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Yolk tumor on testicular ectopia about a case and review of the literature: Contribution of imaging

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Abstract

Vitellin or endodermal sac tumours are rare germ malignant tumours. They account for less than 5% of germ cell tumours. They are aggressive tumors that preferentially affect children and young people. We illustrate here the importance of multimodality imaging for the diagnosis of vitellin sac tumor so we present a literature review about the case studied.

Keywords: Yolk sac tumor; testicular ectopy; young subject; scrotal ultrasound; CT scan; chemotherapy; surgery.

Introduction

Malignant tumors of the testis are in 98% of cases germ cell tumors (GCTs), divided into pure seminomatous GCTs (PSGTs) and non-seminomatous GCTs (NSGTs) in 55 and 45% of cases [1]. Since 2016, the WHO classification takes into account their histogeneses and molecular characteristics. The yolk sac tumor is part of the TGNS, presenting radiologically on a testicular ultrasound as a hypoechoic nodule, more or less homogeneous compared to the rest of the normal testicular parenchyma, sometimes with cystic components or calcifications. Histologically by the presence of tumor cells organized around characteristic vessels forming SCHILLER-Duval bodies. These tumors are rare in adults and represent 72% of testicular tumors in children aged 1 to 2 years [2-3]. In this article, we present the clinical case of a patient with a yolk sac tumor of the testis on testicular ectopy at the metastatic stage under chemotherapy.

Objective

J.O., a 25-year-old patient with a history of right testicular ectopia, was admitted to the emergency room with a history of sub-occlusion (chronic constipation for 6 months) that had been worsening for 2 months prior to his admission. The clinical examination found a conscious patient, hemodynamically and respiratory stable, WHO 1-2, a distended abdomen with a hard abdomino-pelvic mass, fixed in both planes and measuring 10x5cm, the testicular examination revealed an empty right bursa and left testicle in place, the lymph nodes were free. The whole evolving in a context of apyrexia and weight loss quantified at 15kg/ 6 months

A biological assessment was performed and showed normocytic normochromic anemia with hemoglobin at 8g/dl, WBCs at 11,000/mm, and renal failure with urea at 0.70G/L, creatinine at 40Mg/. Scrotal ultrasound and an initial CT scan showed a large retroperitoneal abdominal and pelvic process measuring 23x12.5cm responsible for bilateral UHN. Tumor marker assays showed an elevation of lactic dehydrogenase (LDH) to 1028 IU/L (normal 4.5), as well as alpha-fetoprotein, which was over 400 ng/ml (10000ng/ml). The diagnosis of a testicular tumor was evoked, an ultrasound-guided biopsy of the peritoneal mass and immunohistochemistry were performed. Anatomopathological examination associated with IHC (anti alpha1 FP + and anti pancytokeratine +, anti OCT 4 AND anti CD30 -) oriented the diagnosis in favour of a yolk sac tumor.

The patient benefited from a double J catheter at the UHN with a good improvement of the renal function (creat at 9 and then at 7 Vs 40 and urea at 0.17) and received a primary chemotherapy type BEP (bleomycin, etoposide and cisplatin) with a good clinical tolerance. The thoraco-abdomino-pelvic imaging showed the appearance of a secondary pulmonary location contrasting with the decrease in size of the abdom-ino-pelvic tumor process. Tumor marker monitoring revealed a decrease in LDH levels from 500 to 288 IU/L vs 1028 and AFP from 10,000 to 612 ng/ml. At the end of the treatment, thoraco-abdomino-pelvic imaging showed a decrease in size of the abdom-ino-pelvic imaging showed a decrease in size of the abdom-ino-pelvic imaging showed a decrease in size of the abdom-ino-pelvic imaging showed a decrease in size of the abdom-ino-pelvic imaging showed a decrease in size of the abdom-ino-pelvic imaging showed a decrease in size of the abdom-ino-pelvic imaging showed a decrease in size of the abdom-ino-pelvic imaging showed a decrease in size of the abdom-ino-pelvic imaging showed a decrease in size of the abdom-ino-pelvic imaging showed a decrease in size of the abdom-ino-pelvic imaging showed a decrease in size of the abdom-ino-pelvic imaging showed a decrease in size of the abdom-ino-pelvic process coming in contact with the rectum

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without separation line and stability of the secondary pulmonary localizations. The patient was operated on with resection of the testicular tumor mass and the rectum estimated at R2 and placement of a clip on the right lateral pelvic wall and realization of a colo-supra-anal anastomosis. 4 months later, the evolution was marked by a recurrence of the abdomino-pelvic mass metastasizing to the liver and lymph node level with sign of peritoneal carcinosis. Patient in poor general condition under chemotherapy and radiological surveillance. The followup is 2 years.

