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A RESEARCH STUDY ON LOW GRADE MULLERIAN ADENOSARCOMA OF POUCH OF DOUGLAS RECURRING AS BILATERAL OVARIAN HIGH GRADE MULLERIAN ADENOSARCOMA WITH RHABDOMYOSARCOMATOUS OVERGROWTH

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Abstract:
Mullerian adenosarcoma (MA) of ovary with sarcomatous (rhabdomyoblastic) excess is an amazingly uncommon harmful kind of female genital tract neoplasm. These tumors are exceptionally forceful and nearness of heterologous components is related with more awful visualization. A multi year old female gave lower stomach torment and distension. She had history of expulsion of tumor from pocket of Douglas in 2017 for which she didn't get any extra treatment nor did she keep consistent development. Current preoperative radiological examination uncovered respective ovarian masses. She experienced stomach hysterectomy with reciprocal oophorectomy. Infinitesimal examination uncovered biphasic tumors showing sarcomatous abundance with rhabdomyoblastic separation. Survey of the past biopsy uncovered second rate Mullerian adenosarcoma without sarcomatous excess. Subsequently the present tumor was viewed as intermittent. This report features the forceful idea of MA even with poor quality morphological highlights and underlines the significance of nonstop development and extra treatment.

Keywords: *sarcomatous overgrowth, extrauterine, Mullerian adenosarcoma, rhabomyoblastic differentiation.*

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INTRODUCTION:

Threatening mesenchymal neoplasms represent 1-3% of all female genital tract tumors and Mullerian Adenosarcoma (MA) comprises 8-10% of these malignancies. MAs generally emerge in the uterus and introduces as a polypoid mass. In any case, they can likewise include extrauterine locales and ovary is the most widely recognized among these destinations. Other extrauterine destinations incorporate pelvis, vagina, fallopian tube, peritoneum, digestive tract, liver, and etc.[1-3]. As indicated by the traditional portrayal of Clement and Scully, MA is a blended neoplasm involving harmful stromal and generous epithelial component.[4,5] MA with sarcomatous abundance is characterized as halfway excess of a generally ordinary adenosarcoma by unadulterated high evaluation sarcoma possessing in any event 25% of the tumor mass. Further arrangement of MA relies on separation of mesenchymal components; Homologous tumors are made out of nonspecific axle molded sarcomatous cells, while heterologous tumors are related with myoid, chondroid and osteoid differentiation.[2] Tumors with sarcomatous excess have forceful clinical course when contrasted with tumors without sarcomatous growth.

Presentation:

A stomach hysterectomy and two-sided oophorectomy example was gotten at Histopathology Section, Department of Pathology and Laboratory Medicine, Aga Khan University Hospital, Karachi in April 2017. Understanding was a multiyear old female who displayed to the gynecologist with protest of lower stomach torment. On clinical examination, a delicate mass was substantial in the lower belly. Her preoperative ultrasound uncovered a mind boggling mass in right adnexa. X-ray sweep of pelvis demonstrated respective ovarian masses. Right ovarian mass estimated 8.5 x 8 cm with strong improving and cystic necrotic parts. Left ovarian strong mass estimated 5 x 4.5 cm. Uterus was massive and contained different very much characterized hypo exceptional masses (Fibroids). There was no proof of pelvic instinctive, omental and peritoneal association. Pelvic lymphadenopathy was likewise not seen. Her serum CA-125 dimension was 50.9 u/ml. She experienced absolute stomach hysterectomy with two-sided oophorectomy.

Net examinations demonstrated the uterus, cervix, fallopian tubes and different separate light darker, delicate to firm, unpredictable bits of tumor tissue. The biggest of these tissue pieces estimated 13 x 7 cm and the rest estimated 11 x 9 cm in total. On separating, slice surface was tan to light darker and displayed territories of discharge, rot and various cystic spaces. Uterus, cervix and fallopian tubes were horribly not included by tumor. Myometrium additionally indicated three fibroids.

