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Cell & Cellular Life Sciences Journal
ISSN: 2578-4811

Void and Falcate-Signet Ring Stromal Tumour-Ovary

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Editorial

Volume 9 Issue 1

Received Date: December 28, 2023

Published Date: January 08, 2024

DOI: 10.23880/cclsj-16000192

Keywords: Tumour; Genetic Mutations; Cellular Atypical; Mitosis; Necrosis

Abbreviations: EMA: Epithelial Membrane Antigen; SF1: Steroidogenic Factor 1; PAS: Periodic Acid Schiff's; AUB: Abnormal Uterine Bleeding; AJCC: American Joint Committee on Cancer; IC1: Abdomino-Pelvic Cavity.

Editorial

Signet ring stromal tumour represents as an exceptionally discerned, benign, primary ovarian neoplasm wherein the ovarian stroma is pervaded with signet ring cells. Additionally designated as signet ring stromal cell tumour or signet ring stromal tumour, tumour cells are permeated with cytoplasmic vacuoles, thereby configuring signet ring cells. As the ovarian stromal neoplasm is constituted of variable proportion of signet ring cells and spindle shaped cells, nevertheless, vacuolated signet ring cells are devoid of lipid, mucin or glycogen.

Neoplastic stromal signet ring cells appear focally or diffusely immune reactive to sex cord biomarkers as steroidogenic factor 1 (SF1), inhibin or calretinin. Characteristically, tumour cells are immune non-reactive to epithelial membrane antigen (EMA).

Although no age of tumour emergence is exempt, the neoplasm is encountered within 28 years to 70 years with mean age of disease occurrence at 49 years. Tumefaction preponderantly incriminates the ovary [1,2].

Of obscure aetiology, neoplasm is posited to arise from ovarian stromal cells or may occur due to multifocal conversion of ovarian fibroma. Ovarian signet ring stromal tumour appears devoid of FOXL2 or CTNNB1 genetic mutations [1,2].

Signet ring stromal tumour of ovary is associated with nonspecific clinical features as abdominal pain, pelvic pain or abnormal uterine bleeding (AUB). Additionally, tumour may be incidentally discovered upon imaging [2,3].

Grossly, a characteristic well circumscribed, un-encapsulated, preponderantly unilateral, soft to firm neoplasm is expounded. Tumour magnitude varies from 3.0 centimetres to 20 centimetres with mean tumour diameter of 12.8 centimetres. Cut surface is yellow to tan with foci of grey/white fibromatous areas. Variable foci of haemorrhage, necrosis and cystic spaces may emerge [2,3].

Upon microscopy, tumefaction is composed of variable proportion of dual cellular population. Diffuse dissemination of spherical cells permeated with singular to multiple intracytoplasmic, optically clear vacuoles may occur. Intracytoplasmic vacuoles may peripherally displace the nuclei which appear indented and manifest a 'crescent' shaped outline, thereby configuring the pathognomonic signet ring cells. Additionally, a population of fusiform cells impregnated with eosinophilic cytoplasm and ovoid nucleus is discerned.

Tumefaction depicts fibroma-like areas which intermingle with the signet ring cellular component [3,4]. Cellular atypia, mitotic figures or tumour necrosis are absent to minimal. Occasionally, brisk mitotic activity ranging from 13 mitosis/10 high power fields to 16 mitosis /10 high power fields is observed (Figure 1). An estimated ~50% neoplasms depict intracytoplasmic hyaline globules which may be highlighted by Periodic acid Schiff's(PAS) stain [3,4].

Ultrastructural examination exhibits cytoplasmic vacuoles configured due to generalized oedema within cytoplasmic matrix, distended mitochondria or invagination of cellular membranes by extracellular matrix with configuration of pseudo-inclusions (Figure 2). Basement membrane appears to circumscribe cellular aggregates. Free ribosomes are absent. Hyaline globules simulate features of lysosomes or appear reminiscent of degenerating erythrocytes [3,4].

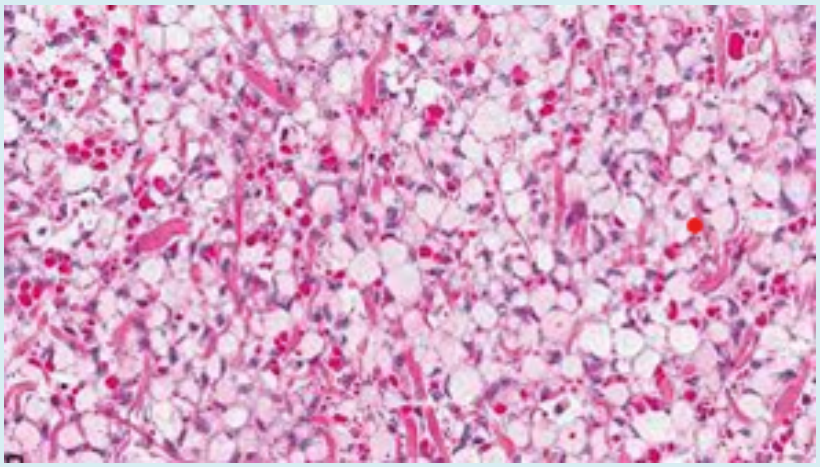

Figure 1: Signet ring stromal tumour composed of ovarian stromal cells impregnated with significant intracytoplasmic vacuoles indenting the nuclear membrane and regular, ovoid nuclei. Surrounding stroma is fibrotic. Cellular atypia, mitosis and necrosis are negligible [5].

