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Case report

The primitive extratesticular seminoma: diagnosis of a rare pathology

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Summary. *Background:* The Primitive Extratesticular Seminoma is a very rare condition and represents 3% of germ cell tumors; it is an indeterminate origin disease, whose diagnosis is often complicated by a nonspecific and highly variable clinical finding. *Case presentation:* A caucasian 55 years old male, non-smoker, arrived to our centre with cough, severe respiratory distress and dysphagia, in a context of the superior vena cava syndrome. A Computed Tomography was performed, which shows the presence of a mediastinal anterior mass with aorto-pulmonary window and left paracardiac invasion. A biopsy of the mediastinal mass was performed with mediastinoscopy; the hystological diagnosis was seminoma. After, first of all the ^{18}FDG PET-CT is performed, which shows the presence of an intense hypermetabolism (SUV max=20.3 and metabolic volume 867 cc) at the level of bulky mediastinal mass, with paratracheal, aorto-pulmonary window and left paracardiac invasion. The mass presents also a sternal manubrium invasion. There were no other detectable tissue metabolic alterations with the ^{18}FDG PET-CT and, in particular, the testicles examination was negative. A bilateral testicular ultrasound (US) was executed, which confirms the absence of testicular germ tumor. Clinical laboratory tests show a significant increase of beta-HCG (123 IU/L); AFP is negative. A final diagnosis of primitive extratesticular seminoma was carried. *Conclusions:* The Primitive Extratesticolar Seminoma is a rare patology, and, for its massive size at the onset diagnosis, curable in early stage often only with radiochemotherapy. The Diagnostic Imaging and Nuclear Medicine, as Testicular Ultrasound, the CT with contrast medium and the ^{18}FDG PET-CT total body examination, are fundamental to the staging and localisation. MRI sometimes is useful for testicular evaluation. The biopsy is needed for tissue characterization. (www.actabiomedica.it)

Key words: extratesticular seminoma, ^{18}FDG PET-CT, Computed Tomography

Background

The Primitive Extratesticular Seminoma is a very rare condition and represents 3% of germ cell tumors (1); it is an indeterminate origin disease, whose diagnosis is often complicated by a nonspecific and highly variable clinical finding (1).

Case presentation

A caucasian 55 years old male, non-smoker, arrived to our centre with cough, severe respiratory distress and dysphagia, in a context of the superior vena cava (SVC) syndrome. It is the first time that he presents so important symptoms in the respiratory tract.

A Computed Tomography (CT) with contrast medium is performed (Fig. 1 a-c), which shows the presence of a mediastinal anterior mass (11 x 9 cm), with aorto-pulmonary window and left paracardiac invasion. The mass present a contact with the sternum (Fig. 1c, arrow), witch it is irregular. No other abnormality have found.

It was decided to make a mediastinoscopy of the solid mass in the antero-superior mediastinum; a bilat-