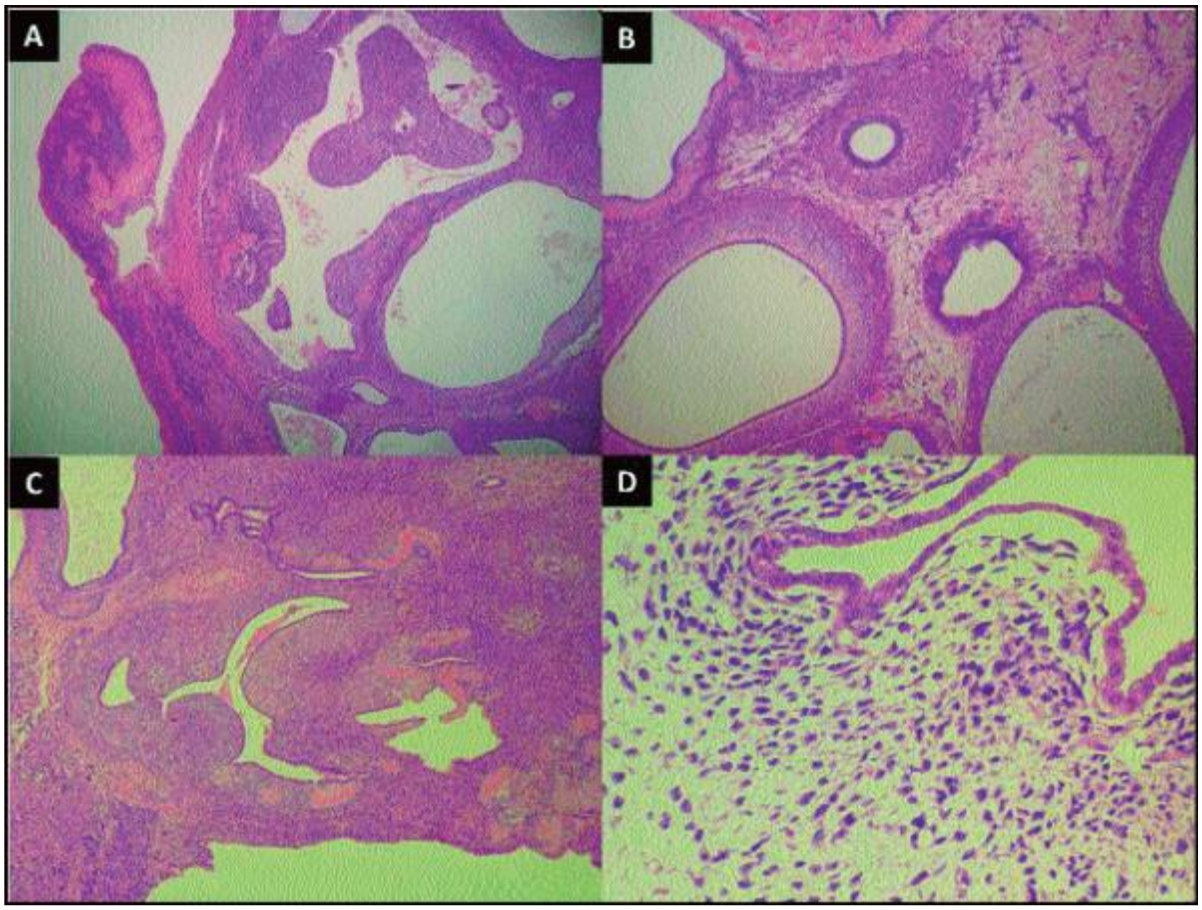

Figure-1: (A) Low power perspective on biphasic tumor displaying cystic spaces. Ordinary ovarian tissue is likewise calculable on left side. (B) Medium power see appearing of stromal part around enlarged glandular structures. (C) Medium power sees appearing of epithelial part by neighboring stromal segment. (D) High power sees appearing low columnar epithelial covering encompassed by poor quality sarcomatous segment

