Article received on August 12, 2017 and accepted for publishing on September 10, 2017.

Yolk sac tumor in a 20 years old patient

Ioana A. Negoiță[1], Bogdan Panaite[1,2], Ovidiu V. Nicodin[1,2], Dimitrie Nanu[2,3]

Abstract: *Yolk sac tumor or endodermal sinus tumor is a rare malignant ovarian germ cell tumor (is a borderline tumor) diagnosed in young females up to 25 years old. Histopathological it resembles the mesenchyme of the primitive yolk sac. Microscopically it presents the following triad: Schiller-Duval bodies, reticular aspect and PAS+ hyaline droplets (α-fetoprotein).*

The diagnosis of Yolk sac tumor is made by dosing serum α-fetoprotein, ultrasound and MRI – DWI imaging.

It requires surgical treatment, followed by chemotherapy (new therapies – platin or carboplatin, etoposide, bleomycin) [1]. Survival prognosis at 5 years is of 80% for stage I.

Differential diagnosis is with Brenner tumor, ovarian clear cell carcinoma, dysgerminoma, malignant teratomas, androblastoma, dermoid cyst.

Keywords: *Yolk sac tumor, AFP, MRI, DWI, abdominal-pelvic pain*

INTRODUCTION

Yolk sac tumor (endodermal sinus tumor) is an ovarian germ-like, extremely rare, (borderline) malignant tumor, which occurs very often at ages < 25 years. Schiller-Duval glomeruloid bodies are specific [2]. They are composed of a papilla with a conjunctive-vascular axis, delimited by round cells; the papilla predominates within a cyst delimited by embryonic tumor cells.

[1] Carol Davila University Central Emergency Military Hospital, Bucharest, Romania

[2] Titu Maiorescu University, Faculty of Medicine, Bucharest, Romania

[3] Saint John Emergency Clinical Hospital, Bucharest

CASE PRESENTATION

The 20 years old patient, R.S., from rural area, is admitted within our service for abdominal-pelvic pain, predominately on the right flank, with onset of approximately 7 days and with progressive intensification. The patient is nulliparous, nuligest, with irregular menstrual cycles, of normal quantity. From her pathological personal history, we retain appendectomy in 2012.

The general objective examination upon admission reveals a patient of good general state, afebrile, of normoponderal condition. The local objective exam reveals a slightly enlarged volume of the abdomen, mobile with breathing, painful spontaneously and upon palpation, predominantly in the right flank, with no abdominal guarding; nulliparous cervix, in axis, closed external cervical orifice, without macroscopic modifications, uterus in anteversoflexion position, of normal consistency and volume, left adnexal area slightly sensible upon palpation, right adnexal area modified by presence of a tumor with an approx. 20 cm diameter. There is no liquid in the Pouch of Douglas.

Biological tests, EKG, pulmonary radioscopy do not

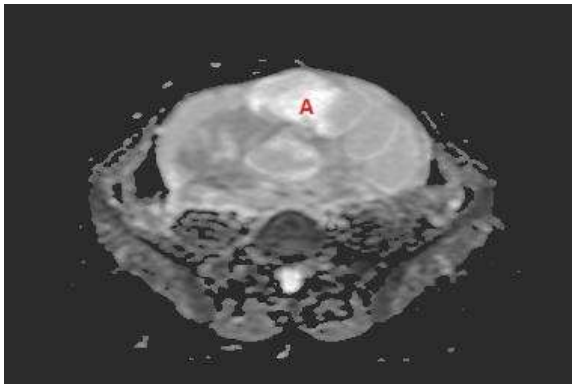
show pathological changes.

Transvaginal ultrasound shows uterus in ante-versoflexion, with diameters of 45/38/42 mm, smooth contour, homogeneous myometrium, linear cavity, endometrium of 3 mm. Voluminous abdominal-pelvic mass extended cranial-caudally on approximately 19 cm and axially on 18 cm, transonic, multi-septated, including hypo-/hyperechoic heterogeneous confluent areas, especially in the caudal portion. There is contact with the uterine wall, without demarcation line with the adnexal areas. Thin layer of fluid in the Pouch of Douglas.

