

THE CLINICAL IMPORTANCE OF ACCESSORY SPLEENS*

(WITH REPORT OF A CASE)

BY ROBERT F. ROBERTSON

Montreal

THE frequency with which the pathologist encounters accessory spleens at autopsy makes it rather remarkable that their presence is rarely noted by the operating surgeon. Orth (quoted by Alexander and Romanes³) mentions as many as twenty, and Albrecht² reported one case of 400 in various locations in the peritoneal cavity. Faltin (quoted by Kaufmann⁹) also reports numerous accessory spleens over the peritoneum six years after splenic contusion, and Küttner (quoted by Kaufmann⁹) found as many as 1,000, five years after a gunshot wound, in which the spleen itself had been injured. According to Adami and Nicholls,¹ accessory spleens are found in 11 per cent of autopsies. Helwig⁷ states that at autopsy he has found them in 10 per cent of cases. Distinction between real splenic substance and hæmo-lymph glands is here not always well established.

The location of accessory spleens varies. According to Schilling¹⁵ the most frequent sites are: (1) the hilus of the spleen; (2) gastro-splenic ligament; (3) splenocolic ligament; (4) pancreatico-splenic ligament; (5) great omentum; (6) along the blood vessels of the spleen, and (7) gastrohepatic omentum. In Albrecht's case the normal spleen was in its usual position, the size of a walnut, with the splenic artery and vein in their normal position; the other spleens were scattered not only in the mesogastrium but also on the peritoneum, the largest number being found on the mesentery and transverse mesocolon and more than 30 in Douglas' pouch. Each of these spleens was enclosed in a separate capsule covered by peritoneum and exhibited the gross and microscopic structure of splenic tissue. Voss¹⁸ reports having found an accessory spleen, 2 cm. in diameter, on the transverse colon, 2.5 cm. from the splenic flexure. Two similar cases have been reported by Hance⁶ and Sneath¹⁶ in which an accessory spleen was found in the inguinal canal and, in Sneath's case, in the scrotum. DeTeyssieu⁵ has reported a very

unusual case in which normal splenic tissue has been found embedded in the liver. One was found by Traver¹⁷ also in the body of the pancreas.

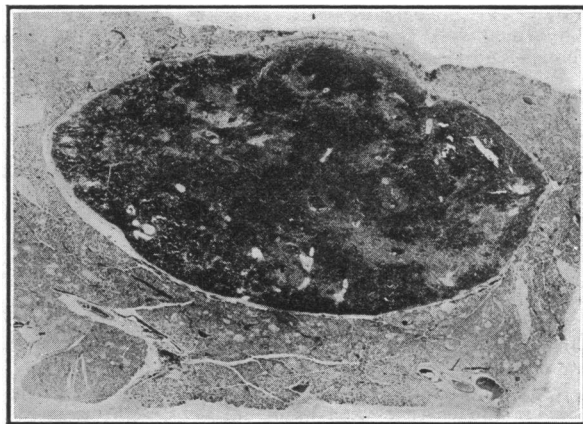


Fig. 1

The specimen here described was found at autopsy at the McGill Pathological Institute. Grossly, a bright reddish-brown oval-shaped nodule, 1 by 0.5 cm., was seen embedded in the tail of the pancreas. Microscopically, it was composed of typical splenic tissue which was completely surrounded by a connective-tissue capsule. Pancreatic tissue was found to surround the splenic nodule on all sides except in a small area where the capsule of the spleen and pancreas fused. Throughout this area were nests of pancreatic cells in irregular patches within the capsule. In the splenic nodule the Malpighian corpuscles were well formed and appeared relatively prominent, the central arteries in many cases having undergone hyaline transformation. The pulp was rather scarce in pulp cells but engorged, with well-preserved red blood cells. Tissue relations were well preserved throughout the entire nodule so that the picture indicated relatively normal spleen.

