MULTIPLE SPLEENS IN A CASE OF ARRESTED ROTATION OF THE MID-GUT LOOP

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A 79-year-old White female cadaver dissected in this Department during 1963 exhibited anomalies of rotation of the fore-, mid- and hindgut, which have been described elsewhere.[29] This individual further displayed a remarkable fragmentation of the splenic tissue into 3 moderate-sized blocks which may be termed anterior-superior, posterior-superior, and inferior spleens and 6 smaller nodules or spleniculi (Figs. 1 and 2). All these were entirely invested by peritoneum, and numerous peritoneal folds linked them to the greater omentum, kidney and posterior body wall.

Accessory spleens may have no Malpighian corpuscles and be intermediate between spleen and lymph nodes—these are termed haemo-lymph nodes.[1] In our specimen even the smallest spleniculus showed lymphoid follicles and so can be classified as a true spleen.

The tail of the pancreas, which was very short,[2] did not nearly approach the hila of any of the spleens.

Anterior-Superior Spleen

By its position and relations this appeared to represent most nearly the normal spleen. It measured 8·75 cm. in long axis (vertically), 5·625 cm. transversely and 5·625 cm. in (antero-posterior) thickness. A deeply-notched anterior border suggested a lien lobatum.

This spleen was related anteriorly to the stomach, posteriorly to the posterior-superior spleen and its accompanying spleniculi, and more medially to the upper lateral aspect of the left kidney.

Posterior-Superior Spleen

This lay immediately posterior to the anterior-superior spleen. It measured 6·25 cm. vertically, 5 cm. transversely, and 3·75 cm. antero-posteriorly, and had a bilobed appearance. It was related anteriorly and above to the fundus of the stomach, and anteriorly and below to the anterior-superior spleen. Medially it was related to the central tendon of the diaphragm at the level of the 10th thoracic vertebra, laterally to the tangle of splenic vessels, and posteriorly to the diaphragm and 11th rib above and to the postero-lateral aspect of the left kidney below.

Related to the lower pole of this spleen was a spleniculus of 3·125 cm. square, and in its hilum posteromedially were 2 additional spleniculi of 2 cm. square (lower) and 2·5 cm. square (upper); these were related anteriorly to the splenic vessels and posteriorly to the diaphragm.

Inferior Spleen

This was the lowest block of splenic tissue, measuring 10 cm. in its longest axis, 7·5 cm. in transverse axis and 5 cm. in thickness, and lying obliquely with its lower medial end in the left iliac fossa and its upper lateral end related to the tip of the 12th rib just posterior to the mid-axillary line. Posterior-inferiorly it was related to fascia iliaca, iliacus, and the iliac crest, and postero-superiorly to quadratus lumborum and the lower lateral surface of the left kidney. Both medially and laterally it was related to vertically placed segments of the colon, which lay entirely in the left lateral region of the abdomen; anteriorly it was separated from the anterior abdominal wall by the greater omentum.