The high-performance imaging examination – MRI with diffusion weighted images (DWI) shows:

- The mass presents areas of restricted diffusion, with hyper-intense aspect on the ADC map (Figure 1), suggestive of benign aspect.

Figure 1: ADC map

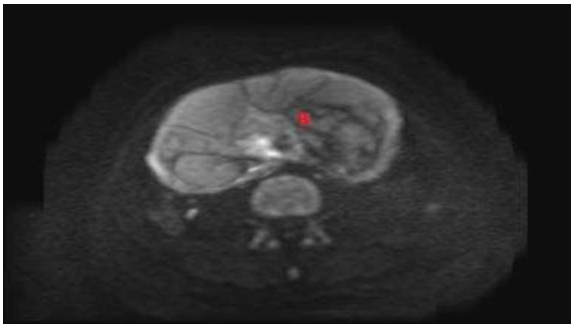


The mass is well delimitated, with predominantly hyper-signal T2-weighted, presents small areas of restricted diffusion (A), with hypo intense aspect on the ADC map (B), suggestive of "malignant foci" within a larger tumor.

- Small areas of restricted diffusion can be appreciated on the DWI image (Figure 2).

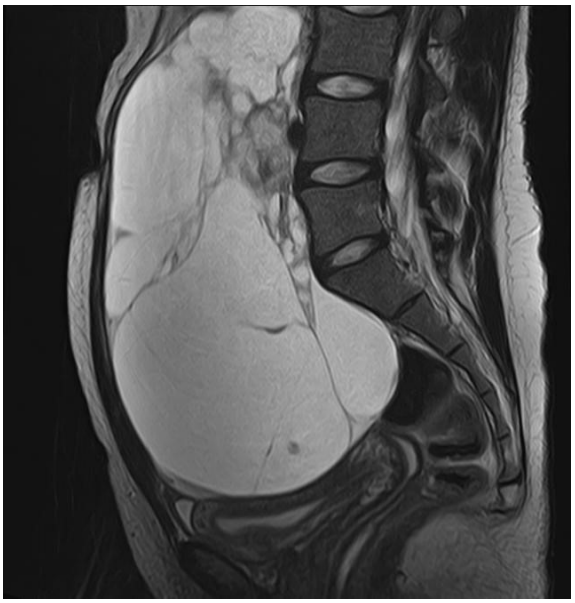
The native and contrast-enhanced abdominal MRI examination shows voluminous cystic multiloculated tumoral mass, with abdominal-pelvic development, of 21.5/18.3/7.9 cm maximum diameters, with adnexa as starting point, connecting both adnexa, including multiple solid areas, with gadolinium uptake and which is developed superiorly up to the inferior renal third, manifesting mass effect on the adjacent structures, to which it maintains the interface of demarcation.

Figure 2: DWI measurement



Uterus of normal size, net, regulated contours and homogeneous structure. There is no sign of abdominal-pelvic adenopathies. Minimum peri-hepatic ascites and in the paracolic gutters. Liver, cholecyst, spleen, kidneys, pancreas with morphology and MRI signal within normal limits.

