

SPLENIC-GONADAL FUSION AND ADRENAL CORTICAL REST ASSOCIATED WITH BILATERAL CRYPTORCHISM

ALEX E. FINKBEINER, M.D.

PAUL A. DERIDDER, M.D.

SALLY E. RYDEN, M.D.

From the Departments of Urology and Pathology,
University of Michigan Medical Center, and
Wayne County General Hospital, Ann Arbor, Michigan

ABSTRACT — A patient with bilateral cryptorchism had ectopic splenic tissue adherent to his left testicle and an adrenal cortical rest adherent to his right testicle. The embryologic basis for these disorders is discussed and the literature reviewed.

Either ectopic splenic or adrenal tissue is occasionally found in the scrotal compartment or adherent to the gonadal-mesonephric structures. Herein we report the first case of both entities occurring in the same patient.

Case Report

This four-year-old white male presented with bilateral cryptorchism. The patient had a past history of ventricular septal defect with probable spontaneous closure, asthma, and multiple episodes of pneumonia. On physical examination the testicles were absent from a slightly hypodeveloped scrotum, and they were not palpable in the inguinal canal. Results of renal function tests, electrolytes, and urine analysis were all normal. A buccal smear revealed a male pattern, and intravenous pyelography was normal. An outpatient trial of 16,000 I.U. of human chorionic gonadotropin did not induce testicular descent.

Consequently, in August, 1974, surgical exploration was performed. Simultaneous bilateral inguinal exploration revealed both canals to be empty except for rudimentary vasa deferentia. These were followed into the retroperitoneum where testicular tissue was noted to be invaginated within the posterior peritoneum bilater-

ally. On the right side the testicle, epididymis, and vas deferens appeared normal. A 2 by 2 mm. nodule was noted to be adherent to the tunica albuginea of the right testicle. This nodule was excised and on frozen section was interpreted to be adrenal rest tissue. An orchiopexy was then performed on the right side. The left testicle was grossly abnormal in appearance and consistency, and appeared to be about twice the size of the right testicle. The left gonad structures were excised. Postoperatively the patient had an uneventful course.

On histologic section, the 2 by 2 mm. nodule adherent to the right testicle was composed of columns of orderly lipid-filled clear cells consistent with adrenal cortex (Fig. 1A). No renal medullary elements were present. The left gonadal structures consisted of a 1-cm., spherical accessory spleen, with a fibrous capsule and fibrous trabeculae throughout, with well-developed red and white pulp and prominent germinal centers (Fig. 1B). On gross inspection this splenic tissue had been thought to be the testis. Also present was a very small, immature testis with poorly developed tubules (Fig. 1C). The splenic capsule and tunica albuginea were joined by loose connective tissue. The epididymis and vas deferens were also small and immature.

