



Recurrent orchitis in a patient with true hermaphroditism

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BACKGROUND

Ovotesticular Disorder of Sex Development (OTDSD - true hermaphroditism) is rare, characterized by the presence of both testicular and ovarian tissue. Usually, these patients seek medical attention due to ambiguous genitalia.

CASE PRESENTATION

A 15-year-old boy, with "atypical" genitalia and breast enlargement came for surgical correction. His genitalia had a more masculine aspect at birth and he had been submitted to six corrective surgeries. Karyotype is 46,XX[20]. Since 10 years of age he is having recurrent episodes of painful testicular swelling and gynecomastia. Tanner stage G4P3 and breast enlargement (T4), hyperpigmented scrotum with palpable gonads and a 6cm long penis with distal hypospadia. Lab work-up: Estradiol = 73.4 pg / mL (<20), Pubertal LH and FSH levels. Testosterone = 156 ng/dL.

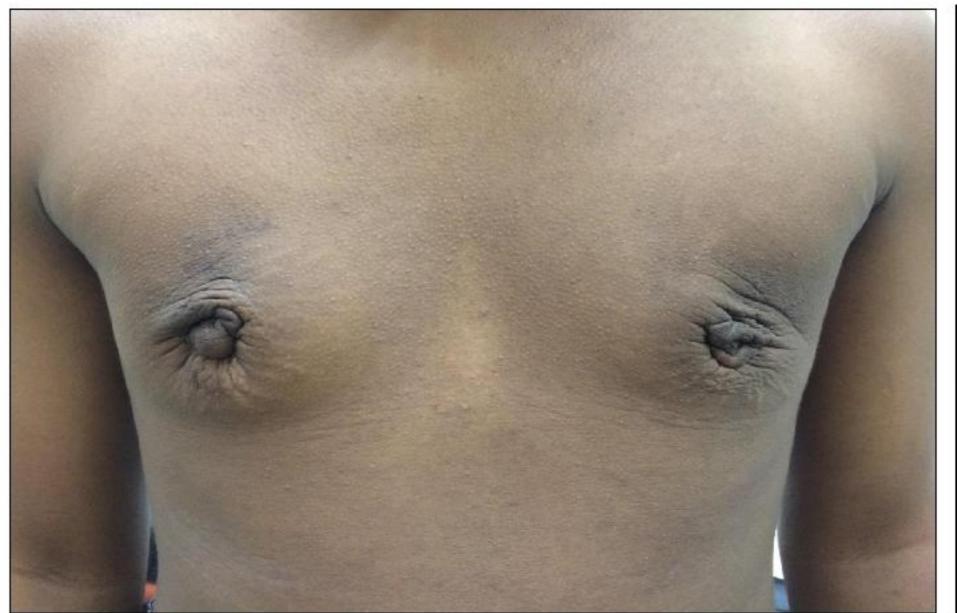






Images showing sexual characteristics before surgery

Ultrasound revealed testes with microlitiasis, bilateral hydrocele and cysts in the left testis. A structure which could resemble a rudimentary uterus or vaginal fornix was also shown. Bilateral mastectomy and a laparoscopy was performed in order to explore the gonads, which turned out to be *ovotestes*. The left gonad was totally excised, whereas in the right one, the macroscopic testis component was preserved.





Images showing sexual characteristics before surgery

CONCLUSION

In OTDSD, the ovarian portion of the gonad in the scrotum may enlarge during the ovulatory phase, which may be misdiagnosed as "orchitis", but the cyclical nature of the episodes should raise the possibility of ovarian tissue present in the gonad. In the ovotestis, the testicular component may be dysgenetic, opposing to the usual normal function the ovarian portion, which favors maintenance of the ovarian portion when possible. In our patient, the social male gender was well established and led to testicular preservation. The possibility of neoplastic degeneration is minimized once Y chromosome is not present but follow-up is mandatory, with the dosage of markers of malignization.



