

POSTERS

POLYPHENOTYPIC EXTRARENAL UTERINE NEPHROBLASTOMA WITH TRILAMINAR DIFFERENTIATION AND A PNET OVERGROWTH

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CASE STUDY: 62-year-old patient was admitted because of methrorrhagia and diagnosed elsewhere of an endometrial stromal sarcoma. TAH-BSO was performed and revealed a globular uterus with a large endocavitary mass. Microscopically, tubular areas positive to WT1, CD10 and CD56 with renal-type glomerulus formation were seen. There were extensive areas of embryonal stroma with cartilage and rhabdomyosarcoma. There were also areas of gland formation of enteroblastic type positive to CK20 and chromogranin, similar to those of "glandular yolk sac tumor". However the predominant pattern was an extensive PNET that invaded the cervix. These areas showed positive Synaptophysin, neurofilaments, NSE, CD56, CD57 and CD99 and areas of neuroblastic and ependymal rosettes.

DISCUSSION: Müllerian carcinosarcomas are the most frequent uterine mixed tumors, which exhibit various tissue components that correspond to various mesodermal type components, although in rare occasions they may differentiate neuroectoderm or other non-mesodermal elements and are thus called teratoid carcinosarcomas. Extrarenal nephroblastomas, a classic example of mixed tumor, may also rarely occur in the female genital tract, where their histogenesis is unknown. Histologically they can reproduce every neoplastic pattern present in renal nephroblastomas, neoplasms that are known to have the potential to differentiate into various tissue components of mesenchymal, neuroectodermal and endodermal nature. The present report deals with a highly complex polyphenotypic extrarenal nephroblastoma in the uterus in a 62 year old patient that differentiated tissues from the three germ layers and was overgrown by an aggressive PNET, establishing the taxonomic difficulties of drawing a boundary between

embryonal type tumors and teratoma. Immunohistochemistry was essential in the differentiation of the various mixed components. This is the first reported case of a trilaminar extrauterine nephroblastoma.

BIPHASIC PLACENTA WITH NORMAL AND MOLAR AREAS COEXISTING WITH LIVE FEMALE FOETUS: DEMONSTRATION BY GENETIC IDENTITY METHODS OF A FUSED TWIN ORIGIN

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INTRODUCTION: The finding of molar areas in an otherwise normal pregnancy coexisting with a live foetus can be explained either by a rare partial mole with embryo or by a twin pregnancy, one of them of complete molar type, with fused placentas. The former situation is invariably associated to triploidy; in the latter, it is mandatory to ascertain the different genetic origin of the two different areas: molar and normal.

CASE STUDY: We present a 27 year old patient 7gesta, 5aborta, with only a live birth in 2003. There were two miscarriages with a pathology study (1999-2005). One of them had abundant trophoblastic inclusions and festooned villa and the second was a partial mole. In the present pregnancy she experienced at 12 week an episode of vaginal bleeding: Ultrasonography revealed a live foetus and areas suggestion of placental hematoma. In the lower implantation edge there were areas of molar change. At 16 weeks a profuse haemorrhage took place and a decision of termination of pregnancy was made due to the intractable hemorrhage. Pathology revealed a female 140gr foetus without any internal or external malformation. Placenta had a normal appearance but at the edge there was a crescent of molar change. After surgery, hCG values returned to normal after 30 days. Pathology showed a normal placenta that continues insensibly with a complete hydatidiform mole. PCR based comparative genetic (forensic) identity studies on maternal and paternal material correlated with

molar and non-molar areas were performed with Identifier® (Applied Biosystems) and showed that complete mole was homozygotic and fully coincided with paternal identity. Triploidy was discarded. Normal placenta and foetus showed both maternal and paternal identities. It was concluded that it was due to fusion of twin pregnancies, one normal and the other molar, of androgenetic type.

