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Refractory Ascites Revealing an Ovarian Yolk Sac Tumor with Intraperitoneal Rupture

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Abstract

Yolk sac tumors of the ovary are rare entities that account for 2% - 5% of all ovarian tumors. They represent the second most common histological variant of malignant germ cell tumors of the ovary after dysgerminomas. Yolk sac tumors are most commonly encountered in women in the second and third decades. Microscopically, they are highly polymorphic and can present in a pure form or associated with another contingent of germ cell tumor. We report the case of a 26-year-old woman, who underwent surgery for a large right ovarian tumor rupturing into the peritoneal cavity. The ovarian tumor was revealed by ascites of great abundance and abdomino-pelvic pain. On histological examination, the diagnosis of yolk sac tumor in its pure and polyvesicular vitelline pattern was made. Through this observation, we propose to discuss the anatomoclinical particularities of these tumors by emphasizing the importance of histology for the diagnosis as well as the need of an early and appropriate management.

Keywords

Germ Cell Tumor, Yolk Sac Tumor, Ovary, Abdominal Distension, Schiller-Duval, Ascite

1. Introduction

Ovarian yolk sac tumors are defined as malignant ovarian germ cell tumors that account for only 2% - 5% of all ovarian tumors and less than 5% of all germ cell tumors [1]. They are mostly seen in adolescents and young women [2].

Yolk sac tumors were initially discovered by Schiller in 1939, who suggested a mesonephroid origin. A few years later, Teilum reclassified these tumors and described their extraembryonal germ cell origin. He designated them as endodermal sinus tumors, which were termed later "Yolk sac tumor" due to their similarities with the extraembryonal yolk sac and vitelline structures [2] [3].

These tumors are rapidly growing and highly aggressive. Quality of diagnosis process and treatment implementation largely determines the prognosis [1] [2] [3].

Herein, we report a case of ovarian yolk sac tumor with intraperitoneal rupture and refractory ascites.

Our purpose was to highlight through this case, the appropriate diagnostic and management procedures in order to improve the prognosis.

2. Observation

A 26-year-old woman, nulliparous, with no other particular medical history, presented with abdomino-pelvic heaviness and pain, radiating to the right, evolving for 2 weeks and associated with an increased abdominal volume. On physical examination, the patient was in good general condition and subfebrile (37.8˚C). Abdominal distension and tenderness in the right iliac fossa have been noticed. The abdomino-pelvic ultrasound revealed an ovarian mass of heterogeneous components, measuring 19 cm along its long axis and associated with abundant peritoneal effusion (**Figure 1**). The contralateral ovary was normal in appearance. AFP and CA-125 serum levels were elevated respectively to 19,206 ng/ml and 619 UI/l. The beta-hCG level as well as the rest of the laboratory testing was normal. The abdominopelvic CT scan showed a large abdominopelvic mass, originating from the right ovary, associated with diffuse and abundant intraperitoneal effusion (**Figure 2**). The contralateral ovary and surrounding organs were normal. Neither adenopathy nor lesion suggestive of peritoneal carcinomatosis was objectified. An evacuating puncture was performed and 2.5 l of yellow citrine ascites were removed. Corresponding cytologic analysis demonstrated non-specific cellular population, essentially composed of few macrophages and mesothelial cells without neoplastic cells. Due to the rapid recurrence of ascites and after a multidisciplinary meeting, a midline laparotomy with right adnexectomy was performed for anatomopathological examination. On macroscopic examination, the specimen showed a focally ruptured right ovarian tumor, measuring 19 × 15 × 6.5 cm with a smooth grayish—brown surface. Cut sections were heterogeneous with gelatinous, myxoid and necrotic—hemorrhagic areas. Cystic spaces with honeycomb patterns were also found (**Figure 3**). Histopathological examination (**Figure 4**) demonstrated a neoplastic cell proliferation. Most of tumoral cells have clear cytoplasm and atypical nuclei with quite numerous mitotic figures and prominent nucleoli. They usually form anastomosing channels and are often arranged around vascular structures to form the pathognomonic Schiller-Duval bodies. Cystic cavities separated by fibrous stroma and lined by