DISCUSSION

An accessory spleen is an anomaly. The explanations given by the various authors for its presence are essentially two—trauma and de-

* From the Pathological Institute of McGill University, Montreal, Professor Horst Oertel, Director.

velopment. Beneke (quoted by Kaufmann⁹) maintains that trauma, with implantation and later regeneration of splenic particles, is responsible for large numbers of accessory spleens. The question of the new formation of splenic substance from the peritoneum has been raised in this connection (Faltin). Helwig⁷ states that intrauterine trauma may be the cause in many cases. He reports two cases of multiple spleens, 14 and 10 in number, in which there was evidence of intrauterine trauma. In both, fibrous bands divided the main spleen into lobes. Also, there were numerous anomalous fibrous bands in the liver and gall-bladder region. One case showed also stenosis of the bile-ducts and peri-insular productive hepatitis, the other, a cardiac anomaly.

The majority of authors support the developmental theory as the most probable explanation for at least many cases of accessory spleens. Although the end-result is a mere displacement of normal splenic tissue, the time and conditions under which the changes take place are important factors in determining the final location of the accessory splenic tissue. Sneath's case¹⁶ is an interesting example of such a misplacement. Here the accessory spleen was in the scrotum, attached firmly to the upper pole of the testis and also to a "tail" of spleen by a fibrous band running from the inner aspect of the spleen, across the peritoneal cavity, down the left side to the left internal abdominal ring. Here it joined the spermatic cord and, passing through the inguinal canal, terminated in a bulbous portion, which was adherent to the upper pole of the testicle. The whole band was 33.4 cm. in length and contained 14.5 cm. of splenic tissue.

The testis in its original position is quite close to the spleen on the left side. Mere propinquity, however, though suggestive, does not completely account for the occurrence. The spleen is developed in the dorsal mesogastrium of the embryo, appearing first as a localized growth in the mesoderm of the mesogastrium at the beginning of the fifth week of fetal life. At this time the growth of the lung buds is forcing the stomach tailwards from its original cervical position, and it takes up its position (with the mesogastrium) level with the lower thoracic segments. At the same time the ridge on the inner side of the Wolffian body, from the hinder part of which the testicle develops, extends from the ninth to the twelfth thoracic segments, the

actual primitive genital gland lying in the iliac fossa.

How then is the association of the spleen (which develops in the dorsal mesentery of a portion of the foregut) with the testicle (which develops in the mesoderm adjoining the mesentery of the midgut) to be explained? Keith¹⁰ states that in the lower mammals the splenic formation spreads caudally, even into the mesentery of the hind-gut, to form the "colic lobe". A persistence of this condition in the human being explains the presence of accessory spleens in Douglas' pouch in Albrecht's case,² since the disappearance of the mesorectum would leave them stranded in this situation. It is not difficult to imagine also that a localized extension to the left of this caudal prolongation of splenic tissue might come into close relationship with the adjacent developing testicle. This relationship might readily extend to a secondary vascular connection, the vessels acting as a suspensory ligament and assuring the migration of the localized extension.

Accessory spleens are often associated with other congenital anomalies. Hyrtl (quoted by Pool¹⁴) cites four cases in which accessory spleens were associated with transposition of other viscera. Helly (quoted by Pool¹⁴) has reported a case in which the only other congenital anomaly was a bilobar spleen.

Warthin and Mayo (quoted by Alexander and Romanes³) maintain that if the main spleen is structurally as well as functionally deficient, there is a compensatory hyperplasia of accessory splenic tissue. This tissue must, of course, have been misplaced before hyperplasia could take place. Warthin cites a case in which there were a large number of accessory spleens with a very small "walnut-sized" major spleen. Mayo¹² states that accessory spleens are often found in the pedicle during splenectomy for tumours of the spleen.

In order to explain the presence of an accessory spleen in the tail of the pancreas, the development of the pancreas must be considered. It arises from two (or three) separate *Anlagen* one dorsal and one (or two) ventral. The dorsal arises as a ridge-like evagination from the dorsal wall of the gut cephalad to the level of the liver. The mass of cells grows into the dorsal mesogastrium and becomes constricted from the parent epithelium, except for a thin neck which becomes the duct of Santorini. One or two ventral diverticula appear and one develops and

becomes constricted, forming the duct of Wirsung. The smaller ventral pancreas grows to the right and dorsally and fuses with the dorsal head in the eighth week. The ducts join up, the duct of Santorini disappears, and with the change of position of the stomach the pancreas comes to occupy its final position. The head and duct of Wirsung develop from the ventral pancreas and the body and tail from the dorsal head. In the fifth week, when the dorsal head grows into the mesogastrium it comes to lie very close to the developing spleen, which is arising from the mesenchyma and coelomic epithelium of the left side of the dorsal mesogastrium. Thus a small amount of splenic tissue may become encapsulated in the developing pancreatic tissue. Further, one can readily see why an accessory spleen in the body and tail would be much more common than in the head, an observation recorded by others (E. Kaufmann⁹).

