A case of extramedullary solitary plasmacytoma arising at the uterine cervix

Ningxia Sun[1*], Lianghe Wang[2*], Wen Li[1]

[1]Department of Obstetrics and Gynecology, [2]Department of Pathology, Shanghai Changzheng Hospital (China)

Summary

Extramedullary plasmacytomas are localized plasma cell neoplasms that arise in tissues other than bone and bone marrow. Primary plasmacytomas of the female genital tract are extremely rare and present a substantial diagnostic challenge. We report a case of a 38-year-old woman who presented with an endocervical polypoid. Surgical removal of the polyp was carried out. The final pathological report revealed primary plasmacytoma of the uterine cervix. The diagnosis was further facilitated by the use of immunohistochemistry and clonal immunoglobulin heavy-chain gene rearrangement. We performed a simple hysterectomy by laparoscopy on the patient and kept a close follow-up. She has remained well for more than eight years. The clinical characteristics and histopathologic findings of plasmacytoma of the uterine cervix are discussed.

Key words: Cervical plasmacytoma; Primary plasmacytoma; Uterine cervix.

Introduction

Plasmacytomas are localized, usually focal plasma cell neoplasms that occur in visceral structures, soft tissue, or bone [1]. Primary plasmacytomas of the uterine cervix are extremely rare and present a differential diagnostic challenge. To our knowledge, only nine cases have been reported in the published literature to date [1-9]. We report a case of a 38-year-old woman who presented with an endocervical polypoid. The diagnosis of plasmacytoma was facilitated by the use of immunohistochemistry and molecular clonality studies. After performing a simple hysterectomy by laparoscopy, the patient has remained well for more than eight years of follow-up.

Case Report

A 38-year-old woman presented for a routine health examination in September 2002 and a uterine cervical polypoid was found. She was referred to the Department of Gynecology in our hospital in October 2002. Physical examination revealed a 1 cm, pink, polypoid lesion extruding from her cervix showing no parametrium or vagina abnormality. Surgical removal of the polyp was done in the outpatient clinic. Unfortunately the final pathological report revealed primary plasmacytoma of the uterine cervix. To make the differential diagnosis of multiple myeloma, a series of laboratory tests were carried out. Complete blood count and urinalysis were normal. Abdominal ultrasound (US) and chest radiograph were unremarkable. Serum immunoglobulin levels were normal: IgG 11.9 g/l (normal range 8-15 g/l), IgA 1.35 g/l (normal range 0.85-3 g/l), IgM 0.56 g/l (normal range 0.5-2.5 g/l), κ 2.71 g/l (normal range 1.72-3.83 g/l), κ 1.46 g/l (normal range 0.81-1.92g/l), κ/λ 1.86 (normal range 1.47-2.95 g/l). Serum and urine immunofixative electrophoresis and bone marrow biopsy showed no abnormality.

After reviewing the case with hematologists, laparoscopic-assisted vaginal hysterectomy was carried out in November 2002. No residual lesions were found in the hysterectomy specimen. The patient recovered soon after the surgery. There has been no clinical evidence of recurrence for more than eight years of postoperative follow-up.

Pathologic findings

Microscopically, the polypectomy specimen was filled with a dense aggregate of mature and immature plasma cells. Plasma cells were present with a morphologic spectrum ranging from mature forms to highly atypical cells with large nuclei, hyperchromatic clumped chromatin, and prominent nucleoli (Figure 1). In poorly differentiated areas, the tumor showed obvious plasmablast differentiation characteristics. Tumor giant cells could be found with brisk mitotic activity and atypical mitotic figures. Immunohistochemical stain revealed CD138 (+), CD38 (+), CD19 (−) (Figure 2), CD56 (−), CD20 (−), λ-light chain (+), κ-light chain (−), CD3 (−), ALK (−), cytokeratin (−), CD10 (−), estrogen receptor (−) and progesterone receptor (−). The monoclonal population with an aberrant phenotype, (CD38 +/CD19−/ CD56−) distinguished the plasmacytoma from the plasma cells with a normal immunophenotype (CD38+, CD19+, CD56−) [10]. Genetically, IgH gene rearrangement was found by using FR3A primer determination.

The subsequently obtained hysterectomy specimen weighed 75 g. It revealed chronic cervicitis in the cervix, and there was also chronic endometritis and multiple myomata in the uterus. We did not find plasma cells in the endocervix nor in the endometrium.

Discussion

Extramedullary plasmacytoma (EMP) is a rare category of plasma cell tumors. It may originate at any site, but occurs predominantly in the upper respiratory tract. The female genital tract is rarely the primary site for hematologic malignancies. Plasmacytomas of the uterine

*The two authors contributed equally to this work.