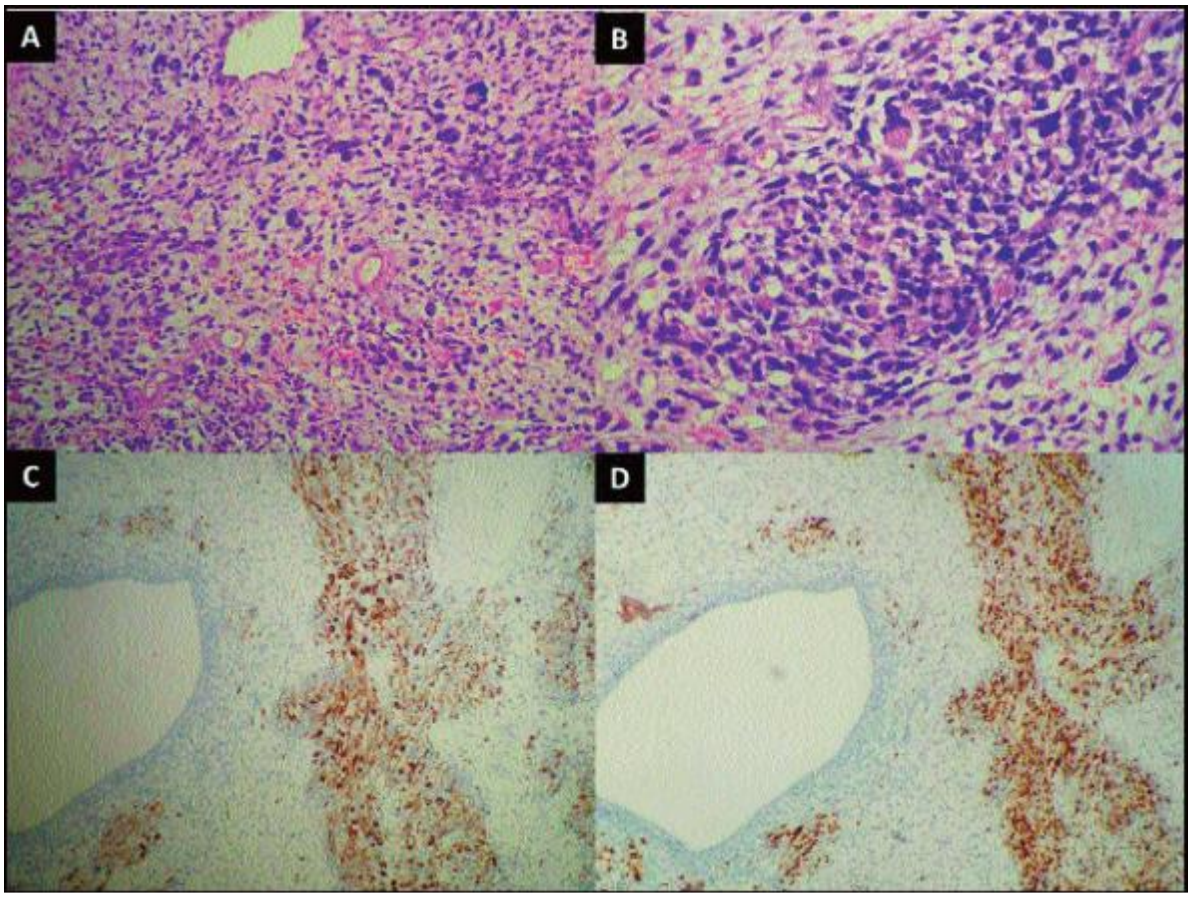

Figure-2: (A) Medium power view and (B) High power perspective on territories indicating rhadomyosarcomatous abundance. (C) Desmin and (D) Myogenin IHC stains exhibiting positive articulation and affirming rhabdomyoblastic separation of tumor cells. Minuscule examination of tumor tissue pieces uncovered a widely necrotic tumor. The feasible regions demonstrated a biphasic development design including dominating stromal segment and dissipated unpredictable, ectatic and packed organs lined by dull looking columnar cells indicating atomic stratification. The stroma segment demonstrated stamped hypercellularity with regions of putrefaction and myxoid change. At a few territories stromal cells showed up dense around the organs (Figure 1A-D). Stromal cells were axle to stellate fit as a fiddle and showed conspicuous rhabdomyoblastic separation. Immunohistochemical (IHC) stains Desmin and myogenin likewise affirmed the rhabdomyoblastic idea of stromal cells (Figure 2AD). The cores were exceptionally pleomorphic and hyperchromatic. Visit mitotic figures (around 22/10 HPF) were likewise valued. At foci, the foundation stroma demonstrated hyalinized plaques taking after "collagen rossettes". Endometriosis and ovarian sex line like components were not found. Centrally, ordinary ovarian stroma was additionally distinguished. Subsequently, the case was analyzed as "Mullerian adenosarcoma with rhabdomyosarcomatous abundance".