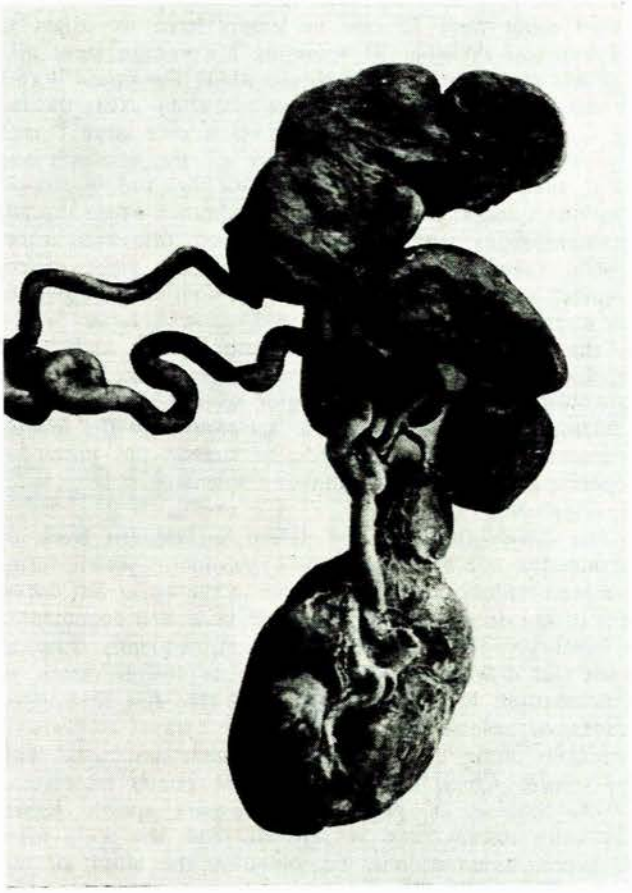

Fig. 1. Mass of splenic tissue. Anterior-superior spleen occupies highest position and inferior spleen the lowest. Other blocks are indicated in the diagram (Fig. 2) and text.

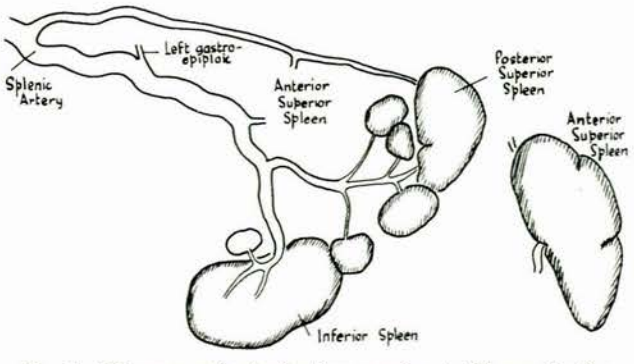

Fig. 2. Diagram of splenic tissue and arterial supply (see text). Anterior-superior spleen has been detached from its vascular connections.

A spleniculus 1·25 cm. square was found in the hilum of this spleen and a spleniculus of 2·5 cm. square was related to its upper pole.

A 6th spleniculus of 0·125 cm. square lay in the greater omentum 2·5 cm. from the greater curvature of the stomach near its point of maximum convexity.

Blood Supply

The exceptionally tortuous splenic artery (Fig. 2) measured more than 25 cm. in length from its origin to its terminal division. It arose by a common stem with the left gastric artery and passed along the upper border of the pancreas to which it gave branches. Near the tail of the pancreas this artery gave off a very large branch which followed the greater curve of the stomach and gave branches to the anterior-superior and posterior-superior spleens; 3 cm. beyond this branch arose the left gastro-epiploic artery, which gave off numerous short gastric vessels and then linked with the right gastro-epiploic artery. The splenic artery continued leftwards, accompanied by the splenic vein, gave a large branch to the hilum of the anterior-superior spleen, and 1 cm. beyond this divided into 2 terminal branches, 1 coursing downwards to supply the inferior spleen, the spleniculus enclosed in its hilum, and the spleniculus in the greater omentum, the other upwards to supply the posterior-superior spleen and the remaining spleniculi.

Development

The development of the spleen in the 5th week of intrauterine life from splanchnic mesoderm which forms localized 'hillocks' of splenic tissue in the upper left dorsal part of the dorsal mesogastrium has been well documented by embryologists. These hillocks subsequently fuse to form the definitive spleen; incomplete fusion results in lien lobatum, while unfused anlagen give rise to isolated blocks of splenic tissue, which may travel in various directions along the peritoneal ligaments associated with the spleen. Curtis and Movitz[3] most clearly interpreted the mechanism of production of extra splenic tissue. Accessory spleens may be separate from the main mass of splenic tissue or may be joined to the hilum of the true spleen by cords of splenic tissue.