It has been said that accessory spleens are more often observed in young subjects. Rocher (quoted by Morrison *et al.*¹³) states that the diminished frequency of their occurrence in the adult is due to atrophy. Jolly⁸ reports having found accessory spleens in 25 per cent of a series of 80 children less than seven years old. Sassuchin (quoted by Morrison *et al.*¹³) observed them in 15 per cent of a series of 153 children under ten years of age. He found them much more abundant in infants.

Not until recently has much clinical significance been attached to accessory splenic tissue. However, of the cases that have been reported, a number are demonstrative of the fact that the clinical importance should not be overlooked. The following cases bear out this statement.

CASE 1. (Reported by Alexander and Romanes.³)

A woman, aged twenty-five, was subject to frequent attacks of pain of a dragging nature in the upper right quadrant of the abdomen which had no relation to the intake of food. The pain was worse when the patient stooped and was unaccompanied by other symptoms. This came on daily suddenly, lasting one to two hours, then passed off leaving no after-effects. At operation an accessory spleen, 2 cm. in diameter, was found attached to the lowest part of the great omentum and lying beneath the diaphragm. Microscopically, the organ resembled normal splenic tissue with the exception that the lymphoid follicles were not so well developed and there was considerable fibrosis of the whole organ with recent and old hæmorrhages.

The cause of the pain was regarded to have been congestion of the veins from torsion of the omentum, obstructing the return blood flow.

CASE 2. (Reported by Témoin, quoted by Morrison *et al.*¹³)

A female, aged thirty-five, had complained of a lump in the abdomen and pain in this region for eighteen

years. It was located in the midline just above the umbilicus. The patient had had attacks resembling those of intestinal obstruction, and in the late stages the mass seemed to have enlarged. At operation the "tumour" was found beneath the omentum and attached to it. The adhesions were separated and the "tumour" was seen to be lying closely against a loop of intestine. It did not form part of the wall. The "tumour" was found to be attached by a small pedicle to the mesentery.

The symptoms were attributed to some interference with the mesenteric circulation and to a kinking of the loops of gut to which the accessory spleen was attached, which gave rise to recurrent attacks of pain and vomiting suggesting intestinal obstruction.

CASE 3. (Reported by Voss.¹²)

This patient had complained of pain in the left hypochondriac region which had lasted for a number of years, gradually increasing in severity and duration of attacks until for the last two months he had rarely been free of pain. He had lost weight, had no inclination for food, and became more and more constipated. He had not had any vomiting nor was the pain related to eating.

On examination, the patient appeared to be suffering intense pain and the abdomen was rigid. He had painful cramps in both legs. Plain enemata gave a copious result of most offensive fæces and all the symptoms subsided. When the enemata were discontinued the symptoms returned. A barium enema showed an acute bend at the splenic flexure.

At operation, a small quantity of free greenish-yellow fluid was found in the splenic region. Running from the free border of the spleen to the splenic flexure was a band, 18 mm. in length and 3 mm. thick. On the anterior surface of the transverse colon, 2.5 cm. from the splenic flexure, was an accessory spleen, 2 cm. in diameter.

The symptoms were attributed to the fact that this body interfered with the normal peristalsis of the bowel, causing at first partial obstruction with localized peritonitis. Following an attack of peritonitis the band between the colon and spleen was formed, which further aggravated the trouble and led up to the condition found when the patient was first seen.

CASES 4 AND 5. (Reported by Morrison, Lederer and Fradkin.¹³)

These were two cases of purpura hæmorrhagica, one of which was followed by apparent cure almost three years after splenectomy, and the other still showing symptoms and signs of the disease almost two years after operation. The latter had an accessory spleen the "size of a walnut" near the hilus of the spleen, which was not removed.