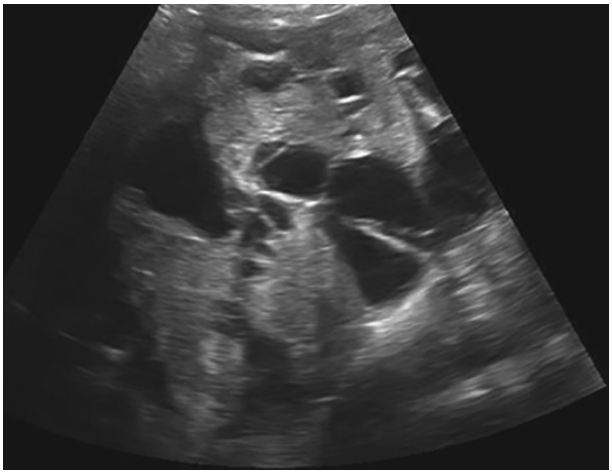

Figure 1. Ovarian mass with heterogeneous appearance on ultrasound. Source: Radiology Department, CHU Andrainjato Fianarantsoa, Madagascar.

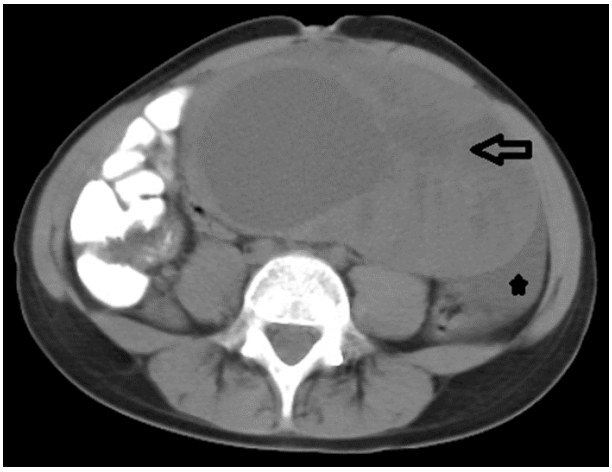

Figure 2. Computed tomography axial slice: ovarian mass (arrow) with ascites (star). Source: Radiology Department, CHU Andrainjato Fianarantsoa, Madagascar.

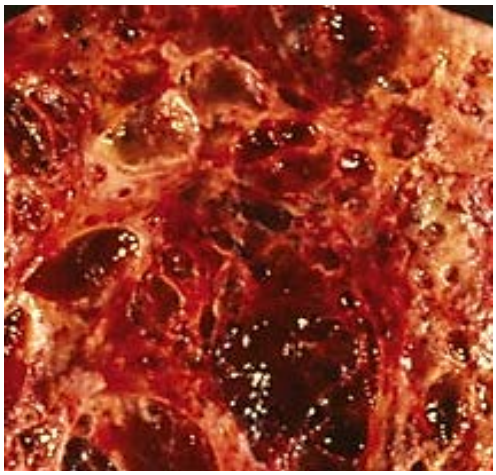

Figure 3. Ovarian tumor: Solid and cystic areas with honeycumb appearance. Gelatinous changes and areas of hemorrhage and necrosis are also present. Source: Pathology Department, CHU Andrainjato Fianarantsoa, Madagascar.

