Primary tumors of the spleen are rare, only about 120 well authenticated examples having been published.

CASE REPORT

A nineteen-year-old boy was admitted to Christ's Hospital (Topeka, Kansas) in 1929, and operated upon for appendicitis. He made a good recovery and appears to have been well until April 1936, when he was readmitted. The family history showed some cancer but no tuberculosis.

On the patient's second admission he complained of abdominal disturbances which had at first been attributed to a gastro-intestinal lesion. Medication had failed to give relief and meantime the abdomen had become distended and respiratory difficulty had developed. On examination the only finding of significance was extreme distention of the abdomen, through which the percussion wave was easily transmitted. The patient was well nourished and there was no chest lesion. The white cell count was 11,000 with 81 per cent polymorphonuclears.

Paracentesis was performed and about 4300 c.c. of serous fluid removed. A mass was then palpable in the upper abdomen and an exploratory laparotomy was undertaken. The left lobe of the liver was found to be adherent to the diaphragm and a tumor was discovered in the left epigastric region. Metastatic carcinoma of the liver was suspected and the abdomen was closed without further surgical procedure.

The fluid re-formed promptly, and repeated paracenteses were necessary until the patient's death on Aug. 6, 1936, between 3000 and 4000 c.c. being removed at approximately weekly intervals. The fluid was serous and only rarely blood-tinged. After his discharge from the hospital, in May, the patient lost weight and during the later course of his illness was confined to bed.

At autopsy the body was extremely emaciated. The abdomen was distended and contained about 4 liters of serous fluid. The intestinal loops were free and moderately distended but showed no lesions. The duodenum and the stomach were normal. The descending colon and the left lobe of the liver were adherent to a mass in the left upper abdominal quadrant. The mesentery contained several enlarged nodes which on section were of a whitish-gray color, some showing calcification. The lumbo-aortic nodes were also enlarged. The left iliac group and those at the hilus of the liver were enlarged and discrete. A mass, some 10 cm. in diameter, was present in the splenic region, globular in shape and adherent to the diaphragm, from which it could not be freed. That the tumor had its origin in the spleen was evident from the occurrence of splenic tissue on the surface and at the lower pole. A splenic notch was also observable (Fig. 1). Toward the hilus the tumor was diffuse; toward the periphery the appearance was of confluent nodules interspersed with splenic parenchyma. The latter formed a shell some 1 cm. in thickness about the tumor.

The tumor tissue was of a grayish color, rather firm, with areas of softening. The splenic vessels were enclosed in it and compressed. The diaphragm was invaded on the left; the liver was adherent to the tumor but not invaded by it. The pancreas and adrenals were of normal appearance. The kidneys were pale, small, and somewhat granular on the surface, but contained no growth. The prostate and the bladder showed no lesions and the vertebrae were not invaded.

About two liters of serous fluid resembling that in the abdomen filled the left pleural cavity, pushing the collapsed lung to the hilus. The right lung showed edema of all three lobes, but no area of consolidation. A group of hyperplastic lymph nodes matted together
was present at the bifurcation of the trachea, but there were no other nodes in the mediastinum. The heart was small (112 gm.), resembling that of a young child. The great vessels at the base were rather hypoplastic, but nearly of adult size, offering a striking contrast to the small organ from which they sprang. The coronary vessels formed tortuous lines on the heart surface, as they had not undergone the same atrophic process as the heart muscle.

The tumor, weighing 850 gm., was elliptical in shape, measuring \(12 \times 9 \times 10\) cm. Nodules were present on the surface and invaded the capsule, being in marked contrast to the dark splenic parenchyma. The tumor appeared to have arisen from the region of the hilus.

Section showed a diffuse tumor of a grayish color, with a rim of splenic parenchyma (Fig. 2). Small islands of dark tissue with the appearance of splenic tissue were found in the mass, particularly in its distal portion. When the edge of the knife was passed over the cut surface, cellular elements were not easily removed and pressure was necessary to

![Figs. 1 and 2. Reticulum-Cell Sarcoma of the Spleen: Gross Specimen](image)

In the uncut tumor the splenic notches are plainly visible. On the cut surface note the narrow band of splenic tissue in the upper part at the right.

scrape out some of the tumor tissue. Around the hilus were three small nodes, which had on section the appearance and consistence of the original tumor.

**Microscopic Examination:** Sections were taken from the diffuse mass, from the small nodules in the splenic parenchyma, and from the marginal zone of splenic parenchyma.

