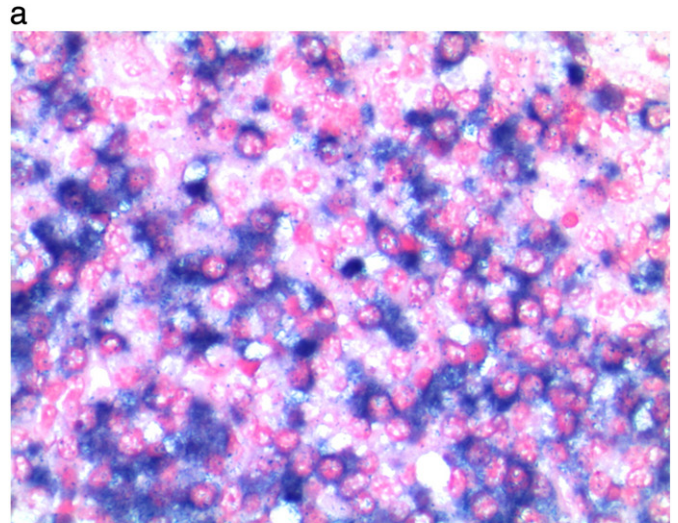


Fig. 2. Microscopic H&E with (a) medium high ($\times 20$) and (b) high power ($\times 40$) shows mostly mature plasma cells with eccentric clock-faced nuclei and infrequent mitoses; no admixed lymphocytes.

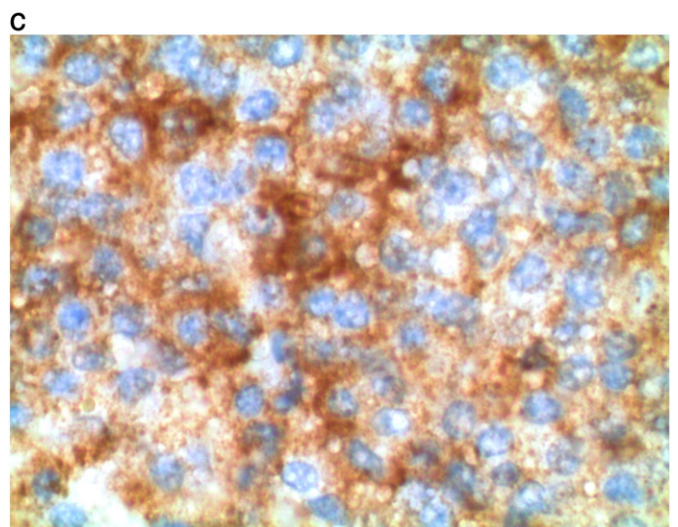


Fig. 3. Special stains using in-situ hybridization (ISH) showing Kappa positive (in blue) (a), Lambda negative (b) and immunohistochemical stain positive for CD138 (in brown) (c).

in the slow gamma region. A full skeletal bone survey, urine free light chain analysis and bone marrow examination were normal. The patient is doing well and currently undergoing active surveillance with her gynecologic oncologist and hematologist.

3. Discussion

Solitary plasmacytomas are an uncommon collection of proliferating monoclonal plasma cells. As in multiple myeloma, the cells are synthesized in the lymph nodes, migrate to the vascular system, and as in the case of MM, eventually settle in the bone marrow. Rarely, for as yet unknown reasons, these cells may collect in soft tissue and do not migrate to the bone marrow. One explanation centers on certain adhesion molecules which may dictate whether the malignant cells gather in the bone marrow or remain in soft tissue. What is clear is that these cells rarely accumulate in the ovary.

EMP shares a common cell type, morphology and pathology with MM, but does not exhibit the systemic properties of its more serious counterpart. Diagnosis of a solitary EMP requires exclusion of multiple myeloma. This distinction is important, since more than 60% of patients who are treated for a solitary plasmacytoma are cured with only local therapies, while the 5-year survival for patients with MM is around 35% (Kumar et al., 2008). Criteria to diagnose solitary EMP include a histologically confirmed solitary plasma cell lesion, less than 5% plasma cells found on bone marrow biopsy of a distant site, absence of end organ damage such as those found in MM, and a full body skeletal survey to exclude intramedullary disease (Dimopoulos et al., 1999). In solitary EMP, all laboratory values are expected to be normal aside from possibly monoclonal gammopathies. These include Beta2 immunoglobulin, blood counts, electrolytes, serum free light chains and serum protein electrophoresis (Alexiou et al., 1999).

The first case of ovarian EMP was described by Voegt (1938), and since then eight other cases have been recounted (Bambirra et al., 1982; Hautzer, 1984; Cook and Boylston, 1988; Emery et al., 1999; Shakuntala et al., 2013). However, when applying the strict diagnostic criteria for solitary ovarian EMP, only one case report fulfilled these required diagnostic tests (Emery et al., 1999). Our case represents the second case of solitary ovarian EMP. Most of the previously reported cases of ovarian EMP were manifestations of fully developed multiple myeloma (Voegt, 1938; Bambirra et al., 1982; Shakuntala et al., 2013), or were lacking the required diagnostic tests e.g. bone marrow biopsy (9) to label the case as a true “solitary” ovarian EMP.

For solitary EMP, Alexiou et al. (1999) recommended active surveillance after complete surgical resection. Adjuvant radiotherapy is suggested where full resection of the lesion is impossible or cannot be confirmed.

Progression to multiple myeloma is the most feared complication of solitary ovarian EMP. Although the 5-year rate of progression to MM in solitary intramedullary plasmacytoma is 30–50%, only 10–35% of patients with EMP develop MM (Dagan et al., 2009; Alexiou et al., 1999). None of the previous case reports have elucidated the criteria for active surveillance. National Comprehensive Cancer Network

(NCCN) guidelines (Anderson et al., 2011) recommend regular complete blood picture, serum chemistries (LDH, albumin, calcium, beta 2 microglobulin), serum immunoglobulins, serum and urine free light chain assays and imaging as clinically indicated.

4. Conclusion

Although rare, solitary plasmacytomas of the ovary can occur without any overt symptoms or laboratory irregularities. These represent a subset of plasma cell dyscrasias and thus require prompt treatment and surveillance due to their ability to progress to frank multiple myeloma. Surgical resection followed by active surveillance is appropriate. Due to the drastic differences in management and prognosis, it is important to distinguish solitary ovarian EMP from ovarian involvement in the context of MM. This can be achieved with the use of strict diagnostic criteria.

Conflict of interest statement

The authors declare that there is no conflict of interest.

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