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HERPES GESTATIONIS: A SUCCESSFUL TREATMENT WITH CORTICOSTEROID IN ASSOCIATION WITH DAPSONE

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Herpes Gestationis is an uncommon pregnancy and/or the immediate post-partum period inflammatory disorder of the skin, characterized by an intense itching and by widespread urticarial papules and plaques accompanied by vesicles and bullae. Individual lesions are indistinguishable from those of bullous pemphigoid, clinically and histopathologically. Although the cause is not known, the mechanism is thought to be immunologic in response to antibodies to an antigen peculiar to pregnancy.

AIM OF THE STUDY: We report this case because of the rare presentation of this skin disorder, and DIF positivity of IgG autoantibody that is infrequently seen. The aim was to underline the successful association between the corticosteroid and Dapsone therapies.

CASE REPORT: A 30 year-old woman came to our Institute because of a strongly itching, erythematous-bullous dermatitis with the onset first on the abdomen, during the 8th month of pregnancy. Secondly, this eruption rapidly progressed to a generalized pemphigoid-like dermatitis, with lesions on arms and legs bilaterally. A skin biopsy had been performed from an abdominal lesion. The histological features revealed a mild perivascular lymphocytic infiltrate and no other alteration of the papillary dermis. The Direct Immunofluorescence

test revealed a positive IgG deposition along the base of the epidermis in salt-split skin, whereas the DIF test was negative for IgA, IgM and C3. We started with a corticosteroid therapy (16 mg twice/day) in association with Dapsone 100 mg/day. The dermatitis completely regressed in three months.

CONCLUSIONS: We would like to underline the important role of Dapsone therapy in association with corticosteroid systemic therapy in this uncommon disorder, despite the usefulness of Dapsone in the treatment of HG is still discussed.

CYTOLOGIC FEATURES OF PULMONARY BENIGN METASTASIZING LEIOMYOMA FROM THE UTERUS: REPORT OF A CASE

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Benign uterine leiomyoma metastasizing to the lung is an extremely rare lesion that has been reported very infrequently in medical literature. We describe the cytologic findings of a case.

CASE REPORT: A 41-year-old woman showing multiple lung nodules during a routine roentgenogram. She had undergone hysterectomy with oophorectomy for uterine leiomyomas 5 years earlier. The physical examination was carried out within normal limits, and blood tests, sputum and bronchoscopy displayed no pathologic findings. The aspirate CT-guided fine needle aspiration cytology was performed using a fine 22-gauge needle. Several samples were obtained, some being immediately fixed in alcohol and others air-dried, being stained with Pap. and M.G.G., respectively. Unstained material was used for immunocytochemical studies with muscle-specific actin, desmin, S100 protein, HMB-45 and estrogen and progesterone receptors. **Cytologic Findings:** The aspirate consisted of a serosanguineous background with scant cellular material, which was composed of spindle cells either alone, or in loosely groups. Mitosis and nuclear atypia were not present. The aspirate was diagnosed as a fusiform tumor without atypia. The microscopic findings of the lung tumor were similar to those of the uterine leiomyoma, and both lesions were histological benigns. Immunocytochemical studies showed expressed positive nuclear staining for muscle-specific actin, desmin and estrogen and progesterone receptors. Staining for HMB-45 and S100 protein was negative.

DISCUSSION: Benign metastasizing leiomyoma is a rare phenomenon with approximately 75 documented cases in the literature, and usually affects women after hysterectomy for leiomyomas. Pulmonary lesions with a significant spindle cell component are infrequently encountered in FNA. They constitute a heterogeneous group and their differential diagnoses range from reactive processes to benign and malignant neoplasms. A specific diagnosis can sometimes be rendered. The use of ancillary studies such as immunocytochemistry and electron microscopy is helpful in narrowing the differential diagnosis. The identification of estrogen and progesterone receptors has therapeutic implications. Even when a definitive diagnosis cannot be rendered, a list of differential diagnoses would guide patients management. We believe that FNA should be the initial diagnostic procedure in evaluating of patients with pulmonary masses.