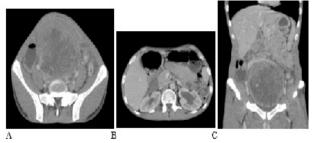
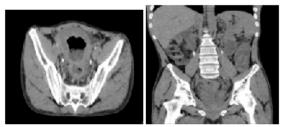
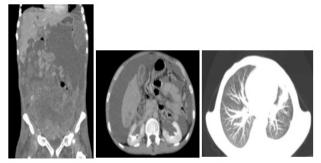


Figure 1: A: axial section of the abdominal and pelvic mass before CT B: Ureterohydronephrosis

C: coronal reconstruction of the abdomino-pelvic mass before CT



D: axial section of the abdomino-pelvic process after 4 CT scans; clear reduction of the tumor volume invading the rectum



E: coronal reconstruction of the abdomino-pelvic mass.

X-ray images in axial and coronal section parenchymal and abdominal window showing recurrence of the tumor mass with secondary hepatic and pulmonary localizations and peritoneal carcinosis.

Discussion

From an epidemiological point of view, testicular germ cell tumors are rare tumors, and represent 1 to 2% of cancers in men [7] (3.5% of tumors of the urogenital system). They are the leading cause of solid tumors in young men with a peak incidence between 30 and 34 years of age [14].

The post pubertal yolk sac tumor, also called endodermal sinus tumor, is a malignant germ cell tumor whose morphologi-

cal aspects are reminiscent of extra embryonic structures: yolk sac, allantois and extra embryonic mesenchyme. Most patients are between 15 and 40 years of age, although later cases have been described [15]. The yolk sac tumor is almost always part of a mixed germ cell tumor and is found in 44% of TGNS.Pure forms are exceptional representing 0.6% of germ cell tumors of the testis [7]. In this case, the yolk sac tumor was a pure form, occurring in a 25-year-old patient with right testicular ectopia; it was a tumor differentiated in the extraembryonic sense. Some risk factors for germ cell tumors are consensual [8] testicular dysgenesis syndrome (cryptorchidism, hypospadias, hypo or infertility), personal or family history of TG, testicular atrophy (<12ml), others are discussed such as heavy cannabis use, pesticides, organic solvents, morphometry [9].

Testicular Doppler ultrasonography is the examination indicated as the first line of treatment for intragonadal germ cell tumors. It has a sensitivity close to 100% for the positive extra and intra-testicular diagnosis of a scrotal mass [4]. It allows confirmation of the presence of a tissue process by specifying the site, the semiological characteristics: size, contours, limits, echogenicity (it typically shows a hypoechoic intra-testicular image), and the vascular nature of the tumour, and thus it explores the contralateral testicle and allows its surveillance. The risk of developing a second tumor is increased if there are grade 3 micro-calcifications [11]. In our series, ultrasound revealed a well-limited oval abdominal and pelvic tumour process, with a tissue echostructure and a minority cystic component measuring 168x102x155m with bilateral pyelocalic dilatation.

The learned societies of radiology (SFR, HAS) consider that CT-PET is systematically recommended for the initial assessment and follow-up of TG without and with injection of iodinated contrast medium, and allows the detection of lesions larger than 3 mm, showing a mixed mass, solid or cystic with significant heterogeneity due to hemorrhage or necrosis, Calcifications are noted in 40% of the situations, as shown in our case where the TAP CT mentioned a voluminous heterodense abdomino pelvic process containing a zone of liquefaction and fleshy contents as well as vessels inside with uretero hydro nephrosis The sensitivity of 80% for the analysis of retroperitoneal adenopathies and the evaluation of their size according to the largest transverse diameter (RECIST criteria) [5] allows the initial staging of this lymph node extension from N1 to N3 (16) (**Table 1**).

