

Bilateral Testicular Tumors in a True Hermaphrodite (46XX): A Rare Case Report

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Citation

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Abstract

A 56 year old female presented with bilateral inguino-labial swellings. She was found to have ambiguous genitalia on examination. Radiological investigations suggested the swellings to be of testicular origin. Both the swellings were excised and histopathology showed "seminoma" testis on one side and "embryonal cell carcinoma" on the other. The case is being reported because of a rare presentation as bilateral testicular tumors in a female phenotype hermaphrodite and also because of presence of synchronous double malignancy.

INTRODUCTION

As many as 1 in 3000 babies are born intersexed – where it is difficult to tell from physical appearance whether the child is a boy or a girl. Although western literature is aplomb with recommendation for early gender assignment surgeries in such patients keeping in mind the psycho-sexual development & risk of neoplasia, the subject still remains an enigma in Indian society where social taboos prevent such patients from having medical treatment at all. We are reporting one such case of intersex female who presented to us at an age of 56 yr with bilateral testicular tumors.

CASE REPORT

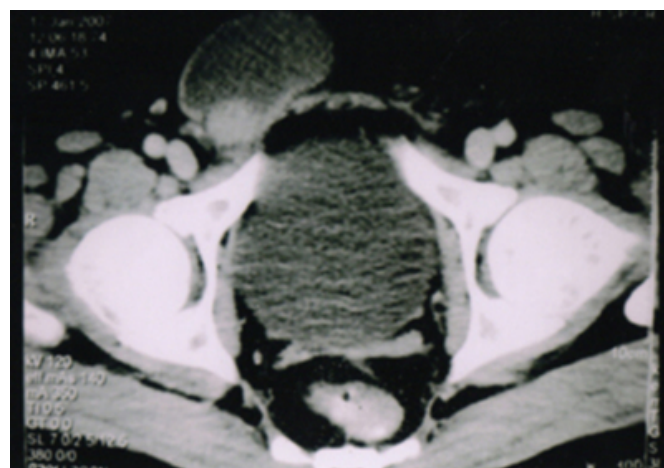
A 56 year old married female presented with a huge swelling in right inguino-labial region for 6 month duration. The swelling was progressively increasing in size without any associated symptoms. She was nulliparous and had primary amenorrhea. On examination, there was an oblong swelling of 12x8 cm size in right inguino-labial region with smooth surface, well-defined margins and cystic consistency. It was non-reducible and without any cough impulse but brilliantly trans-illuminant. Another small swelling of size 3x4 cm was noted in left labium majus. Per-vaginum examination revealed a normal vagina with hypertrophied clitoris. Secondary sexual characters including development of breasts were normal.

Ultrasound examination showed a cystic lesion at the site of swelling extending from right inguinal region into the labium majus with solid component at one pole. Uterus was

rudimentary. CECT showed a hypodense mass of CT no. suggestive of fluid extending from inguinal canal into the right labia majora with a hyperdense area in it showing enhancement on contrast. Another hyperdense area was seen in left labium majus showing enhancement suggestive of bilateral testicular masses (Fig.1).

Figure 1

Figure 1: C.T. film showing hyperdense areas in bilateral labia suggestive of testicular masses and rudimentary uterus.



A hypodense shadow seen posterior to bladder and anterior to rectum was suggestive of rudimentary uterus with bilateral atrophic ovaries. Rest of viscera were normal. Her karyotype was 46XX.

Patient was explored and following findings were noted :-

- (i) 12x8 cm cystic sac on right side containing clear fluid

with solid component of 3x4cm size in it suggesting it to be testis.(Fig.2)

(ii) Small 2x3 cm solid swelling on left side resembling testis.

Figure 2

Figure 2: Operative photograph showing a big cystic mass in right inguinal canal.

