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CASE REPORT

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Abdominal germ cell tumors in children – report of two cases

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Summary

Background: Abdominal germ cell tumors are neoplasms which originate from the primary germ cells. Diagnostic imaging (US, CT and MRI) can detect and localize the tumor as well as show its structure. We present two cases of germ cell tumors, which, despite very good imaging in sonography and CT, caused problems in final diagnosis.

Case report: In a boy, a polycyclic tumor of the liver hilus was detected. In a girl, two tumors in the pelvis and a few others in the retroperitoneal space were detected. Lymphoma, neuroblastoma and PNET tumors were included in the differential diagnosis. In case of clinical suspicion of an abdominal tumor, including germ cell tumors, diagnostic imaging should begin with sonography.

Conclusions: Replacing CT, which is not charged with ionizing radiation, with MRI, especially in monitoring of the treatment effects, should be considered. The ultimate diagnosis of the tumor type must be established on the basis of histopathological examination.

Key words: germ cell tumors • children • US • CT

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Introduction

Germ cell tumors account for 3% of malignant abdominal and pelvic tumors in children. They originate from primary germ cells at various stages of differentiation. World Health Organization divides these tumors into the following groups: mature and immature teratomas, dysgerminomas, seminomas with spermatocytic seminomas, embryonal carcinomas, polyembryomas, chorioncarcinomas and yolk sac tumors [1]. Mixed tumor forms are also observed.

The primary sites of germ cell tumors are gonads (90-95%), sacrococcygeal region, retroperitoneal space, mediastinum and central nervous system [2]. The tumor location is associated with the age of the child. In neonates and infants, the tumors are found most frequently in the sacrococcygeal region, in adolescents in the remaining locations [3]. The prognosis in most childhood germ cell tumors is, in case of early detection, good irrespectively of age at which the diagnosis was established [4, 5].

The clinical symptoms are nonspecific and dependent on tumor location. In case of abdominal tumors they include constipation, miction disturbances, pain, gait abnormalities, nausea and vomiting, sometimes acute abdomen symptoms. The tumor may cause deformation of the abdomen and may be detectable by palpation. Premature appearance of puberty symptoms should be an indication for search for germ cell tumors.

Determination of tumor marker levels: alpha-fetoprotein (AFP) and human chorionic gonadotropin (β-HCG) can be helpful in diagnostics and monitoring of treatment results. Lesions with yolk sac tumor component produce alpha-fetoprotein where as chorionepithelioma – chorionic gonadotropin [3].

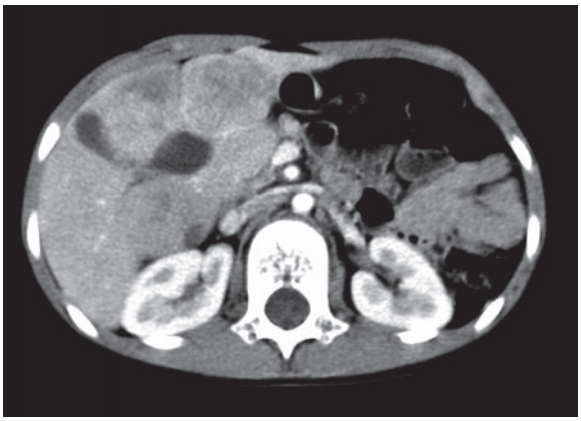

Figure 1. Boy MM. Abdominal CT study after contrast medium administration. A polycyclic, non-homogeneous tumor in the hilus and left lobe of the liver.

Imaging diagnostics

Imaging studies allow to visualize the tumor, to assess its dimensions and pattern of growth, and sometimes to determine the primary site of origin. Germ cell tumors are usually single, cyst-like, solid or mixed structures. They may contain calcifications [1, 6]. The site of origin, localization and appearance allow sometimes preliminary diagnosis of tumor type [6, 7].

In USG, mature teratomas are well-delineated cyst-like structures, often (in ca. 50% of cases) containing peripheral calcifications. Immature teratomas, because of their mixed cystic and solid structure are characterized by inhomogeneous echogenicity, sometimes containing disseminated calcifications [1]. Dysgerminomas and seminomas are homogeneous, solid, well-delineated tumors. In color-coded USG, blood flow can be visualized within these tumors. On testicular USG, attention should be paid to the presence of microcalcifications, which are regarded as the risk factor for seminoma, and by some authors as a precancerous condition [8]. Embryonal carcinomas and chorionepitheliomas are inhomogeneous tumors with hypoechogenic necrotic and hemorrhagic areas.