Incidence

The time-honoured figure of a 10% incidence of accessory spleens would appear fairly accurate; in a recent study Halpert and Eaton[4, 5] found 94 in a series of 1,000 cadavers. There is a gradual percentage decrease in splenic weight from birth onwards and this applies to additional splenic tissue which is more commonly found in younger age groups and is distinctly uncommon over the age of 75 years.[6]

The incidence of supernumerary spleens rises sharply in certain blood disorders; it is at least doubled in cases with acholuric familial jaundice[7] and is estimated at 53% in cases of idiopathic thrombocytopenic purpura.[8, 10] Hypertrophy of splenic tissue commonly ensues when the bone marrow is depressed. Curtis and Movitz[3] go so far as to say that accessory splenic tissue tends to involute unless influenced to remain by some pathologic process.

Number

This varies from 1, by far the commonest, to about 23. Halpert and Eaton[4, 5] in 600 cases found 52 instances of 1 extra spleen, 6 instances of 2 extra spleens, 3 instances of 3 extra spleens and 1 instance only of several spleens associated with absence of a spleen proper. Brash and Stewart[11] found 13 masses of splenic tissue and mention that Testut quotes Otto as having found 23 and that Baillie and Cruvethier note 2 cases with 7 each. Helwig[12] describes 2 cases in infants in which he found 14 and 10 separate nodules of splenic tissue, with a bilobar spleen proper in the latter instance. He quotes Pool, citing 4 cases of Hyrtl's, in which the spleen was broken up into 5 - 11 smaller nodules and 2 cases of Garrod's in which 4 and 9 nodules were noted respectively and also refers to Helly's specimen of a bilobular spleen with 11 accessory spleens preserved in the Anatomical Museum in Vienna. Rhodes and Grunberg[13] describe 9 accessory spleens. Vast numbers, the highest of which is 1,000, referred to by various authors can be traced to Kaufmann's[14] description of Albrecht's and Faltin and Kuttner's cases, both of which are undoubtedly instances of splenosis owing to regeneration of fragments of spleen implanted in the peritoneum following trauma — not true accessory spleens resulting from congenital failure of anlagen to fuse.

Size

Large nodules of accessory splenic tissue are unusual; the range seems to be between 0·2 cm. - 4 cm. with most between 0·5 cm. and 1·5 cm. Prof. J. H. Louw, Head of the Department of Surgery in this Medical School, has told me that he has never seen accessory spleens approaching the size of a normal spleen. Settle[15] describes engorged infarcted accessory spleens removed for torsion of the pedicle with the following measurements:

From a 4-year-old: 6 x 6 x 7 cm.
From a 6-year-old: 2 x 3 x 3 cm.

Brash and Stewart's[11] largest mass of accessory splenic tissue was 6·1 cm. x 4·1 cm.

A common descriptive term employed is 'of reasonable size'. It will be remembered that our largest nodules were: 10 x 7·5 x 5 cm. and 6·25 x 5 x 3·75 cm.

Sites

The sites in which extra splenic tissue have been found in descending order of frequency are:

1. Hilum of spleen proper.
2. Pedicle of spleen.
3. In peritoneal ligaments, gastro-splenic, spleno-renal, greater omentum, pancreatico-splenic, transverse mesocolon, phrenico-colic.
4. Related to gonads on left side. Owing to embryonic contiguity of the splenic and gonadal anlagen, extra splenic tissue may become attached to the left gonad and travel with it: (*a*) to broad ligament where it usually is found in the mesovarium or mesosalpinx, and (*b*) to the scrotum where it may stimulate a tumour of the testis or epididymis. Interesting reviews of splenico-gonadal fusion with case reports are given by Putscher and Mannion,[16] Jayne and Jessiman,[17] Emmett and Dreyfuss,[18] Daniels,[19] and Grossman *et al.*[6]

Not uncommonly such spleens are associated with inguinal hernia and imperfectly descended testis.

Scrotal accessory spleens may be linked by a cord of fibrous and/or splenic tissue to the hilum of the spleen—this cord running anterior to the intestines—but such a connection may be absent.

