Case Report

Signet ring stromal tumor of ovary - Report of an unusual case

Soumya Kundu[1*], Narendranath Hait[2], Priyaranjan Chattopadhyay[3], Taraknath Saha[4], Anjon Debnath[5]

{[1]Demonstrator, Department of Pathology} {[2]Assistant Professor, Department of Gynaecology and Obstetrics} {[3]Assistant Professor, [4]Assosiate Professor, Department of Pathology} Midnapore Medical College and Hospital, Medinipur, West Bengal, INDIA.
[5]Senior Consultant, Department of Pathology, Neotia Getwel Healthcare Centre-Siliguri, West Bengal, INDIA.
Email: drsoumya_2010@rediffmail.com

Abstract

Signet-ring stromal tumor is a rare ovarian neoplasm, only a few cases being reported in the English literature. We report a case of SRST of the right ovary in a 67-year-old woman who presented with abdominal discomfort and underwent total abdominal hysterectomy and bilateral salpingo-oophorectomy. The right ovary showed a part solid part cystic mass 12.5 cm in maximum diameter. On histopathologic examination, the tumor was composed of small round and oval cells with cytoplasmic vacuolization and a typical signet-ring appearance, focally admixed with fibromatous tissue. Special staining revealed that the vacuoles of the tumor cells contained no lipid, mucoprotein, or glycogen. The patient remains free of disease six months after surgery. Signet-ring tumor of the ovary is a rare variant of benign ovarian sex-cord stromal neoplasm and should be distinguished from Krukenberg tumor, the most important differential diagnosis.
Keywords: Signet-ring stromal tumor, ovary, Krukenberg tumor.
Abbreviations: SRST- Signet-ring stromal tumor.

[*]**Address for Correspondence:**
Dr. Soumya Kundu, Demonstrator, Department of Pathology, Midnapore Medical College and Hospital, Medinipur, West Bengal, INDIA.
Email: drsoumya_2010@rediffmail.com
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INTRODUCTION

In 1976, Ramzy first described an unusual ovarian tumor from a 28-year-old woman, which he designated signet-ring stromal tumor.[1] These tumors are rare sex-cord stromal tumors of the ovary and only a small number of cases have been reported subsequently, all of them being in adults, nonfunctioning and benign.[2] We present one such case in a 67-year-old female and discuss the differential diagnoses according to the histopathologic, histochemical and immumhistochemical characteristics.

CASE REPORT

A 67 year-old, post-menopausal woman presented with a 6-month history of lower abdominal discomfort. There was no relevant medical or surgical history. Pelvic ultrasound examination revealed a normal-sized uterus and a semisolid right ovarian mass with heterogeneous echogenity. Serum tumor markers including CA-125, CA 15.3, and CEA were all within the normal range. The patient then underwent total abdominal hysterectomy and bilateral salpingo-oophorectomy. Except for the right ovarian mass, all other pelvic and abdominal organs appeared to be normal. On gross examination, a large bi-lobed cyst measuring 12.5 x 9.5 x 7.0 cm was present in the right ovary. Cut surface revealed the larger lobe was filled completely with straw colored slimy fluid. The other lobe on cutting was mostly solid (about 70%), yellowish in appearance, rest being multilocular cystic. (Figure 1) The external surface was smooth, shiny without any breach in the surface. The uterus, cervix, left ovary and both fallopian tubes were unremarkable grossly. Histopathologic examination of the right ovary showed a circumscribed but unencapsulated tumor surrounded by a thin rim of normal ovarian stroma. The solid area was composed entirely of small round signet

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ring shaped cells containing large round cytoplasmic vacuoles. (Figure 2) These cells stained negative for mucin (by PAS stain). (Figure 3) The stroma was loose fibromyxoid in appearance, (Figure 4) with intervening microcystic spaces often filled with basophilic granular material, negative with PAS stain. There was no significant nuclear atypia or mitotic activity and desmoplastic stromal reaction was absent. No epithelial differentiation (glands, nests, cords) were seen. Other areas of the tumor consisted of cystic areas lined by flattened cuboidal epithelium. The other genital organs did not reveal any significant abnormality. A histologic diagnosis of SRST, based on above findings, was made. The patient was followed up to 6 months; she did not have any complaints.

