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Anti-NMDA-Receptor Encephalitis in a Patient with Ovarian Teratoma, Harboring Brain Histology of Varying Developmental Stages and Regions with Chronic Inflammation

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by solid sheets of histiocytes with abundant granular eosionophilic cytoplasm, and intra- and extracellular basophilic bodies. It should be differentiated from other types of granulomatous inflammation and from signet ring cell carcinoma. Treatment consists of antibiotics, but surgery is sometimes necessary.

Methods/Case Report: A 74-year-old woman experienced postmenopausal bleeding for several years. A previous endometrial biopsy showed a mucinous glandular proliferation of uncertain etiology. She also had coronary heart disease, hypertension, diabetes mellitus and hyperlipidemia. A prior urinalysis showed abundant bacteria, and she had been hospitalized recently for urinary tract infection (UTI). She underwent laparoscopic total hysterectomy. Grossly, the endometrium was mostly uniform, 0.1 cm in thickness, with one 0.4-cm polyp. Microscopically, the endometrium showed simple hyperplasia without atypia, benign endometrial polyp, and multifocal malakoplakia, manifested by sheets of histiocytes with granular eosinophilic cytoplasm, many of them containing variably-sized basophilic spherules imparting to them a targetoid, or bull's eye, appearance. Multiple endometrial plasma cells were also seen.

Results (if a Case Study enter NA): NA

Conclusion: Very few cases of endometrial malakoplakia have been reported in the medical literature. Here we present unexpected malakoplakia coexisting with endometrial simple hyperplasia without atypia. Reports from the electron microscopy literature suggest that the basophilic spherules, named Michaelis-Gutmann inclusion bodies, are electron-dense granules that result from coalescence of phagolysosomes, reflecting failed digestion of bacteria. In our patient, longstanding chronic endometritis, confirmed by the presence of endometrial plasma cells, possibly coexisting with chronic UTI, culminated in malakoplakia, which clinically and pathologically mimicked endometrial hyperplasia or even carcinoma, given her old age, chronic bleeding, and the mucinous epithelium in the endometrial biopsy. The finding of endometrial malakoplakia should remind us of the bacterial etiology of chronic endometritis and the need for timely treatment of urogenital infections with antibiotics.

Primary Malignant Melanoma of Vagina: A Clinicopathologic Analysis of 6 Cases

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Introduction/Objective: Primary malignant melanoma of vagina (PMMV) has a very high rate of recurrence and poor long-term survival. Less than 250 cases are reported in English literature to date, optimal treatments, risk factors, and prognostic predictors for PMMV are still subjects of debate. This study sought to evaluate the clinicopathologic features, initial management, and survival of 6 patients over 7 years follow-up in one institution.

Methods/Case Report: In this study, the clinical and pathologic features of 6 PMMV occurring in patients age 49 to 83 years were evaluated retrospectively. The mean age of the patients was 68.2 years, and all patients were postmenopausal women. At the time of diagnosis, all tumors were limited to the vagina.

Results (if a Case Study enter NA): Vaginal bleeding, discharge and a tumor mass were the chief complaints. These patients were treated by pelvic exenteration, radical surgery and postoperative chemotherapy and/or radiotherapy. Follow-up was available in all patients ranging from 1 to 7 years. Two patients had metastases in other organs died at 16 and 34 months respectively. Two patients had distant recurrence at 1 to 2 years and are still alive. The other 2 patients have no evidence of disease over 2 years follow-up. Grossly, the tumors were mostly polypoid and ranged from 0.25 to 9.5 cm (mean, 5 cm) in maximum dimension. The depth of invasion ranged from 2.2 to 11 mm. A vertical growth phase was present in all tumors. S-100, HMB-45 and Melan-A were positive in 100% of cases tested. Depth of invasion, mitotic index of tumor cells, tumor size, age, menopausal status, pelvic lymph node metastasis and lymphocytic infiltration show no tendency towards progression-free survival. Patients with positive or indeterminate margin status demonstrated a higher risk of recurrence than did patients with negative margins. And adjuvant therapy was associated with progression-free survival.

Conclusion: In conclusion, PMMV is a rare disease, predominantly seen in women of postmenopausal age, and is associated with a poor prognosis. This study confirms S-100, HMB-45and Melan-A remains the most sensitive marker. Conventional predictors were of no prognostic value. Positive margin and adjuvant therapy were associated with progression-free survival. This report can facilitate the expansion of the phenotypic spectrum of gynecologic melanomas, and contribute to the prevention of misdiagnosis and inadequate treatment of PMMV.