5. Fused with the liver.[20]
6. In tail or head of pancreas.[21]
7. On right side of posterior abdominal wall.

A case described by Milroy Paul[22] is of great interest as is his contention that all gnathostome vertebrates have the spleen on the left side and his claim that his case may represent a state homologous to that obtaining in some amphibians and reptiles. Since the entire dorsal mesogastrium has the phylogenetic capacity to form splenic tissue this would seem a more likely explanation of the phenomenon and an additional rotation error as seen in the cases of Brash and Stewart,[11] Rhodes and Grunberg[13] and Hyrtl,[12] might also have been operative here. Alternatively splenosis occurred in his case.

8. In cases of congenital diaphragmatic hernia, splenic tissue has been found in the pleural cavity.

ASSOCIATION WITH OTHER CONGENITAL ANOMALIES

In the present case multiple spleens were associated both with a patent foramen ovale and with 2nd-stage arrested rotation of the mid-gut loop.

Ivemark,[23] while engaged in finding the relation of splenic primordia to Streeter's embryological horizons, found that in 69 cases of splenic agenesis, as well as in some cases of multilobate spleen, there were associated congenital malformations of the heart and great vessels. The development of the spleen takes place concurrently with the partitioning of the truncus arteriosus and the growth of the endocardial cushions, and defects of these structures were the commonest type of anomaly associated with abnormal splenic development. He also made the observation that congenital heart disease associated with absence of the spleen is usually combined with varying degrees of situs inversus. Aguilar and his co-workers[24] expanded this syndrome in reviewing 27 cases of splenic agenesis, and found in addition to defects of partitioning of truncus, endocardial cushion defects and partial situs inversus, septal defects, accessory lobes to the lungs and abnormal mesenteric attachments.

Hyrtl[12] had 4 cases with transposition of the viscera in which the spleens were broken up into 5 - 11 smaller nodules; Brash and Stewart[11] describe a case of transposition of the mesogastric viscera in which they found 13 blocks of splenic tissue, 2 large and the rest smaller. Rhodes and Grunberg[13] describe a case of Hodgkin's disease in which there was transposition of the viscera and 9 masses of splenic tissue were found. Greenberg[25] describes 16 lobules of splenic tissue weighing 6 G associated with transposed stomach, pancreas, spleen and intestinal tract.

CLINICAL SIGNIFICANCE

Although the majority of accessory spleens remain asymptomatic, they participate in all pathologic processes involving the spleen and *de novo* may be subject to torsion[15,26] or rupture.[27] Thus clear-cut indications for removal may develop.

King and Schumacker[28] indicate a greatly increased susceptibility to infection in general, and meningitis in particular, in cases splenectomized in early infancy, and surmised that since the spleen is the largest reservoir of antibody-producing lymphocytes it may act as an important defence mechanism in early life.

Splenectomy for congenital spherocytosis with the removal of accessory spleens would appear to be contra-indicated in young infants.

SUMMARY

A 79-year-old White female cadaver showed a remarkable fragmentation of splenic tissue associated with arrest of 2nd-stage rotation of the mid-gut loop and a patent foramen ovale. Nine separate blocks of splenic tissue were found, 3 of which attained a size comparable with that of a normal adult spleen, the remaining 6 being small nodules or spleniculi.

The large spleens lay in the left half of the abdominal and pelvic cavities; 5 of the spleniculi lay in the hila of these spleens, the 6th in the greater omentum. All were supplied by terminal branches of the splenic artery. The development, incidence, number, size and sites of multiple spleens are reviewed. There is evidence that supernumary spleens may retrogress in later life, which makes the number of bodies of splenic tissue in this individual the more remarkable.

The formation of the splenic primordia coincides with truncus partitioning, development of endocardial cushions and the onset of definitive rotation and attachment of the mid-gut loop.

Splenic anomalies, including multiple spleens, have been found, in a considerable number of cases, to be associated with congenital cardiac defects and with varying degrees of situs inversus. The present case provides a further instance of this association. Clinical implications of multiple spleens are briefly indicated.

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