Conventional radiological examinations (plain radiography, urography) demonstrate calcifications and indirect signs of the tumors, such as abnormal arrangement and separation of intestinal loops, displaced kidneys, ureters, distension of the pyelocalyceal system [10].

In CT, mature teratomas are characterized by low tissue density typical adipose tissue. Immature teratomas have inhomogeneous structure, sometimes reacting to contrast administration. Dysgerminomas are homogeneous, with characteristic reticular pattern of contrast enhancement [1]. The remaining germ cell tumors, are, inhomogeneous, with hemorrhagic and necrotic areas.

Magnetic resonance, associated with lack of ionizing radiation allows multiplanar scanning and is very useful in imaging of pelvis and retroperitoneal space. Fat suppression sequences are useful in visualizing teratomas. No signal in this sequence, corresponding to the presence of adipose tissue and solid parts of the tumor, allows to differentiate between mature and immature teratomas. MR well demonstrates the presence of fibrovascular septa in dysgerminomas [9].

Localization of the tumor, relations with adjacent organs, presence of distant metastases are assessed according to clinical staging classification – SFOP (Tab. 1).

Surgical resection is the main method of treatment of germ cell tumors. Sometimes such treatment is sufficient. According to the protocol (TGM-95), surgical treatment is preceded by determination of tumor marker levels (βHCG, AFP) and imaging studies necessary to assess the extent and structure

Table 1. Postoperative pTNM – SFOP classification.

pT	Primary tumor
pTO	No tumor detected in histopathologic investigation of the specimen
pT1	Complete resection of the tumor with healthy tissue margin (no tumor infiltration in the line of incision)
pT2	Histologically complete resection of the tumor with; the incision extending beyond the organ of origin (T4)
pT3	Residual tumor tissue present
pT3a	microscopic residues
pT3b	macroscopic residues, hemorrhagic exudate or exudate containing tumor cells
pT3c	incomplete resection: simple biopsy
pTx	Infiltration extent impossible to estimate
pNO	No regional lymph node involvement
pN1	Regional lymph node involvement
pN1 a	Complete resection of the affected lymph nodes
pN1 b	Incomplete resection of the affected lymph nodes
pNx	The extent of lymph node involvement impossible to estimate: no surgical resection of lymph nodes was performed or adequate histological information unavailable

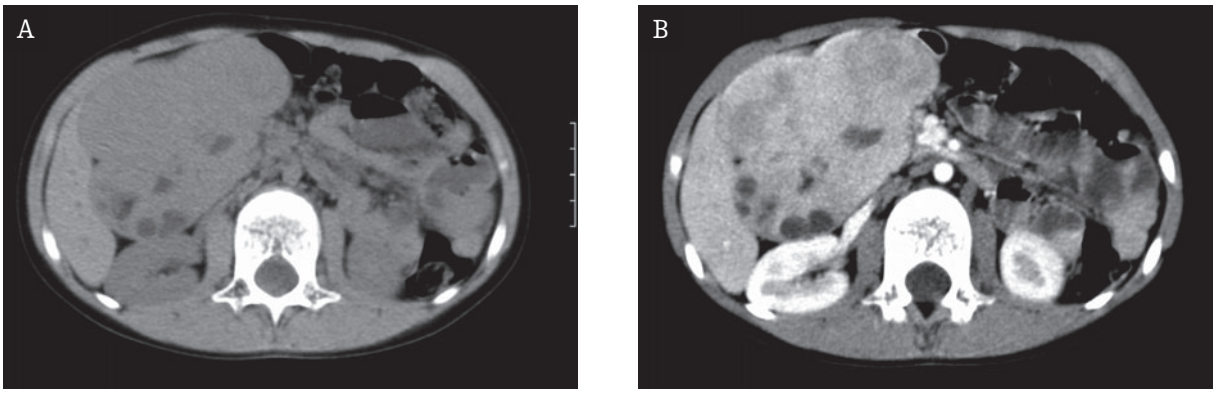

Figure 2. Boy MM. Abdominal CT study before and after contrast medium administration. A polycyclic, non-homogenous, heterogeneously enhancing, well-delineated tumor below the liver hilus.

of the tumor and stage of the process. Determination of tumor markers levels allows to classify the patients into the standard (AFP < 15 000 IU/ml) or high risk group (AFP > 15 000 IU/ml). Detection of metastatic lesions qualifies the patient to the high risk group [19]. The applicable therapeutic protocol specifies the indications for postoperative chemotherapy in standard and high risk tumors. The role of radiotherapy in cases of pure ovarian dysgerminoma has not been established unequivocally [17].