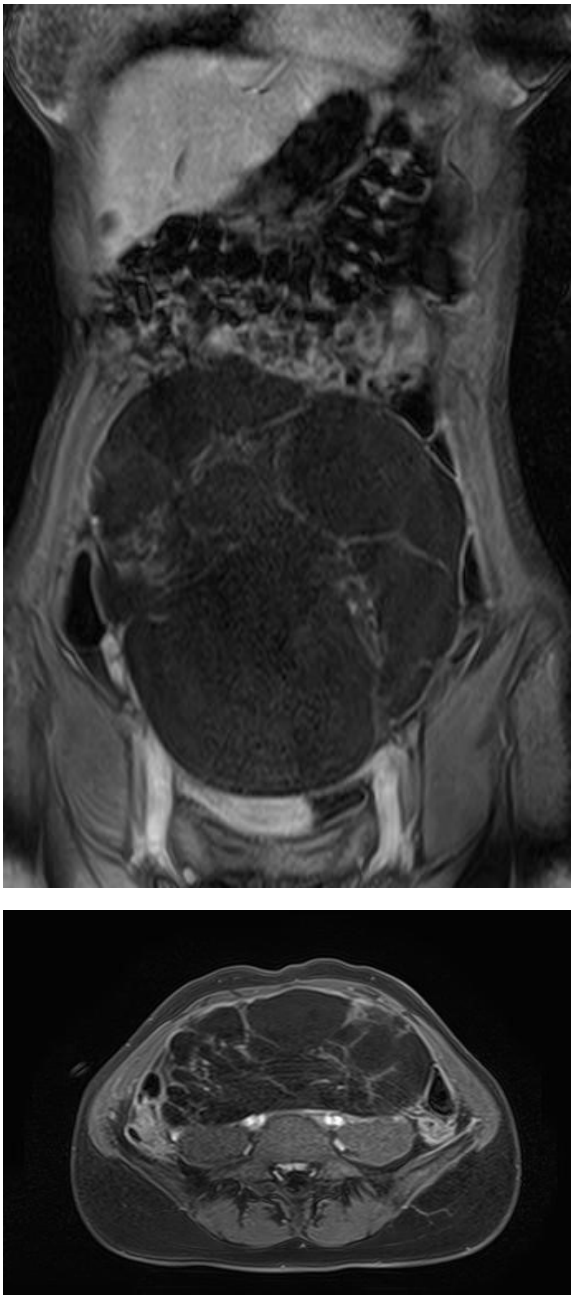
Figure 3: T2-weighted image in sagittal plane. Voluminous abdominal-pelvic mass. Sagittal T2-weighted image with predominantly liquid signal, multiseptate, including solid areas.



After a proper pre-surgical preparation, surgery is performed under general anesthesia, with orotracheal intubation. Upon exploration serosanguineous fluid of approximately 300 ml is found, which is

collected and sent for HP examination. Right adnexa with tumoral transformation, of approximately 20/19 cm, of mixed consistency, with multiple septa, presents on its surface two continuity solutions at capsule level, by which serous, citrine fluid is exteriorized.

Figure 4: T1 FS weighted images – MRI; T1 FS weighted images following intravenous administration of gadolinium contrast shows parietal enhancement also at the level of internal septa.



Left ovary with tumoral transformation, by presence of a 2/5 cm cyst, of serous content. Uterus of normal macroscopic aspect. Right adnexectomy, left cystectomy are performed, as well as hemostasis control, drainage, and parietoraphy in anatomical layers. The evolution following the surgery was favorable under antibiotic, antalgic, anticoagulant treatment, with discharge on the fourth day following the surgery.

Figure 5: Right adnexal tumor of approximately 20/19 cm, of mixed consistency, with multiple septa, presents on its surface two solutions of continuity at capsule level.

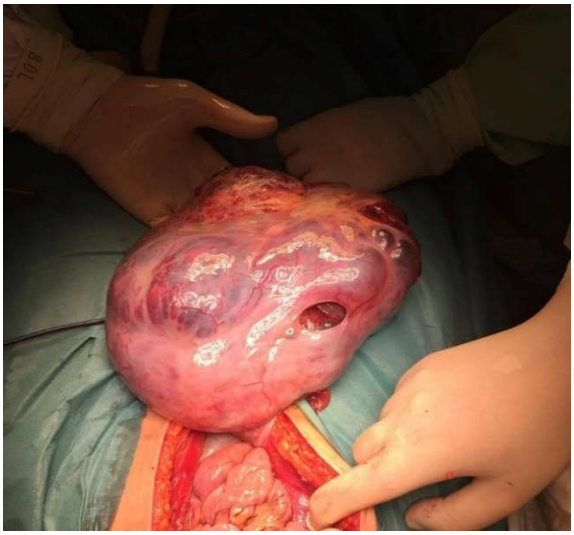
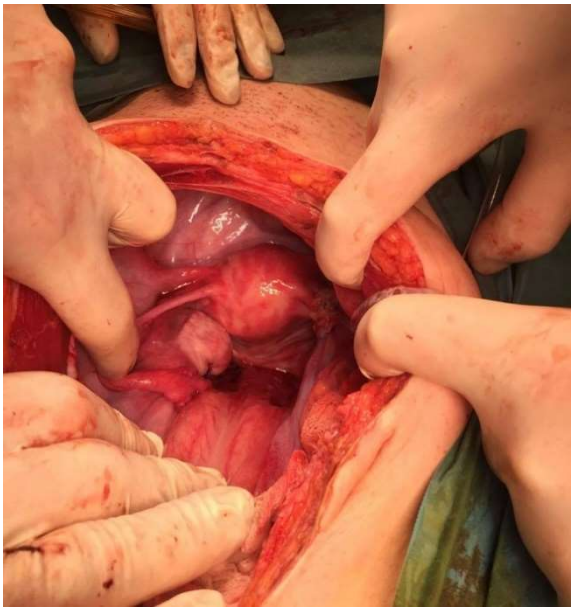


