High-grade metastatic paratesticular cystadenocarcinoma

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1. Introduction

Paratesticular cystadenocarcinoma is a rare form of ovarian epithelial tumor, with less than forty cases reported to date. Clinical presentation is scrotal enlargement with no significant pain. Ultrasound often shows a hypoechoic vascularized mass, with variable amounts of hydrocele. CT and MRI are not able to differentiate this tumor from other differential diagnosis. When normal values of human chorionic gonadotropin and alpha-fetoprotein are seen associated with a high value of serum CA125, this disorder should be suspected and confirmed by immunohistochemistry (IHQ). Herein we report a case of metastatic presentation of a paratesticular cystadenocarcinoma.

2. Case report

A 76 years old, smoker, hypertensive, african-american male, presented at the clinical oncology division with multiple enlarged cervical nodes and significant weight loss of five months duration. The patient referred no personal or familial history of cancer. CT of the thorax showed enlarged mediastinal nodes and a left supraclavicular node. Ultrasound revealed an expanded cavity with 10.8 × 8.9 × 6.0 cm, liquid filled with a round nodule inside. This nodule measured 4.3 × 3.6 × 2.9cm, and had no vascularity on doppler evaluation. A CT confirmed a solid nodule with only peripheral contrast enhancement (Fig. 1). HCG, AFP and lactic dehydrogenase were negative.

An uneventful radical orchiectomy was performed and macroscopy findings showed a tumor mass which weighted 441g and measured 14,2 × 9,0 × 8,6cm with no invasion of the spermatic cord. The mass had a thick external layer, with an oval brown fibrous nodule inside. The remaining testicular tissue could be seen compressed against the thick external layer (Fig. 2 – red arrows). IHQ was positive for CK7 and CA125 and negative for HCG and AFP. The final pathology report was paratesticular serous carcinoma, originating from persistent müllerian duct remnants. The negative Calretinin ruled out mesothelioma. Skin lesions were also positive for CK7, CA125, estrogen and progesterone receptors, and negative for germinative tumors markers (Fig. 3), confirming the metastatic nature of the tumor.

Six months later the patient presented with newly enlarged cervical and supraclavicular nodes, but no new skin lesions.
Sunitinibe was introduced along with five sessions of radiotherapy for the cervical nodes. After seven months of follow up the patient showed partial clinical response and reports mild residual pain on irradiated area.

3. Discussion and literature review

Since the first report by Blumberg and als. in 1991 only a few reported cases of malignant serous adenocarcinoma of testis have been published. Usually, these tumors are found in a paratesticular location and can reach patients in distinctive ages, ranging from 16 to 87 years old.\(^1\)\(^,\)\(^2\)

The correct origin of the ovarian type epithelial tumors of the testis has not yet been established. Some studies refer that these cells are more likely to originate from Müllerian ducts remnants of the paratesticular connective tissue, epididymis and spermatic cord, while others report that they may originate from the mesothelium of the tunica vaginalis after Müllerian remnants metaplasia. Further, in a recent review, it was hypothesised that these cells come from Müllerian metaplasia of intratesticular mesothelial inclusions that occurred during embryogenesis.

Clinical findings include scrotal enlargement and negligible pain. Histological presentations are mainly serous and mucinous, but can be also endometrioid or Brenner tumors. They can be benign or malignant, but are usually described as borderline behavior. One important differential diagnosis is mesothelioma, which can be excluded when the IHC marker caretinin is found negative.\(^3\)

An IHC panel including CK7, CK20, TTF1, progesterone receptor and estrogen receptor is essential for correct diagnosis. Usually serum values of HCG and AFP are normal and serum CA125 is elevated. Chromosomal and genetic mutations have controversial
results, are expensive and were not performed in our case.
Radical orchiectomy is the primary recommended treatment. There is no consensus on the best adjuvant therapy, because these tumors are usually chemo and radio resistant. Of the reported cases without adjuvant therapy, 19% developed metastasis after 48–84 months of follow up. Due to paucity of reports and absence of long term follow up we cannot estimate survival rates.

4. Conclusion
A rare case of metastatic paratesticular serous cystadenocarcinoma is reported. IHQ positivity for CK7, negative germinative tumor markes and positivity for CA125 arises suspicion for this rare diagnosis. Surgery is recommended but there are no guidelines for adjuvant treatment and follow up.

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