### ACCESSORY SPLEEN IN THE SCROTUM

REVIEW OF LITERATURE ON ECTOPIC SPLEENS AND THEIR ASSOCIATED SURGICAL SIGNIFICANCE

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THE PRESENCE of an ectopic spleen in the scrotum, simulating a testicular tumor, is a rare enough finding to warrant recording.

Case Report.—A white male, age 47, was admitted to the Surgical Service of the Chesapeake and Ohio Hospital on March 3, 1942.

About seven months previously, he had noticed, for the first time, a small lump in the right groin. It did not give him discomfort, but he was advised that he had a hernia which should be repaired.

He was known to have a diverticulosis of the colon which caused frequent attacks of abdominal pain. Otherwise, his family and personal history were irrelevant.

Both inguinal rings were patulous and a definite hernial sac protruded on the right side. A smooth, round, nontender, firm mass was found attached to the upper pole of the left testicle. It was about half as large as the testicle, which seemed normal in size and consistency. This swelling had never been noticed by the patient. Preoperative Diagnosis: Indirect inguinal hernia, bilateral. Tumor of left testicle.

Operation.—The right inguinal hernia was repaired; and the left scrotum was incised and the testicle delivered. The tumor was found to be attached, by a broad base, to the tunica albuginea at the upper pole close to the head of the epididymis. It was encapsulated and did not penetrate into the testicle. It was removed by sharp dissection. The testicle, the epididymis and the spermatic cord appeared normal.

Pathologic Examination.—The specimen was almost completely covered with a dense, pearly-grey, glistening capsule. It measured 4 x 3.5 x 2 cm. It was firm and elastic. The cut sections showed brownish-red tissue, with an irregular network of greyish, fine trabeculae. The microscopic sections showed normal splenic tissue, with rather small follicles, and hyperplastic red pulp. The sinuses were almost bloodless and collapsed (Fig. 1).

The patient made an uneventful recovery and was discharged three weeks after operation.

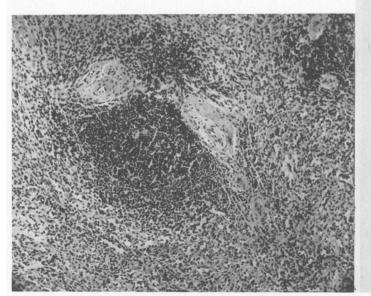
Comment.—Puzzling as such a location of an ectopic spleen may appear on first sight, it will be readily understood if one considers the close topographic relation between spleen and urogenital organs during early intrauterine life. The differentiation of the spleen begins in about the fourth to fifth week. It is completed in the twelfth week. During this time, the spleen is transformed from a trilobated into a single organ. It is situated between the mesonephric and the urogenital fold. The mesonephros (wolffian body) reaches its maximal cephalad extension at the time when the spleen begins to develop. The caudal migration of the genital gland begins in the eighth to tenth week. Therefore, we must place the teratogenetic period, during which portions of the spleen are transported downwards with the genital gland, between the fourth and tenth week; most probably in the sixth

to eighth week (Putschar<sup>20</sup>) because during this time the splenic tissue is already well developed and is still in close contact with the wolffian body. Splenic tissue then accompanies the genital gland in its descensus and is finally found in the scrotum in close association with the upper pole of the left testicle and the epididymis or, in the female, in the region of the mesosalpinx and the mesovarium. The path of this migration is, in some cases, visibly demonstrated by a band of tissue—consisting of fibrous and/or splenic tissue—which connects the normally located spleen with the ectopic spleen in the scrotum or the broad ligament. This band runs from the splenic hilus anteriorly to the intestines and ends near the testicle or the mesosalpinx. A nodule of splenic tissue has been seen, in some cases, at the distal end (Fig. 2).

