Klinefelter's Syndrome and Testicular Teratoma

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ABSTRACT

Testicular teratoma in a patient with Klinefelter's syndrome is described. Relationship between chromosomal abnormalities and the risk for neoplasm is discussed.

A number of reports on the association between chromosomal abnormalities and neoplasm have been done[1], and it is well known that Klinefelter's syndrome is prone to combine with breast cancer[2,3]. The present report describes a patient with Klinefelter's syndrome who had mature teratoma in the testis.

CASE REPORT

A 37-year-old man was presented with a diagnosis of right ureteral stone in November 1976. An excretory urogram (IVP) showed right ureteral stone and hydronephrosis. Physical examination revealed that the scrotal content on the left side was enlarged to about fist size, while the other side was atrophic. He had noticed his enlarged scrotal content since childhood.

The patient was admitted and underwent ureterolithotomy, orchiectomy on the left side, and biopsy of the right testis. He was 164.5 cm. tall and weighed 51 kg., without gynecomastia. The penis was normal although lacked development of the pubic hair. The laboratory data included no spermatozoa, urinary 17-OHCS 1.8 mg/day and urinary 17-KS 7.2 mg/day. Routine blood analyses were within normal limits. The left excised testicular tumor measured about $7 \times 9.5 \times 5$ cm and weighed 150 g and microscopic examination of it revealed mature teratoma containing bone, hair and fatty tissue (Fig. 1). A biopsy of the right testis showed typical changes of Klinefelter's syndrome (Fig. 2). Chromosomal karyotype of peripheral leukocyte was XXY (Fig. 3).

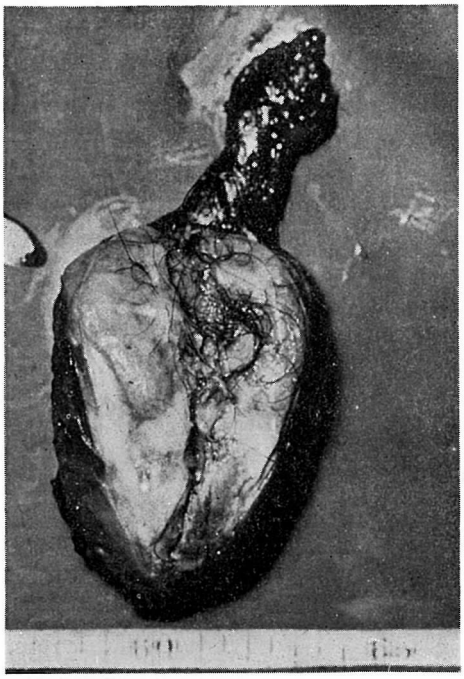

Fig. 1. Cross section of the teratoma showing bone, hair and fatty tissue.

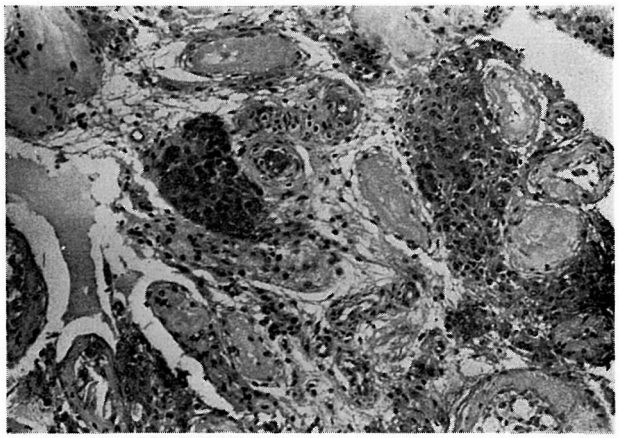

Fig. 2. A biopsy of the right testis reveals destruction of the seminiferous tubles and nests of Leydig cells.

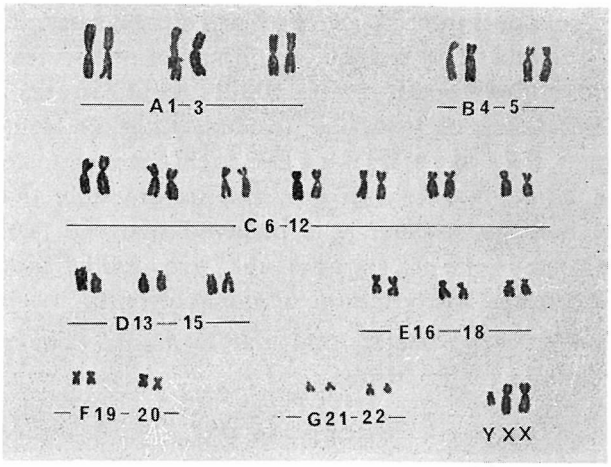

Fig. 3. Chromosomal karyotype showing XXY.

DISCUSSION

Teratomas are germinal cell tumors containing all three germ layers and which form various abortive organs. They frequently occur in male infancy and childhood, and are relatively benign tumors.

Klinefelter's syndrome is a genetic disorder which has 2, or more, X chromosomes and 1, or more, Y chromosomes, or a mosaicism of these. The testicular damage is characteristically seen in biopsy after puberty. Microscopic examination shows an absence of germinal epithelium, hyalinized seminiferous tubules, and nests of Leydig cells.

It is well known that there is a relatively high risk for neoplasm in an individual with chromosomal abnormalities. Cell transformation due to oncogenic viruses of the fibroblast from a cancer patient with chromosomal abnormalities is evidently higher than that of fibroblast from normal individuals[4,5]. Jackson and associates[2] reported that 3 of 21 cases of breast cancer in men were patients with Klinefelter's syndrome. Many other cancers also have been revealed[6,7]. However, the occurrence of testicular germinal cell tumors in Klinefelter's syndrome is very rare. To our knowledge only six cases of these are reported; four cases of teratomas[8], seminoma[9] and embryonal cell carcinoma[10]. Regarding teratomas, Gustavson and associates[8] suggest that the XXY chromosomes may predispose to the development of teratomas, because testicular teratomas frequently have positive X-chromatin.

On the other hand, there is a hypothesis that atrophy of the testes precedes the development of testicular tumors. That is, these testes are

stimulated by gonadotropin in the process of recovery, and then some develop into tumors. For examle, mumpus orchitis, testes in gonadal dysgenesis and undescended testes, which result in testicular atrophy, have a high incidence of testicular tumors. Judging from such facts, it may be assumed that there is a higher incidence of testicular germinal cell tumors in Klinefelter's syndrome. But in practice, the reports on it are very rare. For this reason, it is believed that the testicular damage in Klinefelter's syndrome is so rapid and irreversible that the testes can not be revived by the introduction of gonadotropin.

In this case it is suspected that the teratoma occurred before the chracteristic change of the testis.

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