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YOLK SAC TUMOR OF THE OVARY

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Background: A 10-year-old girl presented to the emergency department with acute abdominal pain in the right fossa. There was no known previous medical history. She had a fever up to 38,4°C without any complaints of nausea, vomiting or diarrhea.
Clinical investigation showed a swelling of the lower abdomen, present since 4 months, a positive McBurney sign and muscular defence. Laboratory findings showed an elevated C-reactive protein of 11,4 mg/dL (normal value 0-0,5 mg/dL) and leukocytosis.

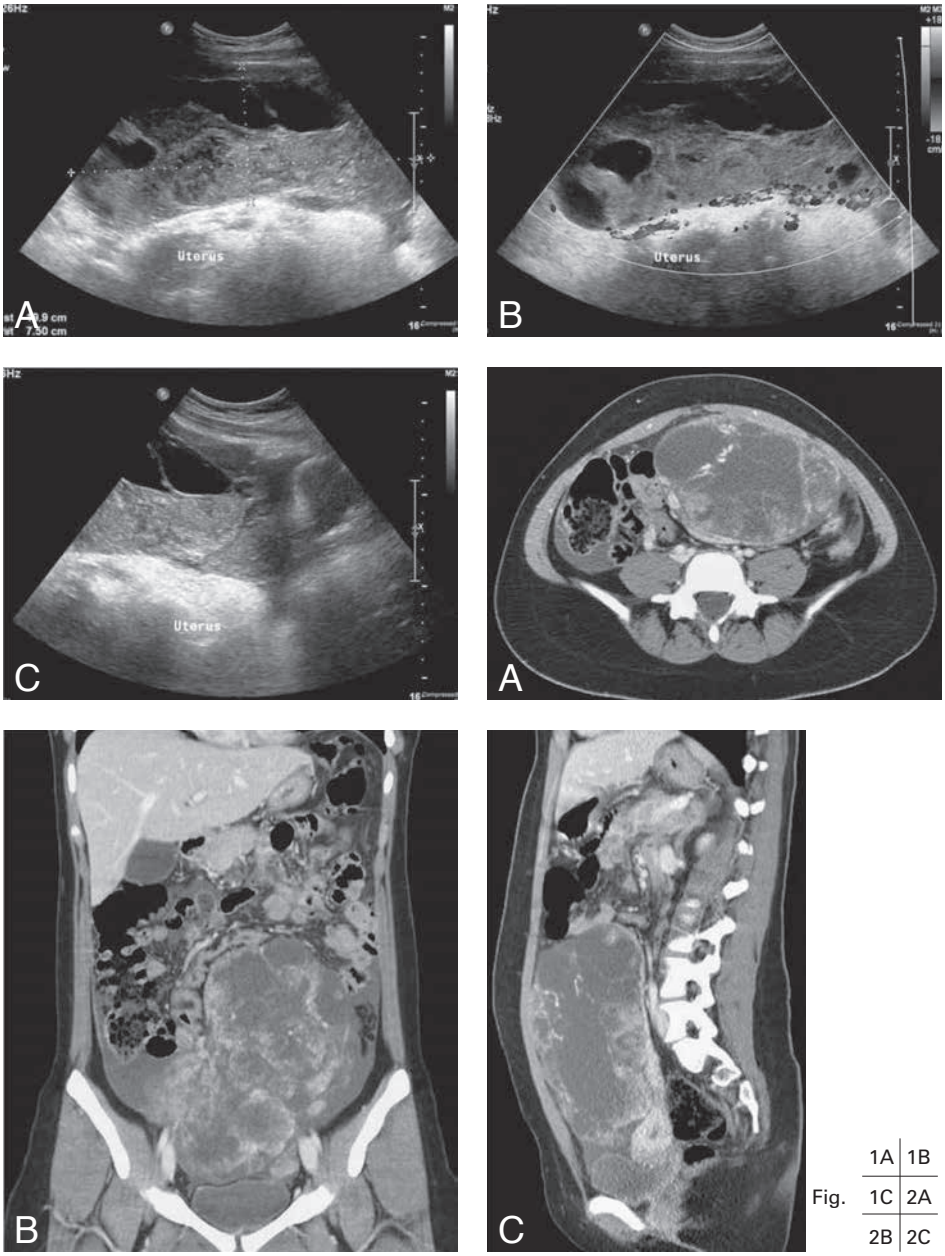

Fig. 1A 1B / 1C 2A / 2B 2C

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Work-up

Ultrasonography of the abdomen (Fig. 1) shows on A (sagittal section on the midline) a large mass with a cystic as well as a solid components. On color–Doppler image (B), increased vascularisation, especially on the edges of the lesion, is observed. On sagittal section through the lower part of the mass (C), a clear separation between the mass and the uterus is shown. Hence the mass is presumed to have on ovarian origin.