Testicular cancers metastasize by lymphatic and hematogenous routes. The first lymphatic relays are the latero-aortic areas immediately under the kidneys: indeed, in the embryo, the testis forms near the kidneys and then descends into the bursa. At a more advanced stage, the supra-diaphragmatic, mainly left supra-clavicular, and mediastinal nodes may be invaded.

Hematogenous dissemination occurs via the spermatic vein and the inferior vena cava: it is mainly responsible for pulmonary localizations, as in our patient's case, and, much more rarely, for hepatic, cerebral and bone localizations. It should be noted that in 10% of cases, the histological nature of the metastases differs from the primary germ cell tumor. Finally, it should be noted that rare germ cell tumors are extragonadal, retroperitoneal, mediastinal or pineal in location including.

Table 1: TNM classification for testicular cancer.

pTIS	Intratubular germ cell neoplasia (carcinoma in situ)	
pT1	Tumour limited to testis and epididymis without vascular/lymphatic invasion; tumour may invade tunica albuginea but not tunica vaginalis*	
pT2	Tumour limited to testis and epididymis with vascular/lymphatic invasion, or tumour extending through tunica Ibuginea with involvement of tunica vaginalis	
pT3	Tumour invades spermatic cord with or without vascular/lymphatic invasion	
pT4	Tumour invades scrotum with or without vascular/ lymphatic invasion	
N - Regional Lymph Nodes - Clinical		
NX	Regional lymph nodes cannot be assessed	
N0	No regional lymph node metastasis	
N1	Metastasis with a lymph node mass 2 cm or less in greatest dimension and 5 or fewer positive nodes, none more than 2 cm in greatest dimension	
N2	Metastasis with a lymph node mass more than 2 cm but not more than 5 cm in greatest dimension; or more than 5 nodes positive, none more than 5 cm; or evidence of extranodal extension of tumour	

Magnetic resonance imaging is not routinely recommended because of its cost and limited availability, but it can replace CT in cases where iodine injection is contraindicated. It can be useful in residual masses by identifying the operative difficulties that may be encountered or in the presence of an atypical lesion.

Positron emission tomography (PET scan) is not recommended for the initial staging of TST. It is currently being evaluated in stage I GIST [6]. For other examinations including spinal CT, bone scan or liver ultrasound, are indicated depending on the clinical context dictated by the metastatic disease. Brain CT or brain MRI are recommended in cases of extensive metastatic TGNS of the lungs and in forms with poor prognosis.

	Recommandation	Grade
Marqueurs tumoraux sériques (hCGt, AFP, LDH)	Systématique	
Echographie scrotale	Systématique	
Scanner TAP	Systématique	
IRM abdominale	En cas de contre-indication au scanner, pour l'évaluation ganglionnaire	
Imagerie cérébrale	En cas de symptômes ou chez le sujet à risque	
Scintigraphie osseuse	En cas de symptômes	
TEP-18FDG	Non recommandée	

The diagnosis remains histological after inguinal orchiectomy in non-metastatic forms [10] and reveals two morphological features: intracytoplasmic or extracellular hyaline globules (SCHILLER-Duval bodies) and intracellular deposits of the basement membrane arranged in a band. The histological study of our case reveals a myxoid, trabecular and microcystic aspect consistent with a yolk sac tumor. However, yolk sac tumors can mimic other types of TG and non-germline tumors especially in metastatic and extra gonadal sites, for this reason immunohistochemical markers are sometimes used to facilitate and support the diagnosis [10].

Sperm cryopreservation at the CECOS is recommended, ideally before orchiectomy and imperatively before any chemotherapy, radiotherapy, or retroperitoneal surgery. It has a medicolegal value [12]. Chemotherapy with four cycles of BEP is the reference treatment for metastatic non-seminomatous germ cell tumors. The other therapeutic options are: surveillance, chemotherapy associated or not with surgery.

Conclusion

The extension workup for testicular cancer uses the international TNM classification. The T stage is a post-operative pT stage (post-orchidectomy). The N and M stages are determined by a thoraco-abdomino-pelvic CT scan (lymph node metastases in the medial retroperitoneum and visceral pulmonary localizations especially). Monitoring under and after treatment is performed by multiple thoracic-abdominal-pelvic scans, the rhythm of which depends on multiple parameters (type of tumor, initial stage) and by ultrasound scans of the remaining testicle. A PET scan can provide information in case of a residual lymph node mass after chemotherapy.

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