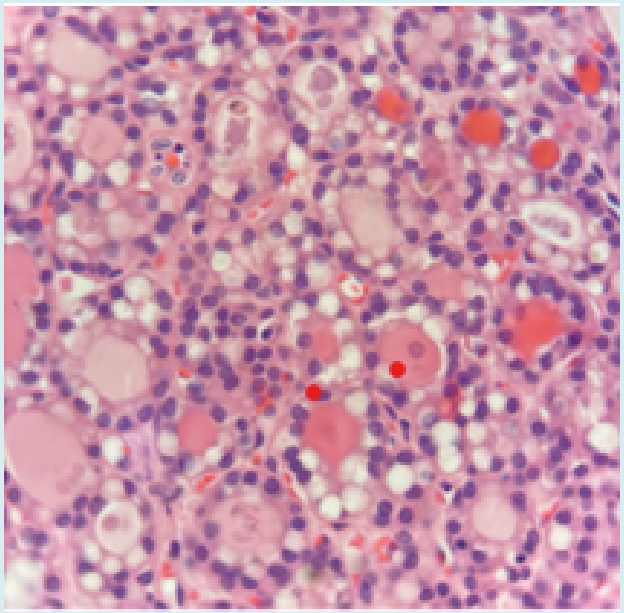

Figure 2: Signet ring stromal tumour delineating ovarian stromal cells pervaded with significant intracytoplasmic vacuoles indenting the nuclear membrane and uniform, ovoid nuclei. Circumscribing stroma is fibrotic. Cellular atypia, mitosis and necrosis are negligible (Table 1) [6].

Tumour	Node	Metastasis
TX: Tumefaction cannot be assessed	NX: Regional lymph nodes cannot be assessed	
T0:No evidence of primary tumour	N0: Regional lymph nodes devoid of tumour cell deposits N0(i+): isolated tumour cells in regional lymph nodes ≤0.2mm	M0: Tumour metastasis within splenic or hepatic parenchyma or distant sites are absent
T1: Tumour confined to ovaries or fallopian tubes • T1a: Tumour confined to single ovary, fallopian tube. Malignant ascites or surface tumour cells or within peritoneal washings are absent. • T1b: Cancer cells in both ovaries and fallopian tubes. Absent malignant ascites. Peritoneal washings are tumour-free • T1c:Cancer cells in both ovaries or fallopian tubes ~capsule rupture with surgical spill(IC1) ~cancer cells on surface of one ovary or fallopian tube or capsule rupture prior to surgery(IC2) ~cancer cells within peritoneal washings or malignant ascites (IC3)	N1:Tumour spreads to retroperitoneal, pelvic or para-aortic lymph nodes N1a: Lymph node metastatic deposit >0.2mm to ≤10mm N1b: Lymph node metastatic deposit >10mm	M1: Distant metastasis M1a:Malignant pleural effusion M1b: Deposits within pulmonary, splenic or hepatic parenchyma, gastro-intestinal tract, bones, extra-abdominal lymph nodes
T2: Tumour within one or both ovaries or fallopian tubes with pelvic spread into uterus, bladder, sigmoid colon, rectum or primary peritoneal carcinoma • T2a: Tumour invasion into uterus or ovaries or fallopian tubes • T2b:Tumour upon surface or invades pelvic organs as bladder, rectum, sigmoid colon		
T3:Tumour confined to one or both ovaries or fallopian tubes or primary peritoneal carcinoma or invasion beyond pelvis • T3a: Cancer cells are invisible to naked eye, microscopic deposits • T3b:Cancer cell deposits ~2cm • T3c: Cancer cell deposits >2cm, upon liver or splenic capsule		

Table 1: TNM classification of carcinoma-ovary [3,4].