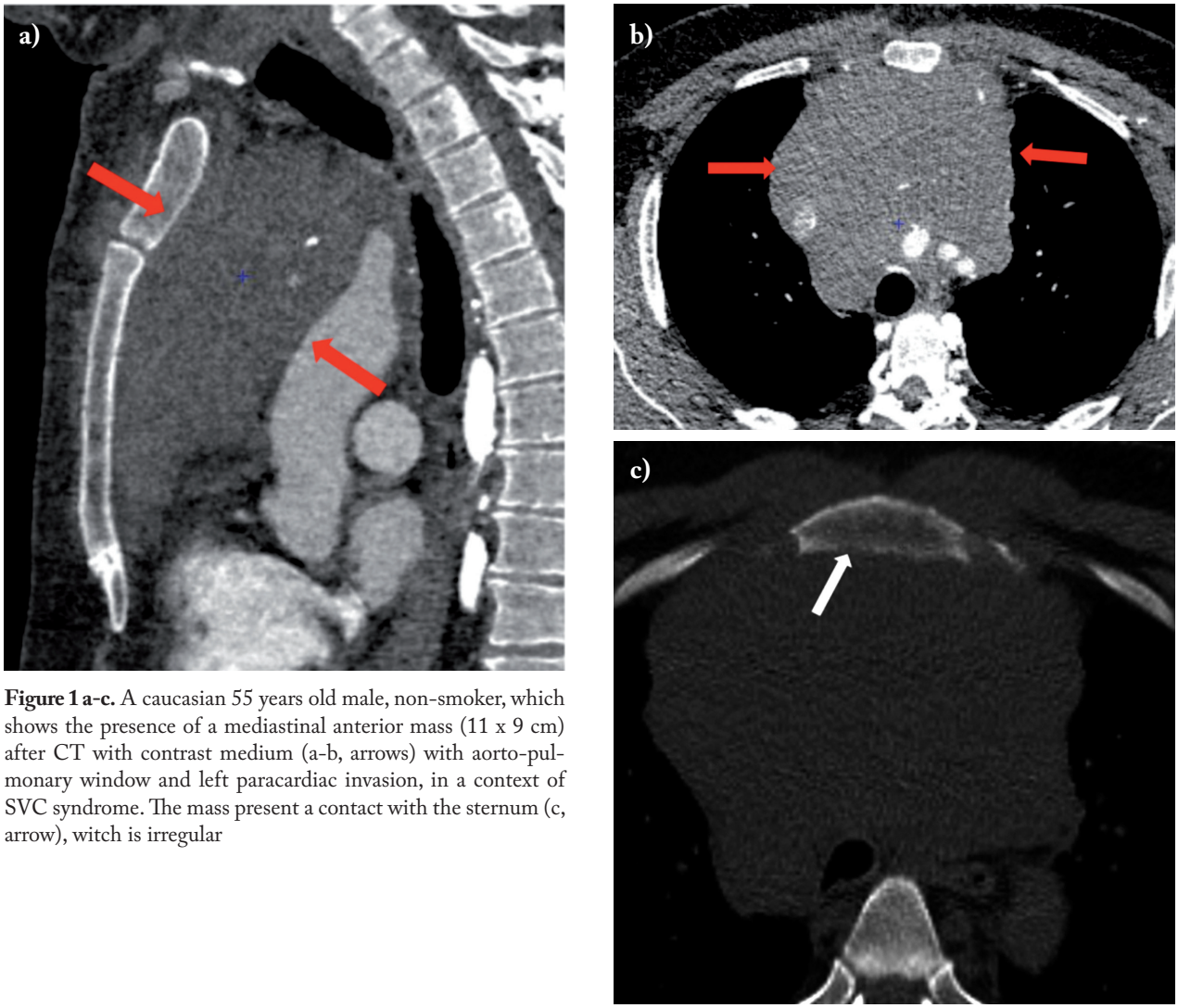

Figure 1 a-c. A caucasian 55 years old male, non-smoker, which shows the presence of a mediastinal anterior mass (11 x 9 cm) after CT with contrast medium (a-b, arrows) with aorto-pulmonary window and left paracardiac invasion, in a context of SVC syndrome. The mass present a contact with the sternum (c, arrow), witch is irregular

eral compression of the arterio-venous vascular structures was present. A biopsy of the mediastinal mass was performed; the hystological diagnosis was seminoma.

The diagnostic process was completed with: first of all the ^{18}FDG PET-CT (Fig. 2), which shows the presence of an intense hypermetabolism (SUV max=20.3 and metabolic volume 867 cc) at the level of bulky mediastinal mass, with paratracheal, aorto-pulmonary window and left paracardiac invasion. The mass presents also a sternal manubrium invasion.

There were no other detectable metabolic tissue alterations with the ^{18}FDG PET-CT and, in particular, the testicles examination was negative.

A bilateral testicular ultrasound (US) was performed (Fig. 3 a-b), which confirms the absence of testicular lesions.

The clinical laboratory tests show a significant increase of beta-HCG (123 IU/L); Alpha-fetoprotein (AFP) was negative.

A final diagnosis of Primitive Extratesticular Seminoma was carried.

Discussion

The testicular cancer incidence in Europe is rising, currently 6.3/100.000/year, with the highest rate

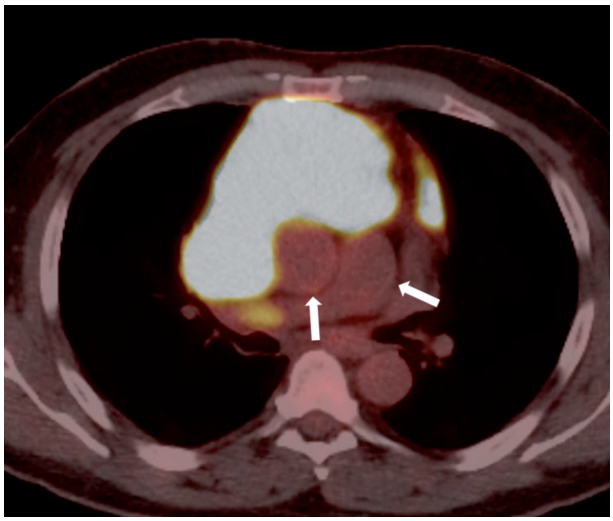

Figure 2. The ^{18}FDG PET-CT is performed, which shows the presence of an intense hypermetabolism (SUV max=20.3 and metabolic volume 867 cc) at the level of bulky mediastinal mass, witch confirm the aorto-pulmonary and left paracardiac invasion (vascular structures, arrows)