Putschar<sup>20</sup> has collected several cases with such bands between the spleen and genital organs. Since then, another such case has been published by Fischer and Gissel<sup>7</sup>:

Case of Fischer and Gissel.7—The patient was a 13-year-old boy who was operated upon for an undescended testicle in the left inguinal canal. A flat, further upward, round band extended from the epididymis into the left side of the abdominal cavity. It was followed for 20 cm. It was as thick as a knitting needle, smooth and dark red. The lowermost 8 cm. were removed and found to consist of typical splenic tissue.

The authors express the opinion that this malformation occurs more frequently than the recorded cases would indicate but that it is either erroneously interpreted or not published.



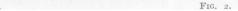


Fig. 1.—Photomicrograph of section from paratesticular tumor showing characteristic splenic tissue. Fig. 2.—Band of splenic tissue connecting the normally located spleen, with an accessory spleen in the scrotum. (Redrawn from Sneath.<sup>23</sup>)

The literature contains, in addition, several reports of cases in which—like in our own case—the ectopic spleen in the scrotum was not connected with the spleen.

Case of Finaly. The patient was a male newborn child with a congenital left inguinal hernia. The left testicle seemed twice as large as the right and had a different shape. A dark brown and purple, round swelling at the head of the epididymis was removed. It was covered with peritoneum and consisted of splenic tissue.

Case of Talmann.<sup>24</sup>—The patient, a 22-year-old soldier, had a painful swelling in his left scrotum. During malarial attacks this swelling reached the size of a goose egg. One nodule of splenic tissue, 2.5 cm. in diameter, was found at the head of the left epididymis, another, 0.4 x 0.5 x 0.8 cm., was present in the spermatic cord.

Case of Osseladore.<sup>17</sup>—The patient, a 19-year-old white boy had, since birth, three nodules in his left scrotum which had grown corresponding to the body development. The largest nodule suggested at the time of operation, as to size and consistency, a third testicle. The two smaller ones were pedunculated. All three consisted of splenic tissue which was, in places, sclerotic and contained several iron-calcium deposits.

Case of Settle.<sup>22</sup>—The patient, an adult, suffered from a painful enlargement of the left testicle during the acute febrile phases of a tertian malaria. An enlarged accessory spleen was found in the scrotum.

Even these few cases illustrate the almost impossible task of a correct preoperative diagnosis. Probably the true condition will only be recognized after microscopic study. However, in the presence of a long-existing tumefaction of the left testicle, particularly when accompanied by an inguinal hernia, the possibility of an ectopic spleen should be considered in the already difficult diagnosis of testicular tumors. (The painful periodic swelling associated with malarial attacks in the cases of Settle,<sup>22</sup>, and Talmann,<sup>24</sup> might perhaps have led to hazarding a tentative diagnosis).

Although it was not possible to inspect the spleen in the present case, we may conclude, from the other similar reports, that a normal spleen was present in normal location. Thus, we would list this case of scrotal spleen among the accessory aberrant) spleens. A certain confusion prevails as to the correct nomenclature of spleens in anomalous location. A clear distinction should be made between the following types: (1) Wandering spleen. (2) Double spleen. (3) Multiple spleen. (4) Accessory (aberrant) spleen. These types differ from each other in their formal genesis as well as in their clinical significance.

The wandering or floating spleen is the type most frequently involved in surgical complications. Since Abell¹ reported 95 cases of wandering spleen, with torsion of the pedicle, additional publications have brought the total to about 120. The wandering spleen may be found in any part of the abdominal cavity and—as in ventral or diaphragmatic hernia—even outside the abdomen. It is much more frequent in women than in men, and its incidence is especially high in multiparae. Only three cases have been reported in children (Truesdale and Freedman²6). It is, therefore, assumed that the relaxation of the abdominal wall plays an important part in the causation of the displacement, although this is probably not the only factor involved. Minor, easily overlooked disturbances of peritoneal development, and consequent faulty attachment of the organ, are most likely equally responsible. The relative frequency

with which wandering spleen is found in malaria, is unexplained. The weight of these spleens is far below that found in leukemia, Banti's or Gaucher's disease, and no case of wandering spleen associated with any of these conditions has been observed.