UNRECOGNIZED EPIGNATHUS FOLLOWED BY AIRWAY OBSTRUCTION AS A CAUSE OF NEONATAL DEATH

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CASE REPORT: We report here a rare case of unrecognized fetal unidirectional oronasopharyngeal teratoma (epignathus) that caused polyhydramnios in 30-year-old pregnant woman. An ultrasound examination revealed the tumor mass in the mouth of fetus several days before an induced delivery in the 29th gestational week. The intubation because of acute respiratory obstruction was unsuccessful and newborn died shortly after delivery. Autopsy revealed the large, tongue-like tumor mass that arose from the hard palate and filled oral cavity and upper aero-digestive tract. In contrast to the most epignathi, neither protrusion from the oral cavity nor intracranial extension of the tumor was noted. Congenital anomalies including cleft palate were not detected. Histopathological examination revealed the mature teratoma consisting of the skin with hair and glands, well organized neural tissue, fat, muscle, cartilaginous tissue, and the bone in the large, central portion of tumor. Careful prenatal recognition of such congenital tumors, followed by a proper planning of its treat-

ment is warranted since all unrecognized cases of epignathi are potentially fatal due to airway obstruction.

ECTOPIC DECIDUA

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INTRODUCTION: ectopic decidua is described as a physiological phenomenon of a pregnancy, which is a result of subserous stromal cell metaplasia due to progesterone activity. Besides ectopic decidua focal site may be found at lamina propria of uterine tubes, cervix and uterus but also submesothelially over abdominal cavity. Cases of ectopic deciduas are described in ovary, omentum, peritoneum, appendix and also in pelvic lymphatic nodules.

CASE REPORT: at a 24-year-old woman who is 39 week into the first pregnancy, due to breech presentation, oligohydramnion, vaginal septum and two-horned uterus, Caesarean section has been done. In that occasion, over peritoneum and greater omentum various sizes of yellowish and soft nodules were noticed, which are described from gynecologist as connective adhesions and they are taken for pathohistological analysis. After analysis of received tissue, around hyperemic blood vessels it can be seen solitary, however accumulated large polygonal cells with eosinophilic cytoplasm and large symmetrical nuclei with nucleoli. Ectopic decidua was infiltrated with mature fatty cells as also poor chronic inflammatory infiltrate.

DISCUSSION AND CONCLUSION: besides ectopic decidua as also decidual mesothelioma, differentially diagnostic it must take into consideration metastatic carcinoma as well metastatic melanoma. Ectopic decidua of peritoneum and greater omentum was confirmed by performed immunohistochemical analysis.

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MECKEL-GRUBER SYNDROME – A CASE REPORT

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A 27-year old woman in 25th week of gestation was admitted to the hospital. Ultrasound examination detected severe oligohydramnion and large multicystic kidneys. Urine bladder and stomach were not visualised. Cordocentesis was performed and normal karyotype, 46 XX, was detected. Hospital ethic committee decided to continue pregnancy. The patient was admitted to the Department of Obstetrics and Gynecology. During 38th week of gestation she vaginally delivered female baby weighted 2250 g. The infant died 1,30 hours later from cardiorespiratory failure. An autopsy was performed and multiple organ malformations were found: occipital encephalocele, bilateral postaxial polydactyly upper and lower extremities, cleft palate, lung hypoplasia, elevation of diaphragm, markedly enlarged multicystic kidneys, and uterine septum. Meticulous morphologic analysis was also made. Meckel-Gruber syndrome, congenital hepatorenal fibrocystic syndrome is rare autosomal recessive disorder which worldwide incidence varies from 1/13250 to 1/140000 live births.