Case reports

Case 1

A 6-year-old boy (M.M.) born from twin pregnancy (initially diagnosed as triplets) by cesarean section in 26th week of gestation with birth weight of 1030 g, was seen by a pediatrician with fever up to 39.5 °C. Anamnesis revealed previous subfebrile states. The pediatrician observed hepatomegaly and ordered USG, which revealed a tumor of the liver. The boy was referred to the Department of Oncology, Hematology and Chemotherapy of Upper Silesian Child and Mother Health Center in Katowice. Physical examination on admission revealed painless palpable mass in the right epi- and mesogastrium, extending down 2 cm below the umbilicus. CT visualized a polycyclic 7.8 x 9.4 x 6.3 cm mass at the level of the hepatic hilus (fig. 1) of heterogeneous density, enhanced inhomogeneously after intravenous contrast administration to 70-100 HU values. The posterior part of the tumor (fig. 2A) contained hypodense areas (ca. 10 HU) up to 1.5 cm, which showed no contrast enhancement (fig. 2B). The tumor caused deformation of the hepatic vessels and the inferior vena cava. The intrahepatic bile ducts and the gallbladder were normal.

Differential diagnosis took into consideration lymphomas, teratoma and PNET type tumors. Laboratory investigations revealed a modest increase of AFP level. The boy was qualified for surgical treatment. An extensive tumor of the right retroperitoneal space was found intraoperatively. The operation was non-radical. Histopathological investigations of the tumor tissue resulted in the diagnosis of immature teratoma *(teratoma immaturum)* with predominant neuroendocrine carcinoma (*Ca neuroendocrinale*) structure.

Case 2

A 9-year-old girl (W.E.), born at term after natural labor, with birth weight of 3100 g, was referred to the Emergency Department of the Upper Silesian Child and Mother Health Center in Katowice with abdominal pain and fever - 38°C. Physical examination revealed presence of a large tumor over the pubic symphysis, which *raised and extended the abdominal wall*. In USG, two tumors of inhomogeneous echogenicity and dimensions: the first 10 x 7 x 5,2 cm, the

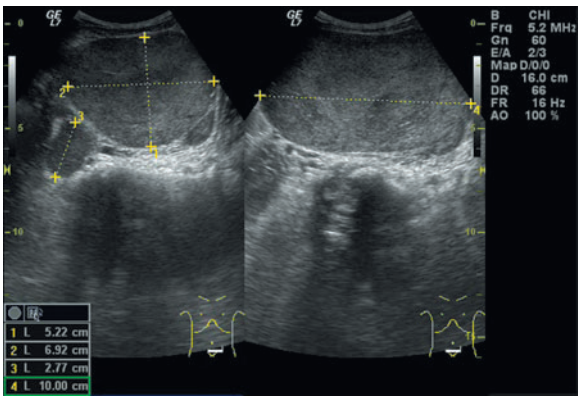

Figure 3. Girl WE. Transabdominal sonography of the pelvis – a well-delineated, solid, non-homogeneous tumor with linear hyperechogenic shadows. A second smaller tumor in anatomical placement of the right ovary. The uterus compressed, slightly displaced to the left.

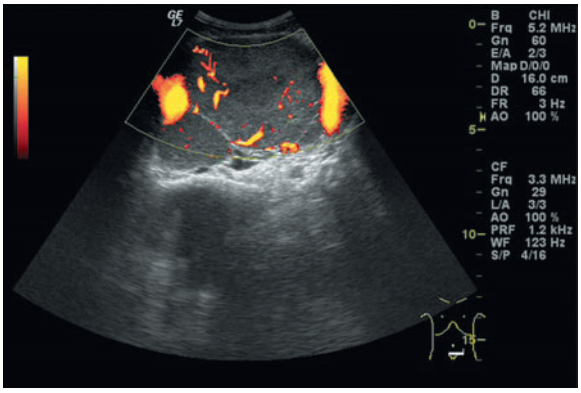

Figure 4. Girl WE. Transabdominal sonography of the pelvis – Power Doppler – blood vessels in the tumor.