Anti-NMDA-Receptor Encephalitis in a Patient with Ovarian Teratoma, Harboring Brain Histology of Varying Developmental Stages and Regions with Chronic Inflammation

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Introduction/Objective: Anti-NMDA-receptor encephalitis is a subacute, autoimmune disorder thought to be caused by autoantibodies directed against the N-methyl-D-aspartate (NMDA) receptor. Clinical symptoms of anti-NMDAR encephalitis may mimic schizophrenia

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and psychotic spectrum disorders or substance-induced psychosis. Although initially described in association with ovarian teratomas in women, anti-NMDAR encephalitis has been reported in individuals without paraneoplastic association, as well as in males. Herein, we report a case of a 29-year-old woman with suicidal ideation and other neuropsychiatric manifestations who was found to have a right ovarian cystic mass by imaging study. Microscopically, the resected ovarian mass is composed of mature skin, fat, cartilage and neural tissues. Nerve, ganglions and multiple brain tissues are present. Cerebellum including external granular cell layer (normally only seen in infants), cerebrum-like, choroid plexus and other neural elements are present. There is peripheral lymphoplasmacytic infiltrates around and within the neuroglial matrix. Cerebral spinal fluid was concurrently tested positive for Anti-NMDAR. The combined clinical, histological, and laboratory findings confirmed the above diagnosis. Although Anti-NMDAR encephalitis is a familiar entity to many clinical psychiatrist and neurologists, it is less commonly reported in the pathology literature. Its resultant relationship to cystic teratoma warrants awareness of this condition by pathologists.
Methods/Case Report: Case Report
Results (if a Case Study enter NA): NA
Conclusion: Anti-NMDA-receptor encephalitis is related to cystic teratoma, therefore pathologists need to be aware of this condition.

Characterization of A Rare Case of Vulvar Epithelioid Sarcoma with local recurrence and metastases

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Introduction/Objective: Epithelioid sarcoma (ES) is a rare, malignant mesenchymal neoplasm that has a known tendency for local recurrence, regional lymph node involvement, and distant metastases. Two histologic variants have been recognized: classic ES also known as the distal type, and proximal-type ES (PES). The classic ES is common in young adults. It occurs more frequently in the distal upper extremities followed by the distal lower limbs and has a male prevalence of 2:1. Conversely, PES commonly involves deep tissues in the pelvic region, including the genital area. It tends to occur in older patients and follows a more aggressive clinical course. In the female genital tract, PES occurs most frequently in the vulva. The incidence of primary sarcoma of the vulva accounts for 1.5-5% of all malignant tumors, making PES a very rare incidence.
Methods/Case Report: Here we report a 60-year-old female diagnosed with vulvar epithelioid sarcoma treated with a right radical vulvectomy and bilateral inguinal lymph node dissection in 2008. In 2017, further surgery and adjuvant radiation were given for local recurrence. In 2020, the patient developed left hip pain and was found to have an expansile lytic lesion in the left proximal femur. Extensive resection was performed. Grossly the vulvar lesion was nodular with diffuse hemorrhage, degeneration, and necrosis. Microscopically, the tumor cells had large vesicular nuclei, prominent nucleoli, and abundant eosinophilic cytoplasm. Histologically, the morphology of the tumor cells are similar for the primary vulvar specimen and the bone metastases. Immunohistochemically, the tumor cells are positive for vimentin, GATA, FLI-1, SMA, SMHC, partially positive for CAM5.2, AE1/AE3, CD31, and CD163. Immunohistochemistry was negative for CDX2, CD56, S-100, TTF-1, CK5/6, CK20, P40, mammoglobin, MOC31, ER, CK7, CK903, HMB45, PAX8. A Ki-67 proliferative index was around 30-40%. NGS molecular testing detected a SMARCB1 mutation with loss of exons 1-3 and exons 7-9 supporting the diagnosis of epithelioid sarcoma.
Results (if a Case Study enter NA): NA
Conclusion: In summary, we report a case of PES of the vulva in a 60 year old female. Grossly, the lesion was nodular with histology showing large vesicular nuclei, prominent nucleoli, and abundant eosinophilic cytoplasm. It showed loss of INI1/SMARCB1 nuclear expression. The patient is receiving further adjuvant treatment and shows no new metastases.

High-Grade Sarcoma Arising in the Background of Growing Teratoma Syndrome; A Rare Case

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Introduction/Objective: Growing teratoma syndrome is a rare condition seen in patients with a history of immature teratomas or mixed germ cell tumor status post-treatment. It usually occurs within the first two years of diagnosis but rarely can be seen years later.
Methods/Case Report: We report a case of a 48-year-old female with a previous history of ovarian malignancy in adolescence status post-TAH/BSO and chemotherapy, now presenting with a 23 cm multi cystic septate mass with some solid components in the subdiaphragmatic area, extending into the liver on imaging. Nodular implants along the surface of the right hepatic lobe and multiple peritoneal implants were seen. Histology of peritoneal implants showed fibro adipose tissue admixed with rare scattered glandular elements, epithelial fragments, and mature neuroglial tissue, consistent with mature teratoma. Liver biopsy revealed short fascicles of spindled, epithelioid and rhabdoid tumor cells admixed with foci of the myxoid stroma. Scattered moderate cytologic atypia, atypical mitosis, and necrosis were appreciated. Tumor

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