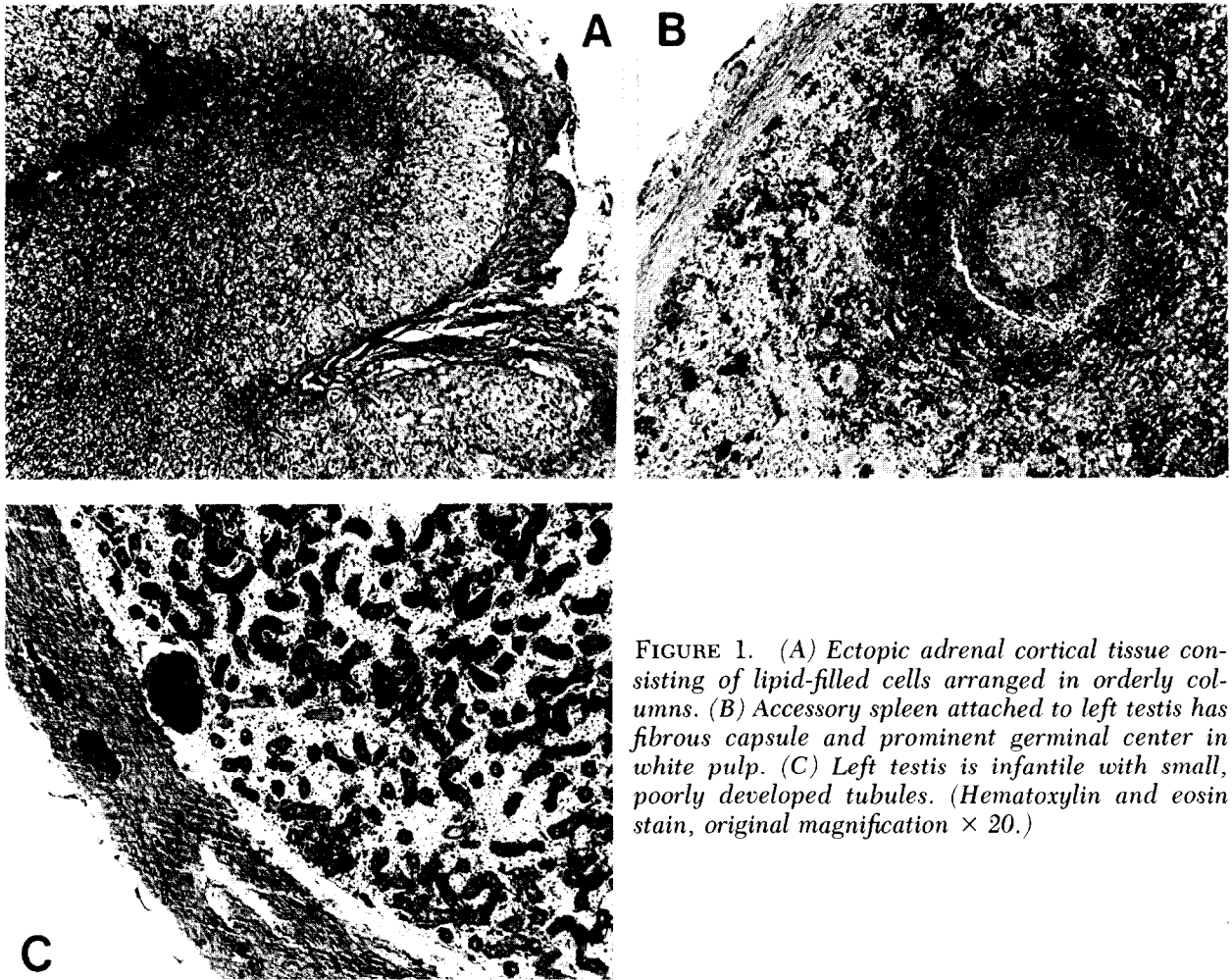


FIGURE 1. (A) Ectopic adrenal cortical tissue consisting of lipid-filled cells arranged in orderly columns. (B) Accessory spleen attached to left testis has fibrous capsule and prominent germinal center in white pulp. (C) Left testis is infantile with small, poorly developed tubules. (Hematoxylin and eosin stain, original magnification $\times 20$.)

Comment

Splenic-gonadal fusion

The incidence of accessory spleens varies between 11 and 35 per cent.¹⁻³ They are most often found in the immediate neighborhood of the spleen, but they may also be found in the upper omentum, gastrocolic ligament, mesocolon, or pancreas. Only rarely are they found in other intra-abdominal locations or in the scrotum.² Splenic-gonadal fusion is an abnormal connection between the spleen and the gonads or mesonephric structures. Scrotal spleens are generally within the tunica vaginalis and separated from the testicle and epididymis by a discrete capsule.⁴ At least 68 cases of splenic-gonadal fusion have been reported;⁵ it is about ten times more common in males than females.⁶ Nine cases of incomplete descent of a fused splenic-gonadal structure have been reported.