A. W. Spence (quoted by Morrison *et al.*¹³) has recently abstracted 101 cases of purpura hæmorrhagica treated by splenectomy. He reports that 15 per cent of cases returned to pre-splenectomy states. This is suggestive when one considers that accessory spleens are found in about 11 per cent of cases.

CASE 6. (Reported by Sneath.¹⁶)

This is the case I have already mentioned of the accessory spleen in the scrotum. In this instance it was clinically mistaken for a third testicle.

CONCLUSIONS

The importance of accessory spleens has been underestimated. They are common in certain locations in the peritoneal cavity, but rarely found in parenchymatous organs (pancreas,

liver). Accounting for this presence, there are two possible theories, traumatic and developmental. The latter accounts better for the intraparenchymatous enclosures and certain other dislocations. Clinically, accessory spleens explain some obscure pains and masses in the abdomen, and may even be mistaken for a third testicle or hernial sac. They may offer an interesting explanation for the unsatisfactory results obtained in the attempts to cure thrombocytopenic purpura by splenectomy. Thus it appears that, besides being essentially splenic tissue in structure, the accessory spleen is splenic in function, taking part in the diseases of the spleen.

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IDENTICAL DUPUYTREN'S CONTRACTURE IN IDENTICAL TWINS

BY HAROLD COUCH, F.R.C.S.

Department of Surgery, University of Toronto

THE accompanying photographs are not pictures of the same hand. They are reproductions of the left hands of identical twin brothers. Not only do they provide an interesting observation on the cause of Dupuytren's contracture but also an illustration of the strange common destiny in matters of life and growth which surrounds twins. So far as we can determine no case of identical Dupuytren's contracture has been described in medical literature.

While Mr. H.S., whose hand is photographed in Fig. 1, was in the hospital for investigation of duodenal ulcer, routine physical examination revealed Dupuytren's contracture of mild degree involving the ring finger of his left hand. A few days later his twin brother visited him and disclosed to the nurse that he had a similar lesion (see Fig. 2).

In the hands of each of these brothers Dupuytren's contracture has occurred, making its appearance at the same age, reaching about the same degree of severity, involving the same finger in the same hand in the same manner, and progressing at the same rate.

One of the criteria for the diagnosis of monozygotic twins suggested by H. H. Newman¹ is that one hand of one twin must be more like one hand of the other twin than like his own other hand. This is well illustrated here. In each hand the contracture, though slight, has

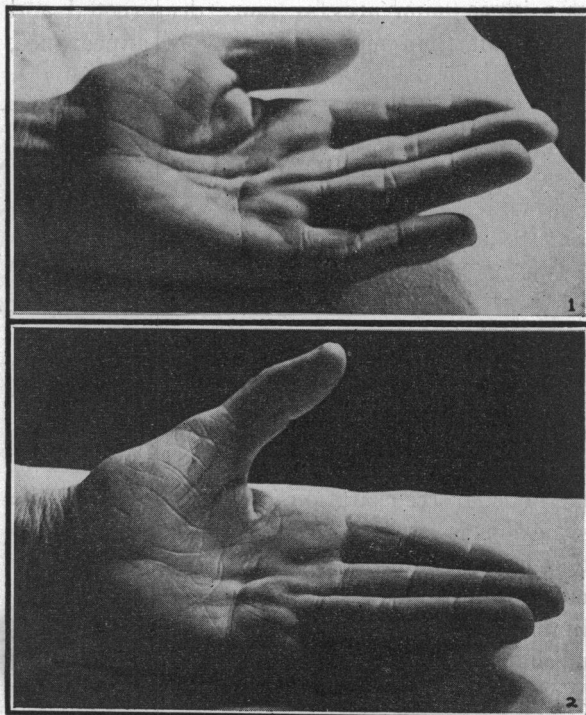


Fig. 1.—Patient's hand showing Dupuytren's contracture and lines.

Fig. 2.—Twin brother's hand showing identical Dupuytren's and similar lines.