Revised manuscript accepted for publication December 30, 2011

cervix are extremely rare [11]. Our review of the literature revealed only nine cases localized to the uterine cervix since 1949 [1-9]. Based on this literature review, the median age at diagnosis was 39.4 years. Clinical symptoms, which are often nonspecific and mimic other disorders, are usually vaginal bleeding or vaginal discharge related to intercourse. One patient presented with pelvic pain. Physical examination findings are frequently inflamed-appearing cervicitis or mass lesions. The diagnosis of primary plasmacytoma of the uterine cervix usually requires biopsy confirmation. Due to the paucity of published data, this rare tumor poses a diagnostic challenge both for clinicians and pathologists. For pathologists, the possibility of a neoplastic process should be considered when highly atypical plasma cells are seen on a Pap test or cervical biopsy. The lesion is histopathologically characterized by infiltrates of plasma cells of diverse maturity and by their monoclonal immunoglobulin products [12]. Immunohistochemical stain and also gene rearrangement analysis should be suggested. The differential diagnosis of multiple myeloma should be excluded by a series laboratory tests such as normal serum immunofixative electrophoresis and bone marrow biopsy. Pathologically, differentiation from plasma cell-rich cervicitis can be excluded by the utility of immunophenotypic and molecular techniques [1].

For the treatment, our review of the literature revealed that one patient received only conization of the uterine cervix, three had a hysterectomy, two received local radiotherapy, two received a surgical excision of the tumor followed with irradiation and chemotherapy, and one patient was treated with three-dimensional conformal radiotherapy after conization.

In most reports on EMP, nearly all patients successfully achieve local control of 80% to 100%. The ten-year disease-free status and overall survival ranges from 50% to 80% [13]. As for plasmacytoma of the uterine cervix, our review of the literature revealed that the clinical follow-up of these patients ranged from three months to three or more years, and local recurrence or persistent disease was documented in three patients. Because of the rarity of this disease, there is no known guideline for the treatment and prognosis. Our patient underwent a simple hysterectomy by laparoscopy. As there was no residual lesion left in the hysterectomy specimen, we did not perform any other medical therapy. The patient recovered very soon and had no complications. However we are continuing a close follow-up every year. The patient has remained well for more than eight years. This is the only reported patient who has had such a long clinical follow-up to now. Thus we think simple hysterectomy may be a good and safe treatment for primary plasmacytoma of the uterine cervix.

Nonetheless, more cases and experience are required to evaluate the optimal treatment and prognosis of this rare disease. Since about 30%-50% of patients will develop disease progression to myeloma [14], we should pay attention to the long term follow-up.

References

[1] Fischer E.G., Bocklage T.J., Rabinowitz I.: "Primary plasmacytoma arising in an endocervical polyp with detection of neoplastic cells on papanicolaou test. A case report and review of the literature". *Arch. Pathol. Lab. Med.,* 2003, *127,* 28.
[2] Johansen B., Ahlbom G., Ostergard B.: "Extramedullary solitary plasmacytoma at the uterine cervix as a cause of postcoital bleeding". *Acta Obstet. Gynecol. Scand.,* 1989, *68*, 279.
[3] Adachi K., Murata N., Fujikawa H.: "A case of primary extramedullary plasmacytoma in the posterior cervical portion". *Gan No Rinsho.,* 1987, *33*, 1281.
[4] Andersen P.E.: "Extramedullary plasmacytomas". *Acta Radiol.,* 1949, *32*, 365.
[5] Barkai L., Keresztury S.: "Extramedullary plasmacytoma of the portio uteri". *Zentralbl. Gynakol*., 1967, *89,* 1728.
[6] Weissbach L., Schopen R.D.: "Solitary plasmacytoma of the cervix". *Geburtshilfe Frauenheilkd,* 1968, *28*, 960.
[7] Neuberg R.: "Immunosuppression and plasmacytoma of the cervix". *J. Obstet. Gynaecol*., 1974, *81*, 165.
[8] Huang C.C., Liu M.T., Pi C.P.: "Primary plasmacytoma of the uterine cervix treated with three-dimensional conformal radiotherapy". *Singapore Med. J*., 2008, *49*, 361.
[9] Schor A.P., Moraes M.P., Bisson F.W.: "Primary plasmacytoma of the cervix in a 21-year-old female patient". *Int. J. Gynecol. Pathol.,* 2010, *29*, 290.
[10] Van Camp B., Durie B.G., Spier C.: "Plasma cells in multiple myeloma express a natural killer cell-associated antigen: CD56 (NKH-1; Leu-19)". *Blood,* 1990, *76,* 377.
[11] Hanley K.Z., Tadros T.S., Briones A.J.: "Hematologic malignancies of the female genital tract diagnosed on liquid-based Pap test: Cytomorphologic features and review of differential diagnoses". *Diagn. Cytopathol.,* 2009, *37,* 61.
[12] Strojan P., Soba E., Lamovec J.: "Extramedullary plasmacytoma: clinical and histopathologic study". *Int. J. Radiat. Oncol. Biol. Phys*., 2002*, 53,* 692.
[13] Chao M.W., Gibbs P., Wirth A.: "Radiotherapy in the management of solitary extramedullary plasmacytoma". *Intern. Med. J.,* 2005, *35*, 211.
[14] Weber D.M.: "Solitary bone and extramedullary plasmacytoma". *Hematology Am. Soc. Hematol. Educ. Program*, 2005, 373.

Address reprint requests to:
W. LI, Ph.D.
Fenyang road 415#
200003 Shanghai City (P.R. China)
e-mail: liwenchzh@126.com