Embryologically splenic-gonadal fusion can be explained as follows: the spleen develops as a

trilobed condensation of mesoderm which later fuses into a single mass. The splenic anlage is visible in the five-week embryo as a proliferation of surface peritoneal cells on the left side of the dorsal mesogastrium. The cells break free and invade the underlying mesenchyma where they differentiate as spleen. Later the splenic anlage comes into close proximity with the left urogenital fold which includes gonadal mesoderm and mesonephric components. The gonad begins its descent about the eighth week and moves caudally during the eighth to twelfth week; thus fusion must occur between the fifth and eighth week. This path of descent is sometimes visibly demonstrated by a band of tissue (fibrous or splenic).^{2,4,5}

Splenic-gonadal anomalies are divided into two types: continuous — a cord of either splenic or fibrous tissue extends from the spleen to the left gonadal-mesonephric structures; and discontinuous — the ectopic splenic tissue presents as a distinct encapsulated mass adjacent to the

gonadal-mesonephric structures with no connection to the main splenic tissue.⁵⁻⁸

Histologically, ectopic splenic tissue may resemble normal spleen, or may show retrogressive changes of fibrosis, thrombosis, or calcification. The associated testicular tissue may be normal or exhibit changes ranging from mild atrophic changes to absence of spermatogenesis.^{1,3,8}

Splenic-gonadal fusion always occurs on the left side^{4,7} and is often associated with indirect inguinal hernias.^{4,7} The relative high incidence of associated cryptorchism suggests that fusion may interfere with normal gonadal descent.^{4,8}

Splenic-gonadal fusion has been noted incidentally at autopsy,⁹ herniorrhaphy,¹⁰ and when performing an orchiopexy for cryptorchism.⁵ Clinically it may present as an asymptomatic or painful, swollen scrotal or testicular mass at all ages.^{1-4,7-9,11} One patient presented with abdominal pain which proved to be secondary to bowel obstruction from extrinsic compression of the colon by a continuous splenic band extending from the spleen to the inguinal canal.¹⁰ Thus, splenic-gonadal fusion frequently presents a diagnostic problem requiring surgical exploration.

A review of 37 cases of splenic-gonadal fusion revealed that 8 of these patients had severe ectromelia (hypoplasia of the extremities), and 4 of these 8 had micrognathia as well. It is postulated that a single intrauterine event is responsible for all three of these congenital anomalies, since the extremity buds and mandibular cartilage form during the time corresponding to fusion of the spleen and testicle.^{6,8,12}

Although this patient had no congenital limb or facial anomalies, he did have a ventricular septal defect demonstrated by cardiac catheterization. Since the interventricular foramen normally begins to close by the end of the seventh week of fetal life, perhaps one embryologic event also induced this patient's gonadal and cardiac anomalies.

Adrenal cortical rests

Schechter¹³ states that adrenal cortical rests occur in 50 per cent of newborns and usually atrophy and disappear within a few years. In 100 males less than one year of age adrenal cortical nodules were found in or near 15 (7.5 per cent) of 200 testes examined at necropsy.¹⁴

Gualtieri and Segal¹⁵ describe four types of aberrant adrenal tissue: (1) Adrenal heterotopia

which is a developmental inclusion of the entire adrenal or a large portion of the adrenal beneath the capsule of the kidney or liver. This usually consists of cortical tissue and is always bilateral. (2) Accessory adrenal whole gland tissue consists of whole gland rests (cortex and medulla) which may be found in various locations. (3) Accessory adrenal chromaffin tissue which is medullary ectopia. (4) Accessory adrenal cortical tissue which appears to be fairly common near the kidney but may be found anywhere in the retroperitoneum or near the genitalia.