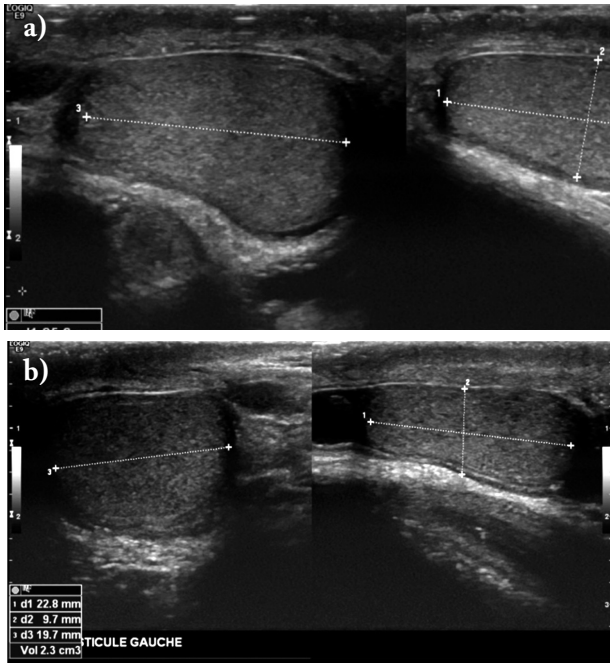

Figure 3 a-b. A bilateral testicular ultrasound (US) was executed, which confirms the absence of testicular germ tumor (the testicles examination was negative at the ^{18}FDG PET-CT)

in Northern European countries (6.8/100.000/year); between testicular tumours, 40% are seminomas and 60% non-seminomas (1-2). The Primitive Extratesticular Seminoma represents a very rare pathology, about 3% of germ cell tumors (1-2).

These tumors occur almost exclusively in males, and the age at presentation is generally 20-35 years (3). More than a third of all malignant germ cell tumors are pure seminomas (3). Seminoma is one of the most radio and chemo sensitive tumors (4). When treated, the prognosis is good, with a mean survival of 90% at five years (5). Aggressive variant of PMS are rarely described in the literature (5). The death rate is very low (0.38 cases/100 000/year) (3).

Extragonadally germ cell tumour may present often in the mediastinum area and in a minority of cases into the retroperitoneum space. There are some study with a description of pancreatic or thyroid invasion (7). These patients present with undifferentiated (adeno)carcinoma of unknown origin, mostly with typical marker elevation and/or elevated copy number of chromosome i12p, which is specific for germ cell tumours (4).

Clinical evaluation of the primitive extratesticular seminoma depend on the location, the size and the initial symptoms (2). Into a mediastinal localisation, back pain is a common complaint in the outpatient setting, often with a persistent cough and dyspnea associated; distention of the jugular veins, swollen lymph nodes were palpable in the subclavicular region; superior vena cava and tracheal compression were presents (9, 10).

The Immuno-hystological evaluation is necessary, considering in particular the dosage of AFP, beta-HCG and D-lactate dehydrogenase (LDH).

For the diagnosis the testicular sonography should be conducted, for evaluate testicular size and possible primitive lesions.

A CT including thorax, abdomen and pelvis is recommended for the cancer staging, often with the execution of the ^{18}FDG PET-CT, that is a possible option for stages II/III, in particular for defining treatment strategy in case of residual tumour. ^{18}FDG PET-CT does not contribute in early stages of seminoma [I, B] (5).

The Magnetic Resonance Imaging (MRI) are being investigated as tools to further characterize testicular lesions; several recent articles have described the utility of MRI for the evaluation and the characterization of testicular lesions (6-8). The MRI showed a 100% of sensitivity and an 88% of specificity for differentiation

of benign from malignant intratesticular lesions (5, 9). It is not useful in the extratesticular localizations.

The histology results confirm the diagnosis of Primitive Extratesticular Seminoma (9-10). The differential diagnosis with Nuclear Protein of the Testis (NUT) Midline Carcinoma, that is a rare aggressive carcinomas characterized by chromosomal rearrangements that involve the gene encoding the NUT, is necessary (17).

Conclusions

The Primitive Extratesticolar Seminoma is a rare patology, and, for its massive size at the onset diagnosis, curable in early stage often only with radiochemotherapy.

The Diagnostic Imaging and Nuclear Medicine, as Testicular Ultrasound, the CT with contrast medium and the ^{18}FDG PET-CT total body examination, are fundamental to the staging and localisation. MRI sometimes is useful for testicular evaluation. The biopsy is needed for tissue characterization.

However, diagnosis is difficult and can be delayed for the extreme rarity of the pathology.

Ethical Statement: All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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