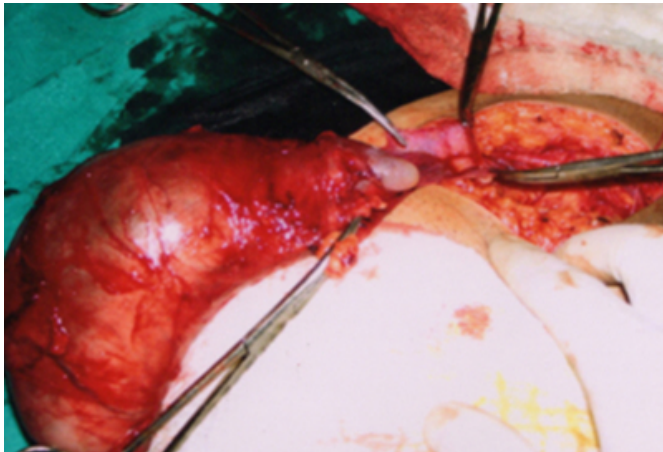


Figure 3

Figure 3: Photograph after final closure showing hypertrophied clitoris.



Both the specimens were sent for histopathology examination which showed a hydrocele sac with a tumor conforming to “seminoma” testis on right side. Normal/atrophic testicular tissue could not be identified, however, structure of vas deferens was seen. Sections from left swelling showed atrophic testis with a tumor conforming to embryonal cell carcinoma.

DISCUSSION

Sexual development is a complex process starting at fertilization of the ovum and continuing during gestation and

after birth and culminating with maturation at puberty. Intersex disorders involve conditions in which the appearance of the external genitalia is either ambiguous or at variance with the chromosomal or gonadal sex of the individual.² Patients with intersex disorders are categorized on the basis of their gonadal presentation into virilized females, undervirilized males, true hermaphrodites and mixed gonadal dysgenesis.

True hermaphrodites have both ovarian and testicular tissue(with germ cells) present in gonads. Ovary and testis occur together as ovotestis or there may be a normal gonad on one side & ovotestis on other or a testis on one side and an ovary on other side.³ Mullerian duct structures are usually present on the side of ovary but regress on side of testis whereas wolffian duct structures are intact on side of testis. External genitalia is variable. Most(70%)of these exhibit 46XX karyotype and in remainder it is 46XY(10%) or mosaicism(20%).³

Patients with mixed gonadal dysgenesis have dysgenetic gonads, retained mullerian structures, asymmetry of both internal & external organs and mosaic karyotype, most often 45X/46XY. ⁴ Many of these have a dysgenetic testis on one side and a streak gonad on other. In 46XY pure gonadal dysgenesis, mixed gonadal dysgenesis and dysgenetic male hermaphroditism, there is a remarkable propensity of the gonads to develop malignant tumors. Gonadoblastoma is the tumor most commonly seen in dysgenetic gonad.⁴ The seminoma-dysgerminoma can also occur and is most commonly seen in streak gonad in mixed gonadal dysgenesis.⁵

The patient in consideration was a 56 year old married female who had either never bothered about her genital appearance (possible in this part of the world with low literacy levels) or had never reported because of the social stigma attached to the condition of intersex; latter possibility seems to be more likely. We could not ascertain the presence of ovarian tissue histologically as that would have required a laparotomy /laparoscopy for which the patient was unwilling. However since patient was having feminine appearance, normally developed secondary sex characters as female like breasts and her CT Scan speculated upon the presence of atrophic ovaries alongside the rudimentary uterus, we assume that patient was having presence of both ovarian and testicular tissues simultaneously.

We are reporting this case as the patient in consideration was a true hermaphrodite according to available evidence and the

finding of bilateral testes and bilateral ovaries in a same patient is unreported in literature. Moreover, the patient had two different primary malignancies which again is very rare.

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References

1. Melton L. New perspectives on the management of intersex. *Lancet* 2001;357:2110
2. Hrabovszky Z, Hutson JM. Surgical treatment of intersex abnormalities : a review. *Surgery* 2002;131:92-104
3. Nihoul Fekete C, Lortat Jacob S, Cachin O, Josso N. Preservation of gonadal function in true hermaphroditism. *J Pediatr Surg* 1984;19:50-5
4. Donahoe PK, Crawford JD, Hendren WH. Mixed gonadal dysgenesis, pathogenesis, and management. *J Pediatr Surg* 1979;14:287-300
5. Salle B, Hedinger C. Gonadal histology in children with male pseudohermaphroditism and mixed gonadal dysgenesis. *Acta Endocrinol* 1970;64:211-227

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