SPLENOGONADAL FUSION—A RARE CONGENITAL ANOMALY DEMONSTRATED BY ^{99m}Tc-SULFUR COLLOID IMAGING: CASE REPORT

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A case report illustrating the value of spleen scanning in the diagnosis of a rare anomaly characterized by fusion of the spleen and left testis is described. This malformation results from faulty organogenesis during the fifth to the eighth week of fetal development. Only 65 cases have been reported in the world literature. In most instances, the anomaly is recognized as an incidental finding at autopsy or at surgical exploration of the abdomen. In 20% of cases the anomaly is associated with osseous malformations such as peromelia, ectromelia, micrognathia, and talipes.

CASE REPORT

A 14-month-old black male infant was admitted because of unexplained high fever. Significant findings on physical examination were hemimelia and ectromelia, undescended left testis, and redundant prepuce with severe phimosis (Fig. 1). The spleen was not palpable.

X-ray films of the chest were negative. An intravenous pyelogram showed normal findings. One urine examination showed E. coli. A complete blood cell count (CBC) showed normal values.

Because of the frequent association of splenogonadal fusion with left cryptorchism and limb malformations, splenic scan was done to verify the presence of the anomaly.

METHOD

Using ^{99m}Tc-sulfur colloid, a dose of 200 μCi was administered intravenously. The scan was performed with an Ohio-Nuclear 54FD scanner at an information density of 500 counts/cm^2 employing medium-energy 85-hole collimators.

The scan showed a normal liver but the spleen configuration was very interesting. The inferior border of the spleen tapered into a narrow tail, extending into the pelvis towards the region of the internal inguinal ring, indicating fusion between the spleen and the undescended left testis (Fig. 2).

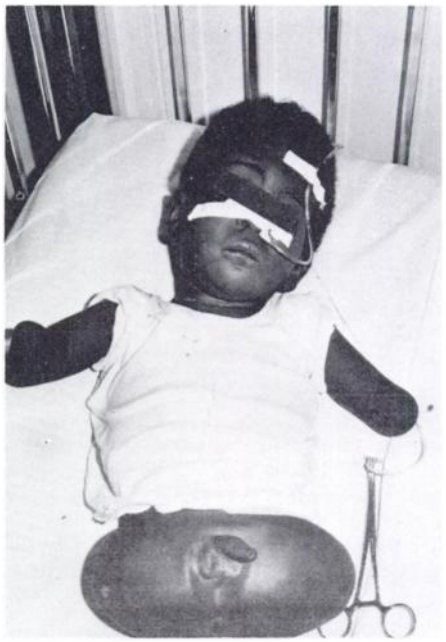

FIG. 1. Black child (14 months old) with hemimelia.

DISCUSSION

Bostroem first mentioned the anomaly in 1883 (*1*). It wasn't until 1889, however, that Pommer (*1*) published a detailed description of a case. Of the total cases reported, all concerned white people except for one Arab, two Orientals, and four blacks. The youngest subject was stillborn and the oldest one was 69 years old. Of all the cases 25% were

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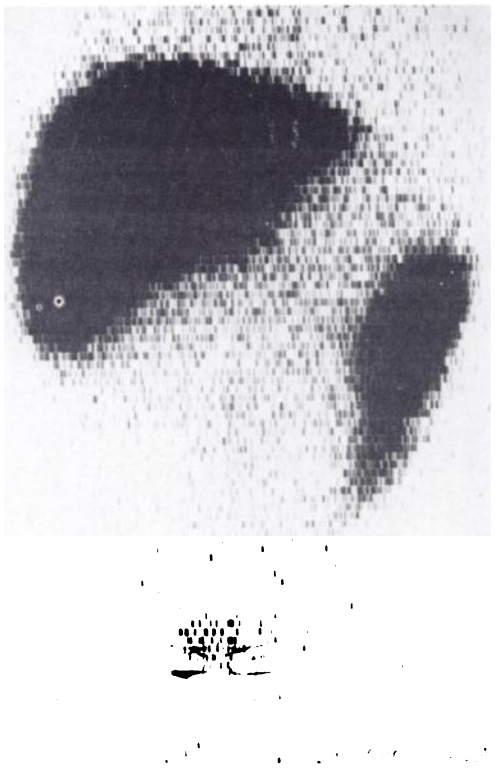

FIG. 2. Anterior rectilinear scan of liver and spleen showing splenogonadal fusion.

incidental findings at autopsy and in 20% there were associated extremity defects. Two forms of the anomaly, continuous and discontinuous, were described by Putschar (*1*) in 1956. The first showed a cord-like extension of splenic tissue connecting the main spleen to the left testis or ovary. In the second variety there was no communicating cord and this probably represented a true accessory spleen.

The cause of abnormal fusion of spleen and testicle is obscure. It may be due to the action of a teratogenic factor during the fifth to the eighth week of fetal life when the spleen and gonads are simultaneously differentiating. During the week when the proliferating gonad separates from the mesonephros, the splenic primordia project above the surface of the dorsal mesogastrium in the form of hillocks. The latter coalesce to form the splenic bud which pushes out laterally to lie in close proximity with the differentiating gonad. It is postulated that faulty union takes place at this time. The occurrence of associated osseous malformations in the form of micrognathia, peromelia, and ectromelia can be explained on this basis. The Meckel's cartilage, which is the anlage of the mandible, and the proximal buds of the extremities also undergo differentiation during the sixth and seventh weeks.

No correct preoperative diagnosis was made in any of the cases reported. Heitzman (*2*) recognized the clinical anomaly in 1917 and is thought to be the first to do so. While operating on a left inguinal hernia, he noticed a scrotal mass that on further abdominal dissection showed connection with the main spleen.

Suspicion of splenic tissue in the left scrotum should be considered in the differential diagnosis when dealing with malignancies of the testes, inflammatory conditions such as epididymitis and epididymo-orchitis, hematoceles with history of injury, hydroceles, torsion of the cord, and diseases affecting testicular embryologic remnants. A splenic cord distinct from the vas deferens may indicate splenogonadal fusion. Careful examination, therefore, is valuable, especially where there is suggestion of multiplicity, i.e., a third testicle.

Patients with cryptorchidism as well as those having vague left scrotal masses with associated inguinal hernia and variegated limb or osseous malformation should be examined for the anomaly. Scrotal exploration can be avoided in a few cases if preoperative scanning using ^{99m}Tc-sulfur colloid is done. Heterotopic splenic masses can be demonstrated easily by this method. The knowledge that a benign condition exists may make salvaging of testicles more likely.

CLINICAL CONSIDERATIONS

Splenogonadal fusion has been reported to cause intestinal obstruction. Hines, et al (*3*) operated on a 15-year-old patient with ectromelia who had a band of splenic tissue causing obstruction of the colon proximal to the splenic flexure.

Unaware of the nature of the left scrotal masses, many surgeons have sacrificed a good testis because they lacked a proper preoperative diagnosis. Surgeons ignorant of the nature of the scrotal mass and fearing malignant degeneration have performed radical orchiectomy on salvageable testes.

Cases of pain and scrotal swelling have been observed in malaria (*4–5*). Alleviation of symptoms followed antimalarial treatment.

Splenic tissue abnormally located in the body behaves as does the spleen itself; therefore, in hypersplenism and other blood dyscrasias requiring splenectomy, heterotopic splenic tissue whether in the pelvis or in the scrotum should be carefully sought and removed to effect complete cure (*6*). Technetium-99m-sulfur colloid scanning is an invaluable tool in locating abnormally located splenic tissue.

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