Contrast-enhanced CT scan of the abdomen (Fig. 2, A: axial section, B: reformatted image in the coronal plane, C: reformatted image in the sagittal plane) demonstrates a complex ovarian mass with displacement of the uterus to the left fossa. The tumor is predominantly cystic but also contains a solid component, which shows contrast enhancement in the solid tumor component.

In between the cysts are large vessels visible (best seen on the axial section (A)).

Radiological diagnosis

Based on the imaging findings, the diagnosis of *yolk sac tumor of the ovary* was suggested.

This diagnosis was confirmed by histopathological examination following surgery.

Discussion

The yolk sac tumor (YST), also known as an endodermal sinus tumor is a rare malignant germ cell tumor. Tumors from germ cell origin represent 15-20% of all ovarian masses, and are malignant in less than 5% of the cases. A YST is the second most common malignant germ cell tumor and counts for only 1% of the malignant ovarian masses.

YST mostly occur in the 2nd and 3rd decades of life and are only rarely seen in patients over 40 years of age. In about 50% the tumor is limited to one ovary, stage II shows pelvic spread (10%) and in 40% of cases metastases are present.

In most cases there are no clinical symptoms. Abdominal swelling may be present in case of rapid growth and can be very pronounced at the time of diagnosis. If complications, such as hydronephrosis or ovarian torsion are present, more acute symptoms can occur as seen in the presented case.

The imaging findings of YST may vary between entirely solid to a predominantly cystic mass. There are also reports of a mixed type tumor with solid as well as cystic components. The cystic components of this tumor are often due to necrosis or cystic degeneration.

This composition determines the ultrasonographic appearance, in the presented case due to the mixed composition, as a large ovarian mass with solid as well as cystic components. The solid components are often seen in the periphery of the mass. As an endodermal sinus tumor is highly vascularized color Doppler flow can show many small arterioles with a low RI value. The cystic components of the tumor have the same echogenicity as the urine in the urinary bladder.

Contrast-enhanced CT examination shows a large mass with well-enhancing solid components and also a cystic or necrotic portion. Large vessels are present in almost all cases of a yolk sac tumor. This phenomenon is described as the 'bright dot sign'.

Because large areas of hemorrhage and necrosis are common findings in yolk sac tumors and by its high capability in demonstration of hemorrhage, MRI might be useful in the diagnosis. T1-weighted images may show several high intensity spots within the solid portion indicating small hemorrhages. As these hemorrhages can be subtle, they can be missed on ultrasonography. Microscopic studies reveal stromal edema, which might explain the high signal intensity on T2-weighted images and the prominent enhancement after administration of Gadolinium. A third finding on MR-images may be the presence of signal voids as the tumor has a rich vascular supply.

Microscopic examination of a YST reveal multiple patterns, with a predominance of reticular honeycombed structure of communicating spaces lined by primitive cells. Shiller-Duval bodies, resembling the endodermal sinus of rodent placenta, are also present. The stroma of the tumor is often edematous with zones of necrosis and hemorrhage.

In imaging studies it is important to distinguish the yolk sac tumor from a sclerosing stromal tumor. A sclerosing tumor shows a strong enhancement compared to the uterine myometrium. The rim of hypo-intensity on T2-weighted images and the presence of low intensity nodules set against high-intensity stroma on T2-weighted images may help to distinguish a sclerosing stromal tumor from a yolk sac tumor. The key diagnosis of a yolk sac tumor is seen in the laboratory results with an elevated alfa-fetoprotein (AFP) level. In the presented case the AFP level was initially 36300 µg/l (normal value 0-12 µg/l) and rapidly decreased to 20667 µg/l after surgery.

The recommended treatment is initially surgical with unilateral salpingo-oophorectomy, with limited debulking of extra ovarian tumor, followed by chemotherapy.

Bibliography

1. Hyuck J.C., Min H.M., Seung H.K., *et al.*: Yolk sac tumor of the ovary: CT findings. *Abdom Imaging*, 2008, 33: 736-739.
2. Seung E.F., Fae M.L., Sung E.R., *et al.*: CT and MR Imaging of ovarian tumors with emphasis on differential diagnosis. *Radiographics*, 2002, 22: 1305-1325.
3. Toshihide Y., Kaori T., Takashi K., et al.: Yolk Sac Tumor of the ovary: radiologic-pathologic correlation in four cases. *J Comput Assist Tomography*, 2010, 24: 605-609.