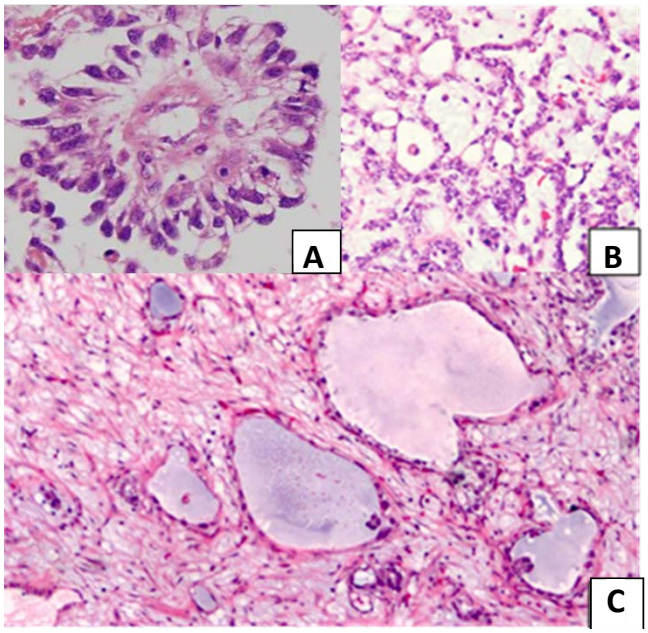

Figure 4. Ovarian yolk sac tumor with polyvesicular vitelline pattern. A, Schiller-Duval body; B, Anastomosing channel patterns; C, Dilated channels with flattened or cuboidal epithelium. Source: Pathology Department, CHU Andrainjato Fianarantsoa, Madagascar.

flattened or cuboidal epithelium were also seen. The area of rupture was covered with a fibrous cap. Hemorrhagic and necrotic changes were found as well as myxoid areas. The fallopian tube appeared normal. Epiploic and peritoneal biopsies were also performed and did not demonstrate metastatic localization. The diagnosis of yolk sac tumor in its pure form with polyvesicular vitelline pattern was made. Immunohistochemical examination revealed strong expression of AFP antigen in tumor cells, confirming the diagnosis (**Figure 5**). Postoperative outcomes were favorable with no evidence of recurrent ascites and a progressive decrease in the serum AFP level. The patient had received adjuvant chemotherapy with BEP (bleomycin, etoposide, cisplatin). Six months after the last chemotherapy course, serum AFP level was 5 ng/ml, abdominopelvic CT-scan was normal and the patient recuperated a regular menstrual cycle. Besides, chemotherapy related toxicity was not found and she has been free of recurrence for 2 years.

3. Discussion

Yolk sac tumors of the ovary are rare tumors that usually occur in young women 20 - 30 years old [1] [2] [3]. They represent the second most common histologic subtype of malignant ovarian germ cell tumor after dysgerminomas [4]. These tumors are rapidly growing and clinical symptoms mainly include pelvic pain with an enlarging abdomino-pelvic mass [5]. They can also be revealed by ascites of variable abundance, secondary to torsion or intraperitoneal rupture [1]. Intraperitoneal rupture generally occurs in 30% of cases and can be pre- or intraoperative. In addition, associated peritonitis can also be observed, secondary to ascitic

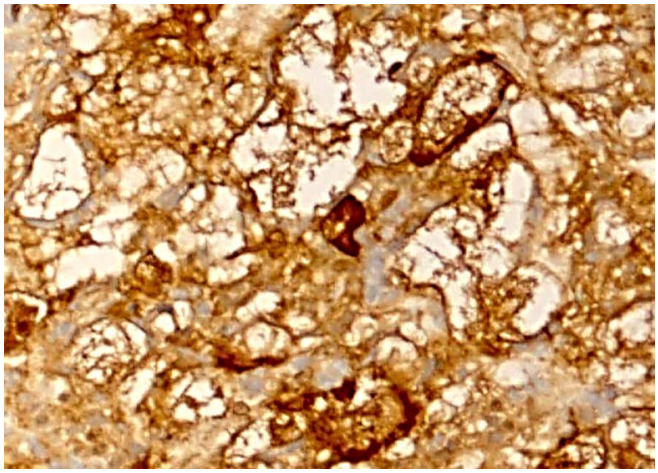

Figure 5. Strong expression of AFP antigen in tumor cells. Source: Pathology Department, CHU Andrainjato Fianarantsoa, Madagascar.