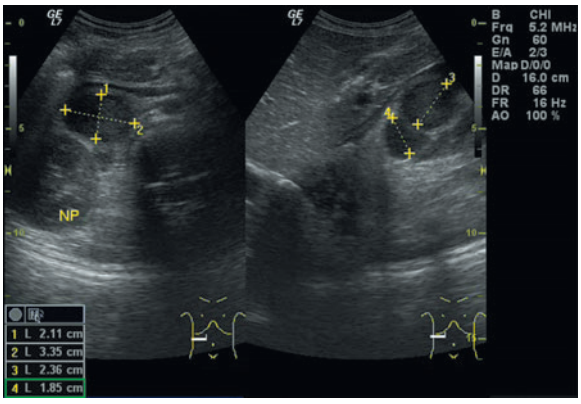

Figure 5. Girl WE. Transabdominal sonography – numerous, hypoechogenic tumors situated in the paravertebral region.

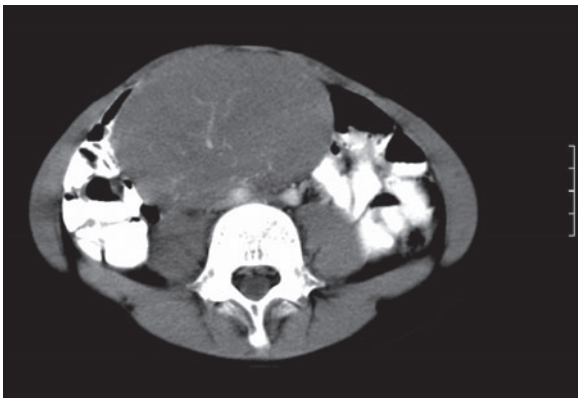

Figure 6. Girl WE. Abdominal CT study after contrast medium administration – a solid tumor of the hypogastrium with „network" enhancement.

second 3 cm in diameter, were seen in the pelvis. (fig. 3). Power Doppler sonography demonstrated blood flow within the larger tumor (fig. 4). Additionally, presence of numerous solid, round tumors up to 5 cm in diameter in retroperitoneal space below the origin of the celiac trunk was observed (fig. 5).

In CT, the pelvic tumors showed ca. 40 HU density. After intravenous contrast administration, they enhanced with a delicate reticular pattern (fig. 6). Tissue density of the mesogastrial tumors ranged from 20 to 40 HU, and after contrast administration showed weak contrast enhancement. One of the tumors demonstrated signs of disintegration and enhanced only marginally (fig. 7). Both imaging modalities did not reveal infiltration of other organs, but only compression of uterus and deformation of urinary bladder. The level of tumor markers was within normal limits.

In differential diagnostics the possibility of lymphoma, embryonal neuroma and PNET type tumor was taken into consideration.

Surgical treatment involved resection of the tumor of the right ovary together with a number of smaller, cyst-like lesions (fig. 8). Histopathologic examination revealed a dysgerminoma; the character of cyst-like lesions was not determined unequivocally. They showed no signs of malignancy. Spontaneous regression of cysts not removed during the surgery was observed in postoperative follow-up.

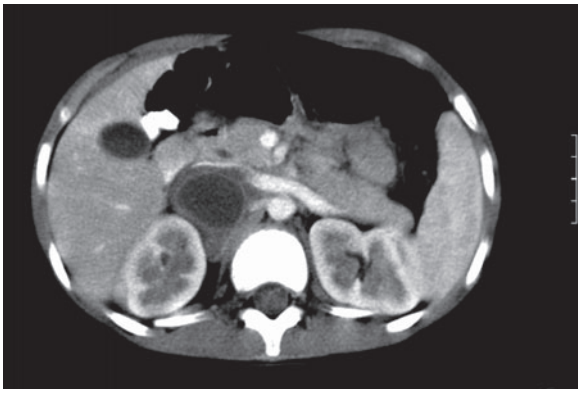

Figure 7. Girl WE. Abdominal CT study after contrast medium administration. Hypodense, cystic, peripherally enhancing tumor situated in the right paravertebral region.

Discussion

The abdominal tumors detected clinically may result from developmental anomalies, inflammatory lesions and neoplastic processes. The latter can be benign or malignant. Germ cell tumors are most frequantly localized in the gonads. Only 1-2.5% of germ cell tumors is primarily localized in the retroperitoneal space [5].

