Available online at www.medicinescince.org

Case Report

Medicine Science 2016;5(4):1021-3

Medicine Science International Medical Journal

Primary pure ependymoma: a rare entity of the ovary

Huseyin Aydogmus[1], Serenat Eris Yalcin[2], Serpil Aydogmus[1], Servet Gencdal[1], Aylin Orgen Calli[1], Sefa Kelekci[1]

[1]İzmir Katip Çelebi University Ataturk Training and Research Hospital, İzmir, Turkey
[2]Suleyman Demirel University, İsparta, Turkey

Received 23 March 2016; Accepted 22 April 2016
Available online 6 May 2016 with doi: 10.5455/medscience.2016.05.8469

Abstract
Ependymomas are an uncommon glial tumors that usually arise from the central nervous system but they can occasionally occur in various extra-axial regions. Pure ovarian ependymoma, classified as differentiated glioma, was first described by Kleinman et al in 1984 and only a few cases have been reported since then. A 48 year-old multiparous woman with the diagnosis of left adnexal mass underwent surgery. Macroscopic evaluation of the left ovary showed 11x8 cm sized smooth-walled mass. Microscopic examination revealed small to medium sized cells with hyperchromatic, round to oval nuclei and scanty cytoplasm, perivascular pseudorosettes. Immunohistochemical staining for GFAP, vimentin, estrogen and progesterone receptors were positive. Based on these histopathologic and immunohistochemical features, the tumor was diagnosed as an ovarian ependymoma. As they seem rare, there is no verified treatment modality for ovarian ependymomas. , it is important to report these rare tumors, to shed light to the management and follow-up of them. To establish standard treatment modalities for neuroectodermal tumors of the ovary, it is essential to discuss all cases of ovarian epenymoma in the literature.

Keywords: Ependymoma, ovarian, immunohistochemistry

Introduction

Ependymomas are an infrequent glial tumors that usually originate from the central nervous system, but they can occasionally occur in various extra-axial regions [1,2]. Pure ovarian ependymoma, classified as differentiated glioma, was first described by Kleinman et al in 1984 [3] and only a few cases have been reported since then. As they seem rare, there is no verified treatment modality for ovarian ependymomas. So it is important to report the management and outcome of this tumor to contribute to the literature.

Case Report

A 48 year-old multiparous woman was referred to our hospital with the diagnosis of left adnexal mass for further evaluation and treatment. On examination, the external genitalia and uterine cervix were normal, but the fornices of the vagina were full. The uterus didn't feel enlarged but was pushed to the front by the mass. It was difficult to specify the origin of the tumor.

Her pelvic ultrasound revealed a large, complex, and predominantly cystic mass approximately 89x80 mm in size, that couldn't be delineated whether it arise from the uterus or the left ovary. The right ovary was normal. A CT scan of the abdomen and pelvis showed 10x9 cm sized, septated pelvic mass in retrouterine region thought to arise from left ovary (Figure 1). No other findings were revealed. Her tumor markers were within normal limits. Laparatomy exposed torsioned cystic mass arising from left ovary. She underwent total abdominal hysterectomy and bilateral salpingo-oophorectomy.

Macroscopic evaluation of the left ovary showed 11x8 cm sized, capsule integrity preserved, smooth-walled mass containing serous fluid. Frozen section of the mass revealed monodermal mature teratoma.

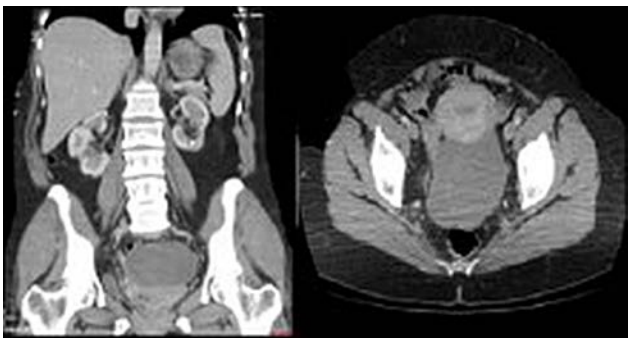

Figure 1. A CT scan of the abdomen and pelvis showing pelvic mass thought to arise from left ovary

Microscopic examination revealed small to medium sized cells with hyperchromatic, round to oval nuclei and scanty cytoplasm (Figure 2). Perivascular pseudorosettes were also observed (Figure 3). Immunohistochemical staining for GFAP, vimentin, estrogen and progesterone receptors were positive (Figure 4). Based on these histopathologic and immunohistochemical features, the tumor was diagnosed as an ovarian ependymoma. Her stage was defined as stage IA and she did not receive additional treatment after surgery. No recurrence was observed in a 2-year follow-up.

