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CASE REPORT

PINEAL BODY SEMINOMA FOLLOWED BY TESTICULAR EMBRYONAL CARCINOMA IN A PATIENT WITH BILATERAL MICROLITHIASIS

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CASE REPORT

A 26-year-old man presented to our hospital in 1993 with a history of double vision and headache that had worsened during the last three months.

Fundoscopic examination revealed evidence of optical nerve compression. The patient subsequently went for computed tomography (CT) scan of the head. CT scan confirmed a pineal body mass with hydrocephalus. A ventriculoperitoneal shunt and biopsy of the pineal body mass was done by the neurosurgeon.

The pathological examination of the biopsy material confirmed the diagnosis of seminoma. Serum alpha fetoprotein and human chorionic gonadotropin were normal. Also abdominal computed tomography did not reveal any abnormality.

Unfortunately there was no record of scrotal ultrasound investigation at that stage although scrotal examination was recorded to be normal. The patient received a course of radiotherapy to the head followed by four courses of chemotherapy (carboplatin, etoposide and ifosfamide). At completion of chemotherapy, the pineal body mass could not be visualized any more on CT scan.

Follow-up evaluation included physical examination, CT scan of the head every six months for the first year followed by yearly examinations, and determination of the serum tumor marker levels every four months for the first three years followed by a six monthly check up. The patient had an uneventful post-treatment course, until - eight years later - he presented with lower abdominal pain and a hard left testicular mass on examination. Ultrasound scan of the scrotum revealed bilateral microlithiasis of the testes and a well circumscribed hypoechoic lesion involving the left testis. (Fig 1)

Subsequent left radical orchidectomy revealed the presence of a local testicular embryonal carcinoma without extension beyond the tunica albuginea, and there was no evidence of lymphatic or vascular invasion. The tumor markers were within normal limits and the abdominal CT scan findings were unremarkable. After careful consideration and discussion with the patient, we abutted for surveillance instead of retroperitoneal lymph node dissection or immediate chemotherapy. The patient was free of recurrence of the disease at twelve months of follow-up.

DISCUSSION

Germ cell carcinoma represents the most common malignancy in men between fifteen and thirty-five years of age.

Extragonadal germ cell tumor is a rare condition representing only 3% to 5% of all germ cell carcinomas. Extragonadal germ cell tumor sites include the mediastinum, the retroperitoneum, the peritoneum, the cranium and the coccyx. A second primary germ cell tumor in patients with a history of extragonadal germ cell carcinoma has been previously reported in only eight cases[1-3].

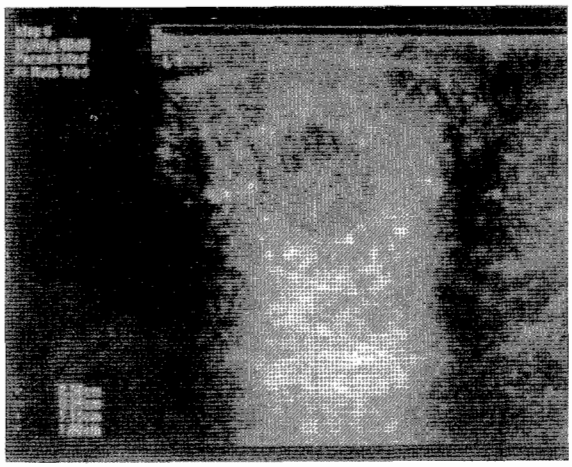

Fig. 1: Left testis with diffuse microlithiasis and a hypoechoic lesion

In all the reported cases the presenting extragonadal germ cell tumour was non seminomatous[1-3]. One reported case of pineal yolk sac tumor was diagnosed after treatment of a testicular seminoma[4]. According to our knowledge, the present case may be the first reported case in the literature of pineal body seminoma followed by testicular embryonal carcinoma with normal tumor markers and bilateral testicular microlithiasis.

The mechanism of development of extragonadal germ cell carcinoma remains unknown. There are three possible theories: The first theory suggests that germ cell malignancy starts from totipotential cells, which are precursors of the germ cells in the extragonadal tissues. The second theory is that there is a migration rest of the germ cells during embryological development. The third theory has been defined as "burned-out theory" postulating that there is a spontaneous regression of the tumor site after dissemination, leaving behind a scarring lesion with typical histological changes[3].

Testicular microlithiasis (TM) is a rare pathologic condition characterized by calcifications within the seminiferous tubules. These calcifications originate from degenerating intratubular cellular debris that is surrounded by concentric layers of stratified collagen fibers[5].

TM is diagnosed by high-frequency (5 to 10 MHz) testicular ultrasound (US) and has been seen with a wide variety of testicular pathologies including torsion of the testis or appendix testis, primary testicular neoplasm, intratubular germ cell neoplasia, benign cystic teratoma, varicocele, infertility, cryptorchidism, Klinefelter's syndrome, and even normal testes. The significance of such association is not known. According to a recent review of the literature it was found that 30% of all reported cases of TM were associated with testicular malignancies[6]. In all these reports, testicular cancer and TM were diagnosed simultaneously. Rarely, TM has been diagnosed prior to the development of testicular cancer. The longest reported interval between the diagnosis of TM and the diagnosis of a testicular malignancy is eleven years[7].

It is not clear, if the discovery of TM places the patient at an increased risk for the development of testicular tumors, and there is no evidence that TM is either a premalignant condition or a causative agent in testicular neoplasia.

Radiotherapy, chemotherapy or a combination of both were used in the reported patients with extragonadal germ cell tumors.

Owing to the fact that these tumors are rare and, in part, biologically distinct from their testicular counterparts, their optimal management still has to be defined.

In conclusion, we would like to emphasize the need of evaluating the testes by ultrasound scan at the time of making the diagnosis of extragonadal germ cell tumor as well as during follow up, which must continue in regular intervals. We recommend a meticulous follow up of patients with TM by means of self-examination and repeated ultrasound scan until the behaviour of the TM is defined.

Retrospectively, we believe that testicular biopsy must be included in the assessment of patients with extragonadal germ cell tumor and TM.

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