Figure 1: Gross Photograph of partly solid partly cystic right ovarian mass

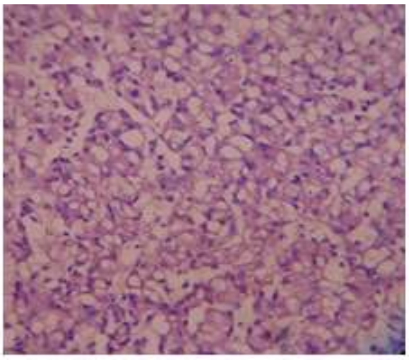

Figure 2: Solid tumor area consisting of small round signet ring cells having large round cytoplasmic vacuoles (H and E 400X)

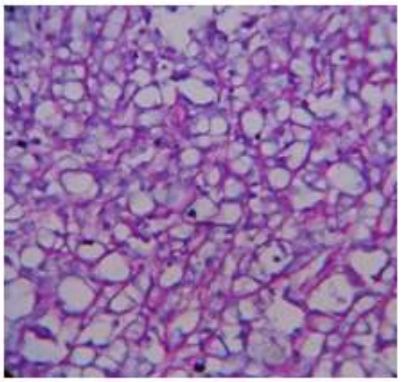

Figure 3: Cytoplasmic vacuoles of signet ring cells staining negative for mucin (PAS 400X)

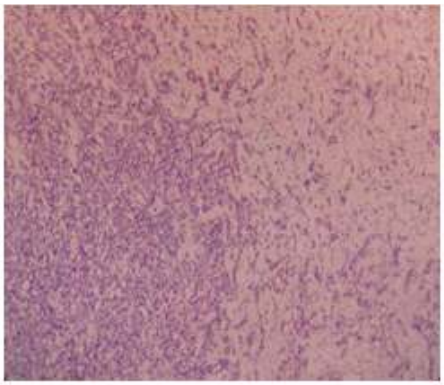

Figure 4: Loose fibromyxoid stroma with solid tumor area in the left hand corner (H and E 100X)

DISCUSSION

According to WHO, SRST is a rare sex cord stromal tumor of ovary composed of signet-ring cells that do not contain mucin, glycogen or lipid.[3] Macroscopically the tumors may be partly solid and partly cystic, as in our case, or entirely solid. The tumor was devoid of mitotic activity, nuclear atypia and pleomorphism, based on which it was designated as benign. After the first description of a SRST of the ovary,[1] 13 other cases of benign signet- ring tumors originating from the ovary were described.[4] In all the previously reported SRSTs as well as in our case, the tumors were unilateral, confined to the ovary and measured 13 cm or less in greatest dimension.[5] The mean age of the patients was 52 years, ranging from 21 to 83 years,[6] while our patient was 67 years old. The most frequent symptom was abdominal pain as in the present case and almost all of the tumors showed a solid appearance under ultrasonographic assessment,[7] which in our case was solid-cystic. The major differential diagnoses of this tumor are Krukenberg tumor and Sclrerosing stromal tumor of the ovary. It is very important to distinguish this tumor from Krukenberg tumor, because a SRST is a benign tumor, whereas a Krukenberg tumor is a metastatic cancer with poor prognosis.[8] Krukenberg Tumor is a metastatic adenocarcinoma to the ovary, often of signet ring cells, arising in the GI tract (stomach, colon, pancreas and gallbladder), breast or bladder. In the literature, bilaterality and extra-ovarian involvement were not seen in any of the SRSTs. Bilaterality has been observed in up

to 93% of Krukenberg tumors.[9] The SRSTs were devoid of epithelial differentiation (glands, nests, cords), as in the present case, whereas all of the Krukenberg tumors contained these epithelial structures at least focally.[2] Ancillary techniques such as special stains for mucin are very helpful in discriminating SRST from Krukenberg tumors; all the SRSTs are negative for PAS, whereas the signet-ring cells component is positive in all cases of a Krukenberg tumor.[8] Immunohistochemically Pancytokeratin is a very useful marker with no expression in the SRSTs but with consistent staining of Krukenberg tumors.[2] Vimentin is also helpful in separating the two lesions, with positivity in SRSTs.[10] Sclrerosing stromal tumor is a benign, usually unilateral, ovarian stromal tumor which occurs in young adult females and is associated with excessive estrogen secretion. The microscopic appearance is of fibroblasts and vacuolated cells supported by edematous hypocellular regions with variable vascularity. The vacuolated cells, probably degenerating lutein cells, resemble signet ring cells, and stains positively for lipid (oil red O stain). SRST lacks the pseudolobulation, lipid-rich cells, and prominent vascularity of the Sclrerosing stromal tumor.[11] In the available literature, follow-up of the cases remained uneventful.

CONCLUSION

SRST is an extremely rare benign ovarian tumor with excellent prognosis according to follow-up data. These tumors may be misdiagnosed as Krukenberg tumors due to the presence of signet-ring cells, with clinical, operative and histological findings along with immunohistochemistry and mucin stains aiding to make a correct diagnosis.

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