In sections from the diffuse zone very fine strands of collagen tissue are seen, formed by young fibroblasts, and thin capillaries subdivide the tumor, giving a pseudo-alveolar aspect or a coarsely lobular appearance. From the perilobular tissue originate other fine fibers, which penetrate into the alveolar spaces between the tumor cells. The cells are separated from one another and each is nearly filled by a dark-staining nucleus, which leaves only a narrow rim of cytoplasm. The nucleus shows two or three masses of heavy chromatin disposed irregularly and united by fine threads; often, also, a dark staining nucleolus. The shape of the cells is globular, like that of the lymphocyte; the size, although varied, is within definite limits, from that of a prolymphocyte to that of a large lymphocyte (Fig. 3). No giant cells are seen, but some binucleated cells are present. Occasionally between the tumor elements there are seen spindle cells with long wavy cytoplasmic fibrils and dark nuclei, or ellipsoidal pale-staining cells, apparently reticular cells. With Mallory's phosphotungstic acid stain, a thick network of fibers is demonstrable in the perilobular spaces, branching
between the cells and breaking up into fine fibrils, which finally encircle the peripheral cells. The fibrils do not penetrate into the inner layer of the alveolar spaces, where the cells appear undifferentiated, nor does their cytoplasm show any tendency to fibril formation.

With the method of Urechia for reticulin fibers a fine network of argyrophile fibers is demonstrable, branching in all the lobules and assuming, particularly at the periphery, a honeycomb appearance. In the central portions of the large lobules or alveoli only short fibrils are found, but in the small alveoli the honeycomb appearance is present throughout.

In the areas of nodular appearance in the splenic parenchyma the tumor elements seem to have no relation whatever to the follicles, which would indicate almost certainly that they originate outside of these, in the splenic reticulum. They exhibit a tendency to form new capillaries, and some endothelial-like cells are seen scattered between the tumor cells in their progressive infiltration of the splenic tissue. Some transitional forms are seen, which suggest a possible capacity of the cell to evolve into another type, particularly the endothelial type. Here and there are small syncytial figures formed by three or four cells, giving the aspect of a multinucleated cell. In the stroma is a small amount of ferruginous pigment, mostly free. There are no inclusions of pigment in the cytoplasm of the tumor cells.

The metastases in the lymph nodes are identical with the original growth. The cells have retained their original capacity to produce argyrophile fibrils.

Microscopic study of the liver, lungs, kidneys and adrenals confirmed the gross impression of absence of metastases.

In determining the exact origin of the tumor three elements are to be taken into consideration: the small lymphocyte of the malpighian corpuscles, the reticular cell or cell of the splenic cords, and the endothelial elements of the sinuses. These three elements, according to the majority of authors, give origin to the three forms of splenic tumors: the small-cell lymphosarcoma, the large-reticulum-cell lymphosarcoma (or better retothelial sarcoma), and the endothelial tumor. The fibrosarcoma is not properly a splenic tumor as it originates from the connective tissue of the capsule or from the trabecular framework supporting the organ, and not from the specific elements of the splenic parenchyma.
In the present case, taking into consideration the areas in which normal splenic elements are still present, the surviving malpighian follicles appear altered, undergoing sclerotic change but never showing evidence of proliferation. They are compressed by the tumor cells, which replace the lymphocytes or push them out, but are always distinct from them. In many areas tumor elements and scattered lymphocytes are seen together, intermingled in the vicinity of the malpighian follicles, but no transitional forms between the lymphocyte and the large tumor cell have been observed. The central artery of the malpighian follicle has usually undergone a hyaline change; the bodies themselves are more homogeneous and are disappearing, while the pulp elements are hyperplastic and appear more active.

The cells are still chromatic, without nucleoli, of uniform size, forming alveoli, separated only by thin capillaries. A fine network of argyrophile fibers encircles the single elements. The reticulum, however, is rather irregular, sometimes fine, at other times absent or coarser, particularly in the vicinity of the trabeculae. No giant cells have been found, although occasionally multinucleated cells of medium size have been observed.

The absence of plasma cells and eosinophils definitely excludes a granulomatous process. Forms representing a transition from the pale reticular cell to the tumor cell are seen, and the presence of numerous thin capillaries indicates an activity which may be attributed only to the reticular cell.

There is no actual new blood vessel formation such as is found in endothelial tumors arising from the endothelial elements of the sinuses. The formation of vessels never goes beyond the stage of fine capillaries.

**DISCUSSION**

Splenic tumors have generally been classified, at least in the American literature, as (1) fibrosarcoma, derived from the capsular or trabecular elements; (2) lymphosarcoma arising from the reticular cells; (3) endothelioma having its origin in the endothelial elements lining the sinuses.

Ménétrier, in France, tried to separate a special type of splenic tumor derived from the large mononuclear cells of the cords. To this he gave the name splenoma. A prominent feature of this tumor is the presence of numerous giant cells containing inclusions or vacuoles. In the portions of the spleen not yet invaded by the neoplastic process are observed an intense sclerosis, an endarteritis obliterans of the medium-sized and smaller arteries, periarteritis, and disappearance or atrophy of the follicular elements. Ménétrier concludes that such a picture may represent the soil on which the neoplasm has developed.