fluid infection [6] [7]. In our case, the patient presented with acute pelvic pain, abdomino-pelvic mass and refractory ascites of great abundance, due to tumor rupture. This tumor rupture, probably preoperative, was confirmed on macroscopic examination of the specimen. Imaging tests could not confirm or dismiss the diagnosis of Yolk sac tumor [1] [2]. Ultrasound characterizes the adnexal mass and shows the presence of ascites or hepatic metastases. CT scan permits detection of peritoneal carcinomatosis or adenopathy [7] [8]. MRI findings are often non-specific, characterized by hypervascular and hemorrhagic appearance [8] [9]. For our patient, imaging examination have demonstrated an abdomino-pelvic tumor, of right ovarian origin, with heterogeneous contents, strongly suspicious of malignancy. Imaging also revealed the presence of ascites and the absence of peritoneal carcinomatosis or tumoral extension to adjacent organs. The diagnosis of yolk sac tumor is essentially based on histological examination [1]-[6]. Macroscopically, the ovarian mass is encapsulated and ranged in size from 3 to 30 cm. The tumor is usually round, oval or globular with smooth and glistening surface that may be firm or somewhat lobulated. Cut sections are solid and cystic with gelatinous changes and areas of hemorrhage and necrosis [10]. The existence of other germ cell tumors components could be sometimes grossly recognized, such as the presence of calcification which could suggest an associated mature teratoma [2]. In our case, gross appearance was typical of a pure yolk sac tumor. On histology, yolk sac tumors show great architectural polymorphism and varying histological subtypes. Three diagnostic criteria are often observed: reticular architecture with anastomosing channels, lined by tumor cells with clear cytoplasm and irregular, hyperchromatic, nucleolated nuclei with strong mitotic activity; Schiller–Duval bodies (pseudopapillary structures formed by tumor cells organized around vascular structures) and PAS-positive hyaline droplets [1]-[6] [11]. In our case, the presence of cystic architecture, that appeared grossly as honeycomb areas was compatible with a polyvesicular vitelline pattern. The latter is often encountered in pure form of yolk sac tumor. On

immunohistochemistry, yolk sac tumor is characterized by a constant expression of AFP by epithelial components [12]. However, other markers are useful for ruling out other tumors. Histologically, clear cell carcinoma constitutes the main differential diagnosis. This latter does not have Schiller-Duval bodies, can express AFP in rare cases and is positive for CK7 and EMA with diffuse staining. Yolk sac tumor is more polymorphic with negative staining for CK7 and EMA [2]. The distinction between these two tumors has a major interest in their therapeutic management. Unlike clear cell carcinoma, yolk sac tumor is very chemosensitive and in some cases, an evaluation of the postoperative chemotherapy response may be useful for diagnosis [2] [13]. In our case, histological findings were typical of a yolk sac tumor with a strong expression of AFP by tumor cells, that facilitated the diagnosis. Besides, AFP serum level constitutes an important parameter for diagnosis and therapeutic surveillance. The decrease in its level may post-operatively reflect the absence of residual tumor [14]. Surgery followed by adjuvant chemotherapy constitutes the standard treatment for yolk sac tumor [6]. After chemotherapy, normalization of serum AFP level may be indicative of therapeutic efficacy. Serum AFP level is also more sensitive than CT for detecting tumor recurrences [1]-[6] [14]. In our case, serum AFP level was extremely high preoperatively (19,206 ng/ml) and decrease gradually after surgery with a normal level (5 ng/ml) after chemotherapy.

For young patients wishing to preserve their fertility like in our case, surgical treatment consists in unilateral adnexectomy via laparotomy with peritoneal washing and complete staging procedure including peritoneal cytology as well as peritoneal and omental biopsies. Chemotherapy combines several molecules and represents a promising strategy to reduce the risk of recurrence [1]-[6]. In our case, the patient has benefited from a BEP-type protocol (bleomycin, etoposide, cisplatin). The VIP regimen (etoposide, ifosfamide, cisplatin) can also be used as a second-line treatment if the tumor persists [6] [15]. For our patient, clinical outcome was favorable, with no residual or recurrent tumor.

4. Conclusion

Yolk sac tumors of the ovary are rare malignant germ cell tumors with a high potential for malignancy. They occur more often in young women. Serum AFP level and imaging examination can be helpful for diagnosis. The definitive diagnosis is based on histology. Conservative management might be proposed in young patients who wish to preserve their fertility. Given the very high chemosensitivity of yolk sac tumors, adjuvant chemotherapy clearly improves survival.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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