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MATURE SOLID TERATOMA OF THE UTERINE CORPUS – A CASE REPORT

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A 59-year old postmenopausal woman was admitted to our hospital due to operative treatment of uterine prolaps. She had two spontaneous deliveries and one elective abortion over 20 years ago. She underwent curettage twice, first seven years ago due to suspected endometrial polyp when endometrium showed irregular proliferative changes and second, two years ago due

to suspected endometrial hyperplasia when endometrium was normal, inactive. Gynecological and ultrasound examination confirmed uterine prolaps and adnexa were normal. Hysterectomy was performed. On gross examination a flat whitish solid polypoid tumor covered with gray hairs was found in uterine cavum. Microscopically a diagnosis of benign teratoma was made. Mature teratoma is a benign tumor composed of mature somatic tissue, derived from three germ layers. Extragonadal teratomas are rare tumors (1-2% of all germ cell tumors) and their primary presentation in uterus is extremely rare. It was first described in 1929. and to our knowledge there have been 18 cases reported since. The etiopathogenetic mechanism is still unknown. Although rare, mature teratomas should sometimes be included in differential diagnosis of polypoid lesions and tumors of uterus.

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PRIMARY UTERINE B-CELL LYMPHOMA - A CASE REPORT

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Primary non-Hodgkin lymphoma of the uterine corpus is rare. We present a case of low-grade B-cell uterine lymphoma in a 70-year old patient with repeated vaginal bleeding. Immunohistochemistry of the curettage tissue was inconclusive; therefore a diagnosis was established by polymerase chain reaction (PCR) technique. Computer tomography (CT) of the pelvis and thorax revealed a solid expansive mass of the uterine corpus and a concomitant tumor in the left lung, respectively. Subsequently hysterectomy with bilateral salpingo-oophorectomy was performed. Transthoracic puncture of the suspicious pulmonary lesion was excluded by PCR. The disease was classified as stage I according to the Ann-Arbor classification. She was treated with standard CHOP chemotherapy. The patient is alive and free of the disease after a follow-up period of five years.

TWIN PREGNANCY WITH A FETUS AND A COEXISTANT ANEUPLOID HYDATIFORM MOLE

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Twin pregnancy with one normal fetus and a coexistent hydatiform mole is rare. With high resolution ultrasound this event can be suspected, but is rarely diagnosed, firstly because of its rarity, and because molar tissue can resemble a placental hematoma, especially during early pregnancy.

We present a case of aborted twin pregnancy with one normal fetus and placenta, while the other placenta showed the morphological and flow cytometric characteristics of a partial hydatiform mole.

A 29-year old woman (P 0, G 1) presented with 13 wks gestation because of pelvic pain and haemorrhagic vaginal discharge. Neither previous ultrasonic examination, nor the one made at admission showed signs of twin pregnancy. Thirty minutes after admission she aborted and the material was sent for pathological examination. It consisted of the placenta that measured 10:6:2 cm and showed 4:1 cm retroplacental hematoma. The placenta was connected with a male fetus (crown-rump length 9 cm) with a paracentrally inserted umbilical cord. An intact 6 cm gestational sac was received separately; it was filled with brownish viscous fluid, without any signs of fetus or umbilical cord, while the placental tissue grossly showed distended, cystic villi with the diameter up to 1 cm. The autopsy of the fetus showed normal findings. Microscopically, the first placenta was immature, appropriate for gestational age, as were the tissues of the fetus. The other placenta showed cystically dilated villi with central cysterns, surrounded with proliferating cito- and syncytiotrophoblast. Tissue sample from both placentas, as well as the fetal tissue sample was sent for flow cytometric analysis. The first placenta and the fetal tissue were showed to be diploid, while the tissue from the second placenta showed aneuploid pattern. The diagnosis of aborted diamniotic dichorionic twin pregnancy with retroplacental hematoma of the first placenta was made, as well as the diagnosis of complete hydatiform mole of the second placenta. On the first day after the abortion the level of beta hCG were 103000 U/L; on the 2nd day it was 12 700 U/L and one week later 2490 U/L.

Most rare, similar cases described in the literature consisted of twin pregnancy with a normal twin and placenta coexistent with complete hydatiform mole, either aborted or rarely, carried to term. The occurrence of twin pregnancy coexistent with a partial hydatiform mole is even less frequent.