Symptoms usually are due to torsion of the vascular pedicle. The symptomatology varies according to the location of the spleen and is that of the torsion of the pedicle of an intra-abdominal tumor. Transitory attacks of colicky pain have occasionally occurred preceding the final attack.

Only in rare instances, has the wandering spleen, as such, caused complications which necessitated surgical relief or interfered, after torsion had occurred, with the function of other organs. A wandering spleen in the pelvis acting as an obstetric impediment has been described by Ottow.<sup>18</sup> Intestinal obstruction has been reported by Mauro,<sup>15</sup> Pacchini,<sup>19</sup> Salvin,<sup>21</sup> and Harris.<sup>10</sup> Cases of gastric volvulus have been published by Lamarque, *ct al.*,<sup>13</sup> Bertone,<sup>5</sup> and Zhdanovich.<sup>28</sup> A wandering spleen was the cause of chronic dyspepsia in the case of Hjort.<sup>11</sup>

Splenectomy is the only method of treatment. Conservative measures, as attempting to untwist the pedicle, are contraindicated. The results are satisfactory, although the mortality rate in pregnant women is considerable.

Double spleen is exceedingly rare. In this condition, two, as to size and shape, equivalent spleens are found in place of the single organ. A discussion as to the origin of this malformation is beyond the scope of this paper. The reader is referred to the monograph of Putschar.<sup>20</sup> The literature contains no reference of double spleens of clinical significance.

We speak of multiple spleen when we find, instead of a single spleen, a varying number of small spleens (3–10) in the left hypochondrium or scattered over the visceral or parietal peritoneum of the entire abdominal cavity (up to 400). They may be due to faulty development of the multiple splenic anlage or to multiple autotransplants after a (intra-uterine) trauma to the spleen. Krueger and Mast, <sup>12</sup> and Hambrick and Bush, <sup>9</sup> have recently described multiple splenic implants after splenic injury and splenectomy, and have reviewed the literature. The multiple spleens are of little clinical significance. However, Leriche and Gravier have reported the torsion of the pedicle in one of several multiple spleens, complicated by rupture and intra-abdominal hemorrhage, and treated by multiple splenectomy.

By far the most frequent type are the accessory (aberrant, supernumerary) spleens in the presence of a normally formed spleen in correct position. They are found in about ten per cent of all autopsies. Their number varies, and is rarely over 40. Most of them are found in the immediate neighborhood of the spleen but they are encountered also in the upper omentum, the gastrocolic ligament, the mesocolon, and in the pancreas. Only very rarely are they seen in other intra-abdominal locations or in the scrotum. Their size varies from that of a pinhead to four centimeters in diameter. They are usually sessile but some are pedunculated. They rarely produce clinical

symptoms by torsion of the pedicle (Alexander,<sup>2</sup> and Alexander and Romanes,<sup>3</sup> Geiger,<sup>8</sup> and Settle<sup>21</sup>) or cause intestinal obstruction (Bainbridge,<sup>4</sup> Voss,<sup>27</sup> and Temoin<sup>25</sup>) or both (Settle<sup>22</sup>). A curiosity is the case of Morris, Lederer and Fradkin.<sup>16</sup> Their patient had his spleen removed for thrombocytopenic purpura. A walnut-sized accessory spleen was left in the abdomen. The patient was well only for a short while but then the blood dyscrasia returned due to the action of the accessory spleen. Considerations along this line would make the search for accessory spleens important, also, where splenectomy for hemolytic jaundice is performed.

#### SUMMARY

A case of accessory spleen in the scrotum is reported and the genesis of the malformation discussed. Similar cases in the literature are summarized. The various types of spleens in anomalous location are surveyed and their clinical importance evaluated.

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