As per American Joint Committee on Cancer (AJCC) and International Federation of Obstetrics and Gynaecology (FIGO), staging of serous carcinoma of ovary is denominated as

- Stage I where tumefaction is confined to singular or bilateral ovaries or fallopian tubes and tumour metastasis within retroperitoneal lymph nodes or distant sites is absent.
- Stage Ia where tumefaction is confined to singular ovary or fallopian tube. Cancer cells are absent from ovarian surface or peritoneal or abdomino-pelvic washings. Malignant ascites is absent. Tumour metastasis within retroperitoneal lymph nodes or distant sites is absent stage.
- Ib where tumefaction is confined to bilateral ovaries or fallopian tubes. Cancer cells are absent from ovarian surface or peritoneal or abdomino-pelvic washings. Malignant ascites is absent. Tumour metastasis within retroperitoneal lymph nodes or distant sites is absent.
- Stage Ic where tumefaction occurs within singular or bilateral ovaries or fallopian tubes and tumour metastasis within retroperitoneal lymph nodes or distant sites is absent [3,4].
- Capsule rupture with surgical spill of neoplastic cells into abdomino-pelvic cavity (IC1) cancer cells disseminated upon surface of singular ovary or fallopian tube with

capsule rupture prior to surgery (IC2).

- Cancer cells discernible within peritoneal or abdomino-pelvic washings or malignant ascites (IC3) [3,4].
- Stage II where tumour incriminates unilateral or bilateral ovaries or fallopian tubes with extension into pelvic organs as uterus, urinary bladder, sigmoid colon, rectum or primary peritoneal carcinoma. Tumour metastasis within retroperitoneal lymph nodes or distant sites is absent.
- Stage IIa where tumour infiltrates into uterus or ovaries or fallopian tubes. Tumour metastasis within retroperitoneal lymph nodes or distant sites is absent [3,4].
- Stage IIb where tumour deposits appear upon the surface or infiltrate pelvic organs as urinary bladder, rectum or sigmoid colon. Tumour metastasis within retroperitoneal lymph nodes or distant sites is absent [3,4].
- Stage III subdivided into
- Stage IIIA1 where tumefaction involves unilateral or bilateral ovaries or fallopian tubes or primary peritoneal carcinoma with infiltration of pelvic organs and dissemination into retroperitoneal, pelvic or para-aortic lymph nodes. Distant metastasis is absent [3,4].
- Stage IIIA2 where tumefaction involves unilateral or bilateral ovaries or fallopian tubes or primary peritoneal carcinoma with infiltration of pelvic organs. Carcinoma cell deposits are invisible to naked eye although can be discerned microscopically. Dissemination into retroperitoneal, pelvic or para-aortic lymph nodes may or may not occur. Distant metastasis is absent [3,4].
- Stage IIIb where tumefaction involves unilateral or bilateral ovaries or fallopian tubes or primary peritoneal carcinoma with infiltration of pelvic organs. Carcinoma cell deposits are ~ 2 centimetre magnitude and discernible to naked eye. Dissemination into retroperitoneal, pelvic or para-aortic lymph nodes may or may not occur. Distant metastasis is absent [3,4].
- Stage IIIc where tumefaction involves unilateral or bilateral ovaries or fallopian tubes or primary peritoneal carcinoma with infiltration of pelvic organs. Carcinoma cell deposits appear upon hepatic or splenic capsule and exceed > 2 centimetre magnitude. Dissemination into retroperitoneal, pelvic or para-aortic lymph nodes may or may not occur. Distant metastasis is absent [3,4].
- Stage IV where tumour involves unilateral or bilateral ovaries
- Stage IVa where malignant pleural effusion occurs. Neoplastic dissemination into liver, spleen, gastrointestinal tract or extra-abdominal lymph nodes is absent [3,4].
- Stage IVb where tumefaction extends into hepatic, pulmonary or splenic parenchyma, bones or extra-abdominal lymph nodes [3,4].

Signet ring stromal tumour appears immune reactive to sex cord biomarkers as steroidogenic factor1 (SF1), calretinin, inhibin or Wilm's tumour 1(W1) antigen. Variable immune reactivity to CD10, cytokeratin AE1/AE3 and Cam 5.2 is observed [5,6].

Intracytoplasmic vacuoles appear immune non-reactive to mucins upon staining with mucicarmine, Periodic acid Schiff's stain with diastase resistance or Alcian blue, glycogen as highlighted by Periodic acid Schiff's stain or Alcian blue and lipid as discerned by Sudan III or Oil Red O stains. Tumour cells are immune non-reactive to cyclin D1, cytoplasmic β catenin, epithelial membrane antigen (EMA), CK7 or CK20 [7,8].

Signet ring stromal tumour of ovary requires segregation from neoplasms as Krukenberg tumour, microcystic stromal tumour or signet ring cell change as encountered within fibroma, adult granulosa cell tumour , Brenner's tumour or serous cystadenofibroma.

Cogent histological evaluation of surgical tissue samples as oophorectomy may appropriately categorize signet ring stromal tumour of the ovary [7,8]. Characteristically, serum levels of CA-125 appear within normal limits or may be minimally elevated [7,8]. Upon imaging, neoplasm expounds non-specific features and appears non discernible from diverse ovarian neoplasms. Tumefaction may appear solid or cystic or enunciates a complex countenance [7,8]. Signet ring stromal tumour may be appropriately alleviated by surgical extermination procedures as oophorectomy. Tumefaction is devoid of distant metastasis or tumour reoccurrence [7,8].

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