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MALIGNANT TERATOMA OF THE SPLEEN

The following presentation represents the first case report of a malignant splenic teratoma.

CASE REPORT

First admission

A 69 year old white woman was admitted to the Norwalk Hospital April 15, 1968 complaining of left upper quadrant pain and vomiting of two days' duration. She was acutely ill, temperature 101°F., with a somewhat distended abdomen. There was generalized abdominal tenderness particularly in the left upper and right lower quadrants. A large firm, tender mass was felt in the left upper abdomen. Laboratory data were: hemoglobin 14.4 gm/100 ml.; wbc 7,400/cu mm; polymorphonuclears 83%, stabs 1%, lymphocytes 9%, eosinophils 3%, monocytes 3%, basophils 1%. Urinalysis, blood urea nitrogen and serum electrolytes were within normal limits. X-ray film of the abdomen showed a soft tissue mass in the left upper abdomen. On intravenous pyelogram, this mass appeared to displace the left kidney inferiorly and medially.

She underwent surgical exploration that night through a midline incision. Findings included an acutely inflammed appendix and a huge freely moveable spleen but no ascites. The ovaries were atrophic and the pancreas unremarkable. No disease of other organs was found. The spleen and appendix were removed. The patient's postoperative course was uneventful and she went home on the 17th day.

Findings in the appendix grossly and microscopically were typical of acute appendicitis.

The spleen measured 18 × 15 × 15 cm. and weighed 2,500 grams. Most of it was occupied by a large monolocular cyst (Fig. 1) the wall of which averaged 2 mm. in thickness. There were saccular outpouchings and papillary projections of the lining (Fig. 2). There were large areas of calcification in the wall, appearing as thin, large, flat plaques. The cyst contained about 1,500 ml. of a thin yellow-brown turbid liquid. Cytologic examination of the fluid did not yield atypical cells. This cyst was intimately attached to

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the remaining 150 grams of red-pink fleshy splenic tissue covered by a thin wrinkled capsule. Although most of the cyst wall was thin, there were some thicker gray-red areas in which were foci of yellow opaque discoloration. In other regions there was gray rubbery tissue resembling cartilage. Microscopically there was a very mixed appearance. The thinner portions of cyst wall were made up of relatively acellular collagenous tissue. The inner lining in these areas was eosinophilic material identified as fibrin rather than epithelium and in the wall were extensive areas of old hemorrhage containing striking cholesterol clefts and amorphous calcium deposits. The more cellular areas of viable tumor demonstrated three main elements.

First, there were papillary carcinomatous structures (Fig. 3) with a loose connective tissue core covered by one or two layers of high columnar epithelium. The epithelial cells varied moderately in nuclear size and chromatin content and mitotic figures were rare. These cells contained much mucus demonstrated by Alcian Green stain, and there was also much mucus in the spaces between the papillae. In some foci there was considerable vascular stroma moderately infiltrated with plasma cells and lymphocytes and frequently the core of the papilla contained a psammoma body (Fig. 4).

A second type of growth was in the form of spindle cells with a whorled pattern (Fig. 5). The cells were irregular and mitotic activity was moderate. The stroma stained brown in a Masson's stain, suggesting origin from muscle cells rather than fibroblasts.

Finally, in the areas which grossly resembled cartilage, the cells were ovoid, clumped in small groups, and surrounded by a small clear halo separating them from abundant intercellular material which stained pale blue in the H & E stain (Fig. 6). The pathologic diagnosis was malignant teratoma. The slides were reviewed by Dr. Robert V. Hutter, Professor of Pathology, Yale University School of Medicine, and he concurred in our diagnosis.

Last admission

Four months after discharge the patient was brought to the Emergency Room because of difficulty in breathing. She was in acute distress, dyspneic and jaundiced. Blood pressure was 90/70, respirations 28 and pulse 120 and irregular. There was dullness on both sides of the chest particularly on the left. The heart sounds were decreased. No murmurs or thrills were present. There was moderate ascites and marked pitting edema of the legs. Laboratory data were: hemoglobin 13.5 gm/100 ml.; wbc 14,200/cu mm; polymorphonuclears 86%, stabs 6%, lymphocytes 8%; platelets 93,000/cu mm; BUN 76 mg/100 ml., creatinine 1.6 mg/100 ml.; HCO_3 28 mEq/L, Cl 78 mEq/L, Na 120 mEq/L, K 5.9 mEq/L; total bilirubin 18.6 mg/100

FIG. 1. Deflated cyst of spleen. Normal spleen in left lower section of photography.