***Corresponding Author:** Irem Pembegul Yigit, Department of Nephrology, Malatya Government Hospital, Malatya, Turkey
E-mail: pembegulmd@yahoo.com

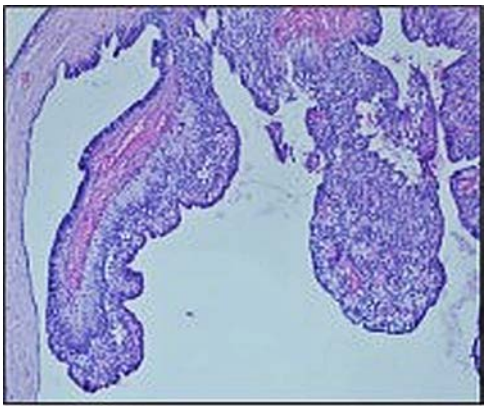

Figure 2. Histopathology of ovarian ependymoma (H&Ex100)

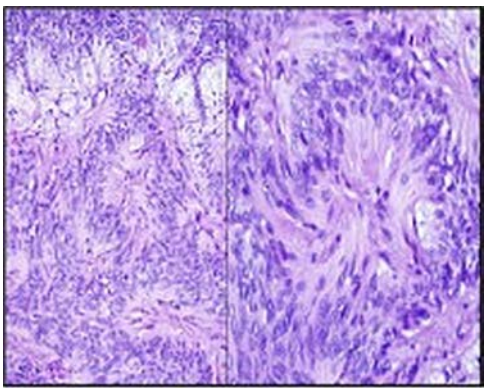

Figure 3. Typical ependymal rosette: a gland-like space lined by small to medium sized round or oval cells with fibrillary processes towards the lumen (H&Ex400)

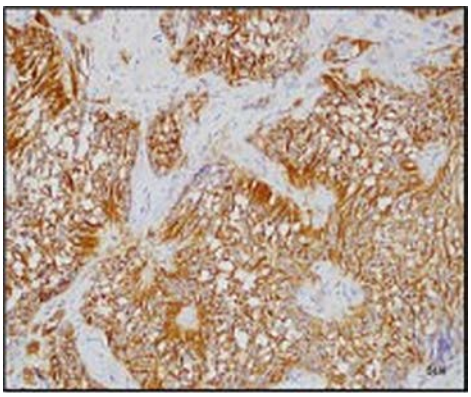

Figure 4. Immunohistochemistry showed positivity for GFAP in the cytoplasm of the cells of the ependymal rosettes (x200)

Discussion

Different versions of neuroectodermal tumors have been described in the ovary such as ependymomas, primitive or anaplastic neuroectodermal tumors and astrocytomas. They are microscopically identical to their analogues in the nervous system and can be subdivided into three groups: 1) well-differentiated, 2) anaplastic and 3) poorly-differentiated. The group of well-differentiated tumors include ependymomas and astrocytomas [4]. Anaplastic and primitive tumors predispose to occur in younger patients than well-differentiated tumors [4]. Diagnosed women are in their third and fourth decade of life mostly as in our patient. Reported cases of ovarian ependymomas in the literature have been shown in Table 1.

Table 1. Reported cases of ovarian ependymomas in the literature

Reference	Age	Size and Localization	Stage	Treatment	Chemotherapy /Radiotherapy	Recurrence during follow-up
Carr et al. [7]	25	20x20cm right, 8x16 cm left; Bilateral	Stage IV	Cytoreductive surgery	CT and RT	No
Madabhavi et al. [1]	67	98x107 mm right, 110x94 mm left; Bilateral	Stage III	Cytoreductive surgery	CT	No
Erdoğan et al. [8]	76	14x14 cm left	Stage II	Cytoreductive surgery	Not Taken	?
Stolnicu et al. [2]	32	15cm right	Stage IIIB	LSO OT pelvic node sampling	CT	Yes (8 months later cytoreductive surgery+ CT) Alive
	22	14 cm right	Stage IIIA	RSO OT appendectomy	CT	Yes (1 year later tumor resection+CT) Alive with recurrence
	35	5-7cm Bilateral	Stage IA	TAH BSO Lymphadenectomy	CT	No (Stage III colonic carcinoma diagnosed after 14 years-died)
Garcia Barriola et al. [6]	30	20cm right, 11x7 cm left; Bilateral	Stage III	TAH LSO OT (RSO was done before referral)	CT	No
Hirahara et al. [5]	19	?, right	Stage I	RSO	Not Taken	Yes (8 cm left sided Stage III tumor- Cytoreductive surgery+ CT+ RT+ hyperthermotherapy) Died 9 years after initial diagnosis
Kleinman et al. [3]	25	?,left	Stage IA	TAH BSO	Not Taken	No
	16	16cm left	Stage IA	TAH BSO	Not Taken	?
	36	16 cm right	Stage IIA	TAH BSO	Not Taken	No
	35	10x10 cm left	Stage III	TAH LSO	CT	Yes
	30	8 cm right	Stage III	TAH BSO OT	CT and RT	Yes
Guerrieri and Jarlsfelt [10]	68	18x11 cm left	Stage IA	TAH BSO OT	Not Taken	No

TAH: Total abdominal hysterectomy; BSO: Bilateral salpingo-oophorectomy; LSO/RSO: Left/Right salpingo-oophorectomy; OT: Omentectomy; CT: Chemotherapy; RT: Radiotherapy.