Foix and Roemmele described a tumor corresponding to Ménétrier's splenoma and called it a nodular reticulo-splenoma. They believed that it was derived from the reticular elements of the malpighian follicles. Another case, described by Duchemin as a primary sarcoma of the spleen, should, according to Ménétrier, be included in his group. Although he regards these tumors as derived from the mononuclear elements of the splenic cords, he agrees that their origin is sometimes difficult to trace and finds no objection to the conception of Foix and Roemmele of a possible follicular origin.

The term splenoma was also used by Cesarsis Demel, but the lesion which he describes and illustrates was rather a hyperplasia of the splenic parenchyma than a true neoplasm.

Langenstrasse and Neumann have recently described a case of primary reticulo-endothelial sarcoma of the spleen, which fits into the group of tumors described by Ménétrier as splenoma and by others as endothelial sarcoma. The tumor evidently originated from the reticulum of the organ, but from the
description no conclusion can be reached as to its endothelial nature, as the cells had no angioplastic activities. The only difference between this tumor and the one here presented is the presence in the former of numerous giant cells and rather pale nuclei. The argyrophile fibers formed a complete network and the authors appear to conclude that the presence of such fibers is indispensable for a diagnosis of endothelial sarcoma.

Such is not our opinion, as the presence of fibers, the so-called Gitterfasern, is not a specific feature of reticular cells, but is found almost constantly in connective-tissue tumors, even the benign fibroma. The property of fiber formation belongs to the mesenchymal cells, and in their evolution they preserve this exclusive characteristic. The more primitive the cell, the less the capacity to produce a complete network. In the more undifferentiated tumors no real network is seen; only fine fibrils, mostly intracytoplasmic. To rely on the presence of a complete argyrophile network for a diagnosis of reticulum-cell sarcoma would entail the risk of overlooking the more malignant forms.

Oberling classifies the reticulosarcomas as: (1) undifferentiated reticulum-cell sarcoma; (2) differentiated reticulum-cell sarcoma, and (3) evolitional forms, as the reticulo-endothelial sarcoma, reticulomyelosarcoma, reticulolymphosarcoma. In the description of their case, Langenstrasse and Neumann stress the epithelioid aspect of the cells, the almost complete absence of vascular activity, the presence of fibrocytes and lymphocytes, and the fine argyrophile network. The tumor would thus belong to Oberling's third group, in which the reticular elements are already differentiating toward those of fibro- or polymorphous connective-tissue type. There is no ground for considering it as endothelial and the terminology which the authors propose as the most suitable for such a tumor is in our opinion quite unsuitable, since it fails to take into account the potentiality of the tumor cells. In regard to the origin, there is no proof of a derivation from endothelial elements, as they cannot be differentiated in their more immature aspect from the reticulum cells, and, on the other hand, they are derived from these cells.

In our case the formation of reticulin is not a constant feature, for some cells show only intracytoplasmic fibrils, but these, too, are reticular elements which possess evolutionary properties. In some areas the cells have assumed a lymphoblastic character, in others they tend to differentiate in other directions. The actual position of this tumor remains uncertain, for in some respects it would appear to belong in Oberling's second group of differentiated reticulosarcoma, while in others it could be grouped with the forms showing a lymphoblastic evolution. We prefer this latter classification, as it reflects the evolutionary potentiality of the tumor. In many areas the cells preserve a definite reticular aspect but do not show any definite orientation; in other areas the orientation is apparently lymphoblastic, and this must be taken into consideration. The lack of reticulum in the more immature areas does not exclude the fact that we are dealing with a tumor derived from the reticular cells, but only implies the immaturity of the cells.

In conformity with the conceptions and classification of Oberling it seems very probable that the tumor originated from the reticulum cells of the splenic cords and should therefore be classed as reticulum-cell sarcoma. In view of
the probable orientation of the elements, it might be called a reticulolymphosarcoma or lymphoblastic reticulosarcoma.

**Summary**

A case of primary splenic tumor has been reported with metastases confined to the lymph nodes. It appeared to be derived from the reticulum cell, as the malpighian follicles showed degenerative lesions. The tumor was not uniform in differentiation, as evidenced by reticulin impregnation. Besides areas in which the cells were undifferentiated there were others in which they showed a differentiation to lymphoblasts.

In classifying the tumor, we adhere to the conception of Oberling and adopt his classification, which is the only one taking into account the nature and the future evolution and possible differentiation of the growth. The diagnosis, therefore, is reticulolymphosarcoma or lymphoblastic reticulosarcoma of the spleen.

**Note:** The author is indebted to Dr. A. J. Briar for the photographs of the tumor.

**References**