Embryologically the adrenal gland has a double origin: the cortex arises from coelomic mesodermal epithelium and the medulla from chromaffin ectodermal cells of the neural groove. The adrenal primordia and primitive gonads appear in the vicinity of the mesonephros during the fourth week of fetal life. At this time the adrenal primordia lie at the base of the dorsal mesentery, medial to the cephalic pole of the mesonephros, and the primitive gonads form ventral to the mesonephros immediately adjacent to the adrenals. The cluster of cells of the adrenal primordium does not become encapsulated until the sixth week, at the time of separation of the adrenal gland from the gonad. As the gonads descend, adrenal cortical tissue which may adhere to the gonad becomes separated from the main adrenal body and descends with the gonad. Ectopic adrenal cortical tissue can thus be expected anywhere along the course of the gonadal vessels.¹³

Accessory adrenal cortical tissue found along the course of wolffian ducts or their adult derivative structures has been described frequently.¹⁶ This tissue has been found along the course of the gonadal vessels, within the broad ligament, connective tissue of the inguinal canal, spermatic cord, canal of Nuck, uterus, hernia sacs, and epididymis or tunica albuginea, and adherent to the ovary or testes.¹³⁻¹⁶ Adrenal cortical tissue has never been found within testicular or ovarian parenchyma.¹³⁻¹⁵ Histologically, medullary cells are absent and the tissue consists mostly of zona fasciculata or reticularis although some rests may contain glomerulosa as well.¹⁵

These cortical rests may have clinical significance if they present as a scrotal mass; aberrant nodules may undergo compensatory hypertrophy after adrenalectomy; heterotopic implants may be excised inadvertently resulting in adrenal insufficiency; and aberrant masses may themselves become neoplastic.¹³

Department of Urology
University of Arkansas College of Medicine
Little Rock, Arkansas 72201
(DR. FINKBEINER)

References

1. Lynch JB, and Kareim OA: Aberrant splenic tissue in the scrotum, *Br. J. Surg.* **49**: 546 (1962).
2. Emmett JM, and Dreyfuss MI: Accessory spleen in the scrotum, *Ann. Surg.* **117**: 754 (1943).
3. Daniel DS: An unusual case of ectopic splenic tissue resembling a third testicle, *ibid.* **145**: 960 (1957).
4. May JE, and Bourne CW: Ectopic spleen in the scrotum: report of 2 cases, *J. Urol.* **111**: 120 (1974).
5. Mizutani S, Kujohara H, and Sonoda T: Splenic-gonadal fusion in a Japanese boy, *ibid.* **112**: 528 (1974).
6. Sommer JR: Continuous splenic-gonadal fusion with ectromelia: teratogenesis, *Pediatrics* **22**: 1183 (1958).
7. Mendez R, and Morrow JW: Ectopic spleen simulating testicular tumor, *J. Urol.* **102**: 598 (1969).
8. Putschar WGJ, and Manion WC: Splenic-gonadal fusion, *Am. J. Pathol.* **32**: 15 (1956).
9. Keizur LW: Accessory spleen in the scrotum: report of two cases, *J. Urol.* **68**: 759 (1952).
10. Watson RJ: Splenogonadal fusion, *Surgery* **63**: 853 (1968).
11. Bennet-Jones MJ, and St. Hill CA: Accessory spleen in the scrotum, *Br. J. Surg.* **40**: 259 (1952).
12. Hines JR, and Effum PR: Splenic-gonadal fusion causing bowel obstruction, *Arch. Surg.* **83**: 887 (1961).
13. Schechter DC: Aberrant adrenal tissue, *Ann. Surg.* **167**: 421 (1968).
14. Dahl EV, and Bahn RC: Aberrant adrenal cortical tissue near the testis in human infants, *Am. J. Pathol.* **40**: 587 (1962).
15. Gualtieri T, and Segal AD: Report of a case of adrenal-type tumor of the spermatic cord: a review of aberrant adrenal tissues, *J. Urol.* **61**: 949 (1949).
16. Freeman A: Adrenal cortical adenoma of the epididymis, *Arch. Pathol.* **39**: 336 (1945).