Microscopically ovarian ependymomas can be found in cellular, papillary, myxopapillary or mixed patterns [2-6]. Perivascular pseudorosettes can be seen as well as ependymal rosettes. Psammoma bodies can be found. This variability expedites difficulties for differential diagnosis with various primary and metastatic ovarian tumors. They can enclose large gland-like areas and tubules appeared like endometrioid carcinoma of the ovary. The presence of papillary structures may pose papillary serous carcinoma of the ovary. Ependymal rosettes can be similar to Call-Exner bodies of granulosa cell tumors [4-6]. Gland-like areas lined by cells with fibrillary cytoplasmic processes, perivascular pseudorosettes and positive satining for GFAP confirm the diagnosis of ovarian ependymoma [7,8]. Ependymomas can also stain positively for S-100, epithelial membrane antigen (EMA), neuron specific enolase (NSE), cytokeratin, CEA, vimentin, estrogen and progesterone receptors [9,10]. Our case showed positivity for GFAP, vimentin, estrogen and progesterone receptors as in line with previous studies.

There are no standard guidelines for the management of ovarian ependymomas because of the rarity of these tumors. In previous reports most patients with early stage received operation only [3,5,8] while patients with advanced stage administrated with adjuvan radiation and/or chemotherapy after cytoreductive surgery [1,3,6,7]. Patients with early stage have longer overall survival with less recurrence rates, so stage seems to be the most important prognostic parameter of survival. The expression of estrogen and progestin receptors may suggest that hormonal responsiveness of this tumor can be used as a treatment modality. An aromatase inhibitor can be initiated for these tumors [9,10].

In conclusion, it is important to report these rare tumors, to shed light to the management and follow-up of them. To establish standard treatment modalities for neuroectodermal tumors of the ovary, it is essential to discuss all cases of ovarian epenymoma in the literature.

References

1. Madabhavi I, Patel A, Choudhary M, Anand A. Paraneoplastic internal jugular vein thrombosis leading to diagnosis of bilateral ovarian ependymoma. Case Rep Obstet Gynecol. 2014;2014:324509.
2. Stolnicu S, Furtado A, Sanches A, Nicolae A, Preda O, Hincu M, Nogales FF. Ovarian ependymomas of extra-axial type or central immunophenotypes. Hum Pathol. 2011;42(3):403-8.
3. Kleinman GM, Young RH, Scully RE. Primary neuroectodermal tumors of the ovary. A report of 25 cases. Hum Pathol. 1984;15(8):632-8.
4. Morovic A, Damjanov I. Neuroectodermal ovarian tumors: a brief overview. Histol Histopathol. 2008;23(6):765-71.
5. Hirahara F, Yamanaka M, Miyagia E, Nakazawa T, Gorai I, Minaguchi H, Kakei M, Yamamoto M, Kitamura H. Pure ovarian ependymoma: report of a case treated with surgery, chemotherapy, irradiation and hyperthermotherapy. Eur J Obstet Gynecol Reprod Biol. 1997;75(2):221-3.
6. Garcia-Barriola V, De Gómez MN, Suárez JA, Lara C, González JE, García-Tamayo J. Ovarian ependymoma. A case report. Pathol Res Pract. 2000;196(8):595-9.
7. Carr KA, Roberts JA, Frank TS. Progesterone receptors in bilateral ovarian ependymoma presenting in pregnancy. Hum Pathol. 1992;23(8):962-5.
8. Erdoğan G, Ozel E, Peştereli HE, Salar Z, Tirak B, Karaveli S. Ovarian ependymoma. APMIS 2005;113(4):301-3.
9. Auerbach R, Mittal K, Schwartz PE. Estrogen and progestin receptors in an ovarian ependymoma. Obstet Gynecol. 1988;71(6 Pt 2):1043-5.
10. Guerrieri C, Jarlsfelt I. Ependymoma of the ovary. A case report with immunohistochemical, ultrastructural, and DNA cytometric findings, as well as histogenetic considerations. Am J Surg Pathol. 1993;17(1):623-32.