Figure 6: Intraoperative image following right adnexectomy and left cystectomy



The histopathological examination of the macroscopic piece shows a solid mass of 20/18/6 cm, with cystic areas, necrotic and hemorrhagic areas. The microscopic examination on the piece shows the following: tumoral mass consisting in linear channels of atypical epithelial cells, islands with clear cells and aspect of Schiller-Duval bodies, myxoid hypocellular stroma and areas of tumoral necrosis; aspects suggestive of Yolk tumor; salpinges without tumoral infiltrates.

Immunohistochemical study is recommended for confirmation and differential diagnosis with ovarian clear cell carcinoma. Upon immunohistochemical study the diagnosis was of Yolk sac tumor.

DISCUSSIONS

Yolk sac tumor is also known as endodermal sinus tumor and represents an ovarian germ-like tumor. It is an extremely rare malignant tumor that appears predominantly in the second decade of life. Yolk sac tumor may occasionally arise from a pre-existing dermoid cyst. An increased value of AFP can be relevant for this diagnosis. Symptomatology is contoured by intense abdominal-pelvic pain.

Bilateral presence of this tumor is extremely rare. It is described as a pelvic mass that can also be extended abdominally, is complex and contains both solid and cystic portions. Cystic areas are often composed of epithelial cysts produced by the tumor or by coexisting mature teratomas.

Ultrasound in Yolk sac tumor describes echoic and hypoechoic areas.

Magnetic resonance imaging demonstrate on DWI sequences, presents small areas of restricted diffusion, with hypo-intense aspect on the ADC map, suggestive of "malignant foci" within a larger tumor [4]

Yolk sac tumor increases quite rapidly in size, the symptomatology being especially described in intense abdominal-pelvic pain. Lymphatic dissemination may occur early in some cases.

The treatment is surgical, followed by chemotherapy (platinum or carboplatin, bleomycin, etoposide)..

CONCLUSIONS

There are difficulties in the diagnosis and surgical staging of ovarian tumors of low malignant potential. AFP utility together with ultrasound examination and magnetic resonance imaging with DWI sequence helps in reaching an accurate diagnosis in Yolk sac tumoral masses.

Being a borderline tumor, a prompt diagnosis represents a success in the long-term prognosis of the disease. Transabdominal surgical approach is of choice in endodermal sinus tumors.

The success of subsequent therapy is largely determined by the initial surgical procedure. Surgical treatment is the therapeutic method of first choice, which can lead to healing in the early stages I-a and I-b and which decisively influences the results of complementary treatments, chemo- and radio-therapy.

References:

1. Motegi M, Takakura S, Takano H, Tanaka T, Ochiai K (February 2007). "Adjuvant chemotherapy in a pregnant woman with endodermal sinus tumor of the ovary". Obstet Gynecol. 109 (2 Pt2): 537–40.

2. Kumar, Abbas, Fausto. Pathologic Basis of Disease, 7th edition. Philadelphia; Elsevier-Saunders, 2005. 1101.

3. Levitin A, Haller KD, Cohen HL et-al. Endodermal sinus tumor of the ovary: imaging evaluation. AJR Am J Roentgenol. 1996;167 (3): 791-3. AJR Am J Roentgenol (abstract) -Pubmed citation

4. Levine D, Kruskal JB. Overview of use of magnetic resonance imaging in obstetrics and gynecology. Updated September 2013. Available at: http:\\www.uptodate.com. Accessed on September 15, 2013.