MORPHOLOGICAL CHARACTERISTICS OF MECKEL GRUBER SYNDROME

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We describe a case of Meckel or Meckel Gruber syndrome in a fetus that was aborted because of suspected multiple malformations at 20 wks gestation.

The fetus was a male, that on gross examination showed an edematous nuchal region, ranging from the tip of the head until the lower posterior neckline, postaxial hexadactyly on both hands and enlarged abdomen. On autopsy, both kidneys were enormously enlarged, with smooth, unlobulated surface covered with petechial haemorrhages. The cut surface was spongy, containing numerous small cysts. The renal pelvis was normal, but both ureters were narrow and the urinary bladder was small and empty. The adrenals were small and compressed. Both testes were intraabdominal. Grossly, the liver showed only the remains of a ruptured subcapsular hematoma. The head showed a 0,4 cm defect in the region of the posterior fontanelle with protruding 0,7 cm sacular cavity with smooth whitish inner surface. The cavity was connected with both posterior horns of lateral ventricles with a thin cord of translucent tissue. The frontal regions of the brain were not divided into two lobes, showing only one enlarged ventricle surrounded with thinned layer of cerebral tissue. In the temporal and occipital region the brain was divided into two hemispheres, with enlarged posterior horns of lateral ventricles and the reduction of the surrounding brain parenchyma. On microscopic examination the kidneys showed marginally preserved cortex with small, immature glomeruli, while most of the kidney parenchyma was replaced with irregular cysts of different diameter and orientation, covered with flattened epithelium and separated by fibrous interstitium. The liver showed enlarged and irregular portal spaces with peripheral flattened irregular bile ducts, and foci of extramedullary hematopoiesis. The sac in the region of posterior fontanelle was

covered with thin regular epidermis, with edematous fibrous tissue underneath and a cystic cavity covered with a layer of epithelium that focally showed papillary features and resembled the chorioid plexus. The lungs were hypoplastic. Other findings were unremarkable.

The diagnosis of Meckel Gruber syndrome was made showing most of the described characteristics of this syndrome: posterior encephalocele, postaxial hexadactyly, polycystic kidneys (of undetermined type), biliary dysgenesis (ductal plate malformation) and semilobar holoprosencephaly with hydrocephaly.

OBSTRUCTIVE LESIONS IN THE “OUTLET” AND “INLET” TRACTS OF THE HEART CAUSED BY TUMORS IN INFANTS

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AIM: Our study aimed to diagnose and identify tumors in infants with congenital heart malformations (CHM).

METHOD: 260 hearts with CHM were analyzed. Histological sections were stained with HE, Trichrom,

VanGieson-Elastica, as well immunohistochemical analyzes for tumor reactivity to: actin, desmin was done.

RESULTS: In five of the analyzed cases (about 1%) rhabdomyoma was found and the tumor was cause of various hemodynamical disturbances. In the first case (pre-maturus, male with g.w. 1200gr/36cm), a well circumscribed, and non-encapsulated gray-white tumor was found with dimension of 25x20x15 mm, located on the anterior wall of the LV. Histological analysis has shown a tumor with striated muscle cell differentiation and so called “spider cells”. The second case (fetus mortus in utero, female with g.w. 3350/52cm) presented with cardiac rhabdomyoma. Further morphological analysis revealed well circumscribed tumor nodules in the myocardium of the RV, LV and LA. Tumor nodules were with various dimensions: in the RV with d=10mm, LV with d=15mm and in the LA with d=32mm. The autopsy protocol disclosed presence of glial tubers, the so called tuberous sclerosis. There were multiple tumor nodules in the myocardium of the LV and RV in the other 3 cases.

CONCLUSIONS: Out the 5 cases in our study, where rhabdomyoma was found, tumor was localized in LV in one case and it led to obstruction of the “inlet” of the LV and finally caused subaortic stenosis. Rhabdomyoma was associated with tuberous sclerosis in 2 of the cases.