Calcifications are seen in less than 50% of cases. In two cases of germ cell tumors described above no calcifications were observed. In one case (girl W.E.) a necrotic area was present. The symptoms of abdominal and pelvis minor tumors in children are often nonspecific, which is confirmed by the reported cases.

Differential diagnostics of our patients took into consideration lymphoma, tumors of embryonal origin, and PNET. Abdominal lymphomas can be located in lymph nodes

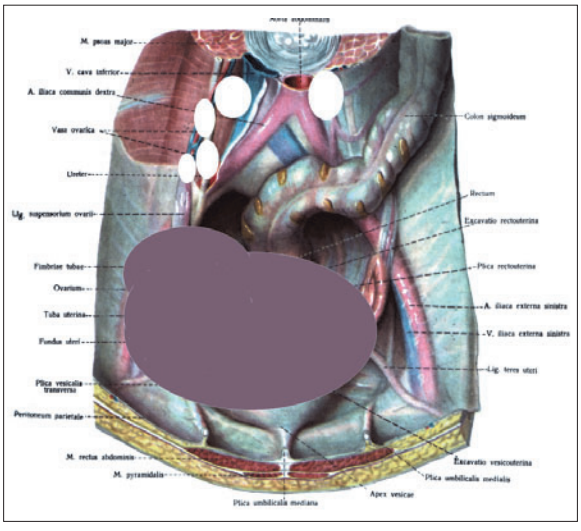

Figure 8. Girl WE. Postoperative scheme with locations of numerous tumors in the abdominal cavity.

or abdominal organs. In lymphadenopathy, a group of enlarged lymph nodes is visualized as well-delineated, usually homogeneous tumor-like lesions, showing weak contrast enhancement. Organic lesions may be focal or disseminated (diffuse) in character, localized in the liver, spleen, kidneys, adrenal glands, gastrointestinal tract [18]. Unlike lymphomas, PNET type tumors can be inhomogeneous [11]. In the reported cases, the detected tumors were inhomogeneous in CT scans (fig. 1, 2B, 7).

In boy M.M., the tumor in the hepatic hilus with polycyclic outline and inhomogeneous density caused problems with determination of the site of origin. Despite considerable mass effect, no distension of the bile ducts was observed, that is why we took into consideration germinal tumors, lymphomas and PNET type tumors in differential diagnosis. The pelvic tumor with reticular contrast enhancement suggested the diagnosis of dysgerminoma [1]. The presence of additional nonspecific tumors in the vicinity of the spinal column prompted us to consider also the possibility of a lymphoma, PNET and embryonic neuroma in differential diagnostics.

The reason for presentation of the two cases were diagnostic doubts.

Because of their nature, extragonadal germ cell tumors (originating from 3 primordial cell layers) develop in the body axis. In this case, right-sided location of the abdominal tumor suggested the presence of more common systemic diseases, embryonic neuroma or PNET. Suggestions from clinical examination could correspond to a primary hepatic tumor. Despite detailed imaging, only histopathology allowed to establish the ultimate diagnosis of immature teratoma.

In the second case, we observed extensive involvement of the retroperitoneal space with tumors arranged along the large abdominal and iliac blood vessels. Their character in CT imaging suggested solid lesions – lymphadenopathy. Simultaneously, the presence of a large mass in the pelvis minor corresponded best to advanced systemic process. In that case, intraoperative findings elucidated the original site of the lesion – the right ovary, the suspected solid tumors turned out to be retroperitoneal cysts. Histopathology led to the diagnosis of dysgerminoma of the right ovary, with no signs of neoplastic process in the observed cyst-like lesions. The fact of spontaneous regression of cysts not resected during the surgery is worth emphasizing.

Conclusions

1. Imaging diagnostics in case of clinical suspicion of an abdominal tumor, including germ cell tumors, should begin with USG. This modality provides good visualization of the tumor and allows to plan further diagnostics and differentiation.
2. CT demonstrates more precisely the character of the tumor (solid, cyst-like, mixed), presence of calcifications and relations with adjacent organs: infiltration, compression, displacement and involvement of other structures, including lymph nodes. Reaction to intravenous contrast administration allows to determine vascularization pattern of the tumor.
3. Replacement CT with MR, especially for monitoring of treatment results, should be considered. For the time being, the number of MR scans is limited by insufficient availability of MR units.
4. The ultimate diagnosis of tumor type must be established on the basis of histopathological investigations.

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