Later the patient likewise uncovered that she had experienced laparotomy and extraction of a tumor in the pocket of Douglas in 2017. There was no proof of sickness in the ovaries, other pelvic organs and omentum around then. The slides of the past biopsy were evaluated which additionally uncovered second rate adenosarcoma with no other genealogy separation. Thus, we view our case as repeat of the underlying tumor. Since the patient had a place with are mote provincial zone of the nation, she neither kept constant follow up nor got extra treatment around then. Quiet was sans malady 4 months after the evacuation of the intermittent tumor. Since the tumor had obtained high evaluation and sarcomata's abundance, tolerant was offered adjuvant radiotherapy which she declined because of budgetary reasons. She was then encouraged to keep close catch up with her gynecologist and oncologist.

DISCUSSION:

The expression "Mullerian adenosarcoma" was utilized first in 1974 by Clement and Scully for an

unmistakable uterine tumor portrayed by a dangerous, typically poor quality, stromal part, and a by and large kindhearted, yet at times atypical, glandular epithelial component.[5] Since that time, comparative cases have been accounted for from different places in the writing, the vast majority of which incorporate few patients.[1-3], [6-8] The biggest arrangement on MA included just 40 patients.[2]. Mama can happen at any age including puberty. Extrauterine MAs happen at more youthful age than their uterine partners as found in our patient. Middle age for uterine tumors is 71 years when contrasted with 53 years for extrauterine tumors. In lion's share (97.5%) of cases may will in general happen unilaterally.2 conversely, the ovaries were reciprocally engaged with our case. The regular displaying indications incorporate stomach torment, stomach enlargement with substantial mass and sporadic vaginal seeping as likewise saw in our case.3 Some of these extrauterine tumors have been related with and apparently emerge from endometriosis. While others are not related with endometriosis, they are thought to emerge from surface epithelium, ovarian stroma, or peritoneal mesothelium as a component of optional Mullerian system.[6, 7]

Mullerian adenosarcoma of ovary is commonly comparative infinitesimally to uterine adenosarcoma with the exception of a couple of contrasts like sarcomatous excess is seen in 30% of ovarian cases when contrasted with 8% in uterine adenosarcoma and SCLE (Sex rope like components) were seen in 15% of ovarian cases when contrasted with 7% in their uterine counterparts.[2,4]

Practically all endometrioid tumors of the ovary are carcinomas with the exception of uncommon tumor types for example nadenofibroma and MA. Adenofibroma takes after MA with the exception of that the stroma of adenofibroma needs atomic pleomorphism and mitotic action found in adenosarcoma. The differential conclusion in anyindividual case relies upon the morphologic highlights, for example, nearness or nonattendance of heterologous components, sex rope like components, and sarcomatous excess. The differential conclusion of MA with sarcomatous abundance likewise incorporates endometrial stromal sarcoma [ESS], juvenile teratoma, dangerous mullerian blended tumors (MMMT) and unadulterated sarcomas when heterologous components are available. ESS happens in same age gather as mullerian adenosarcoma and is typically one-sided. ESS looks like stromal segment of adenosarcoma however does not have its glandular part. Exhaustive inspecting ought to be done to look for epithelial part of MA as the forecast of MA is more regrettable than ESS. Youthful teratoma can be avoided based on age as these tumors most normally happen in the initial three many years of life and are practically nonexistent after menopause, and contain embryonal neuroectodermal components and endodermal derivates in practically all cases. Rather than MA, threatening mullerian blended tumors (MMMT) regularly have a high evaluation mesenchymal part and an obtrusive carcinomatous segment. The nearness of average adenosarcoma territories discount the potential outcomes of unadulterated sarcomas [7]. Ovarian MAs have poor visualization, higher repeat and higher death rates than their progressively normal uterine partners. Multiyear ailment free survival is under 25%. [8,9]. The fundamental reason is likely identified with the area of tumors regarding the stomach cavity and the absence of an anatomic boundary to spread. Another reason may likewise be the aftereffect of the bigger size, higher stage, and higher recurrence of crack of the ovarian tumors. Medical procedure is the pillar of treatment and extra chemotherapy as well as radiotherapy is controlled in an extent of patients.

CONCLUSION:

MAs of ovary are uncommon forceful tumors which can give repeat after numerous years, even with second rate morphological highlights. Extra forceful treatment modalities and close clinical follow up ought to be considered in all cases.

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