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CASE REPORT

A testicular tumour in the left adnex. The persistent Mullerian duct syndrome with testicular malignancy

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Cryporchidism is known to increase the incidence of testicular malignant tumours. In this report a patient is described with the Mullerian duct syndrome in connection with a cryptorchistic malignant testicular tumour.

Key words: persistent Mullerian duct syndrome; testicular malignancy.

Introduction

Cryptorchidism is known to increase the incidence of testicular tumours.[1 4] A rare cause of non-descending testicles is persistent Mullerian duct syndrome, one of the types of male pseudo-hermaphroditism.[5] In this type, as distinct from other types, virility is not disturbed yet a deficiency in regression of the Mullerian duct occurs, in genotypic and phenotypic male persons. Up to 1981, about 80 patients with this syndrome have been described in the literature.[6 9] A testicular tumour, complicating this syndrome, is a very rare condition.

Case report

A 45-year old married man, with no children was hospitalized because of the following complaints, malaise, night sweat, loss of weight and pain localized at the left side of the lower abdomen.

Patient history revealed no abnormalities, but physical examination revealed a large, solid tumour in the lower abdomen. No testicles could be found in either the scrotum or both inguinal regions. A solid irregular resistance could be felt in Douglas cave. Inguinal lymph nodes were not enlarged, the remaining external genitals were normal, and the patient characterized his sexual functioning as normal.

Laboratory findings showed a considerable increase in tumour-markers (BHCG 16.3 μg/l, aFP 700 μg/l). Computed tomography of the abdomen showed a big solitary tumour, extending into the small pelvis. Histologic analysis of tissue obtained by means of trans-abdominal needle biopsies showed a non-seminoma testicular tumour. The poor quality of the specimen rendered greater definition impossible.

The tumour was classified as T × M3N0 (stage IIc). The patient was treated with polychemotherapy: bleomycin 30 mg, VP-16 180 mg, cisplatinum 40 mg at day 1 and at day 8 30 mg bleomycin, with an interval of 3 weeks. After four treatments there was considerable tumour regression. The BHCG dropped to 5 μg/l and aFP to 8.2 μg/l.

A CT-scan of the abdomen still showed a large quantity of pathologic tissue, so a staging laparotomy was performed to evaluate the result of therapy. Inspection of the abdomen revealed a small uterus with both adnexes, and a large remnant of tumour, 15 cm in diameter located in the left adnex. The uterus ended up blindly on the pelvic floor and was excised totally with both adnexes including the tumour.

Pathology showed a necrotic tumour, due to

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chemotherapy. No vital tissue tumour could be found. In addition an atrophic uterus, Fallopian tubes, a testicle with seminiferous tubes at the right adnex, vital testicular reticulum and remnants of the epididymis were found. No vasa deferentia were found. Spermatogenesis appeared to be non-functioning. In the bioped para-aortal lymph nodes no tumour tissue was found.

Chromosomal investigation revealed in this patient a normal male genotype 46xy. Post-operative recovery was uneventful and during a 2-year follow-up until July 1989, there were no signs of tumour relapse.

Discussion

In the persistent Mullerian duct syndrome caused by male pseudohermaphroditism, no regression of the Mullerian duct by the Mullerian Inhibiting Factor (MIF) seems to occur. MIF is known to be produced by fetal Sertoli cells. The regression of the Mullerian duct fails when MIF synthesis is disturbed, when there is a structural deformity in the MIF or when the Mullerian duct is too small for MIF.

As there generally is normal androgen and testosterone production normal virility occurs as does sexual differentiation. Some cases of spermatogenesis have been described, but without fertility.

The persistent Mullerian duct syndrome frequently appears in combination with uni- or bilateral cryptorchidism, transverse ectopia testis (both testicles localized at one side of the midline)[10,11] and hernia uteri inguinalis. A hereditary component has been suggested, lacking any clinical or genetic support so far. Although the location of a testicular tumour as presented in this case report is a rare condition, some authors indicate that in the persistent Mullerian duct syndrome there is an enlarged incidence of testicular malignancy.[9,12] Whether the malignant deformation in the persistent Mullerian duct syndrome is merely due to absent descent of the testicle into the scrotum or to factors producing the defect in fetal Mullerian inhibiting substance is not clear.[13]

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