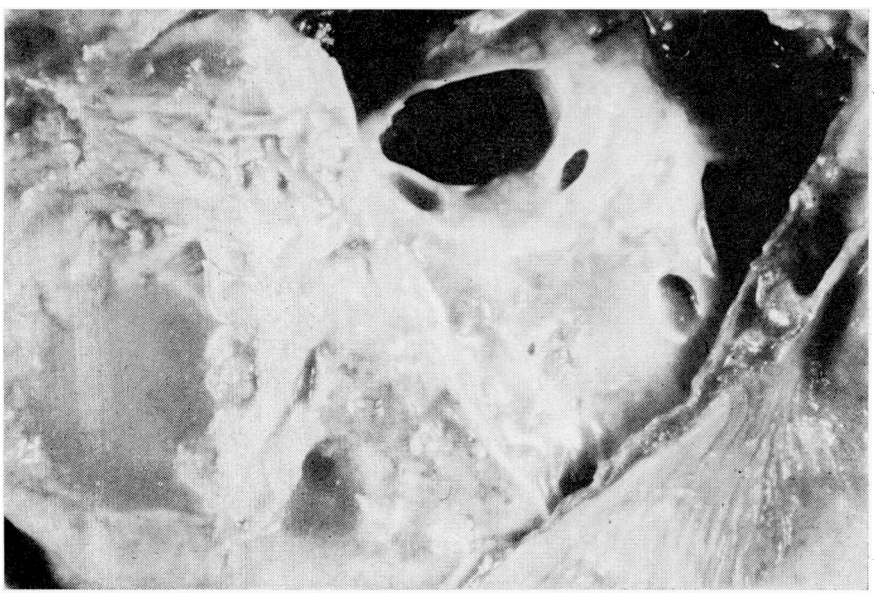

FIG. 2. Lining of splenic cyst. Note thin wall in right lower region, papillary projections into lumen and outpouchings of the cyst.

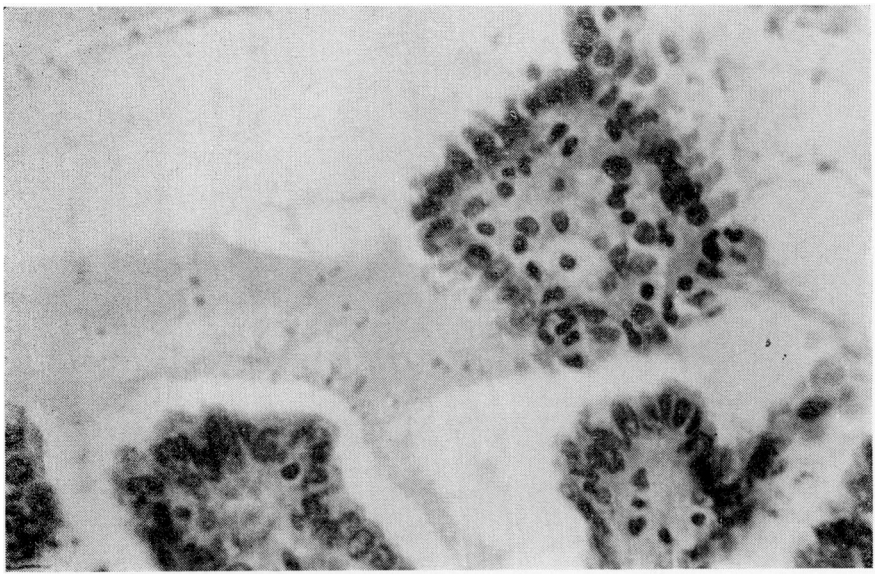

FIG. 3. Papillary adenocarcinoma. Material in lumen and in apices of epithelial cells stains strongly positive for mucus. Original magnification *500* ×.

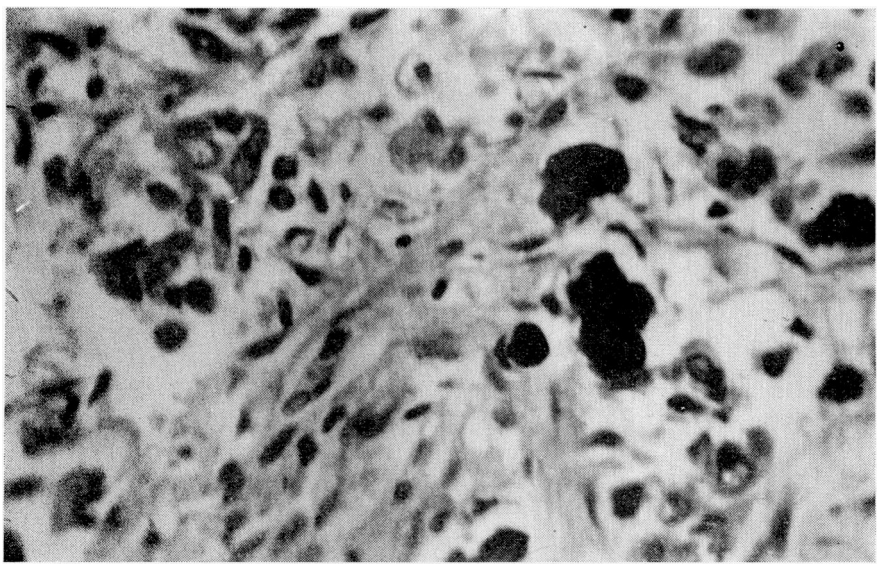

FIG. 4. Psammoma bodies, dark irregular calcifications in stroma of papillary adenocarcinoma. Original magnification *500* ×.

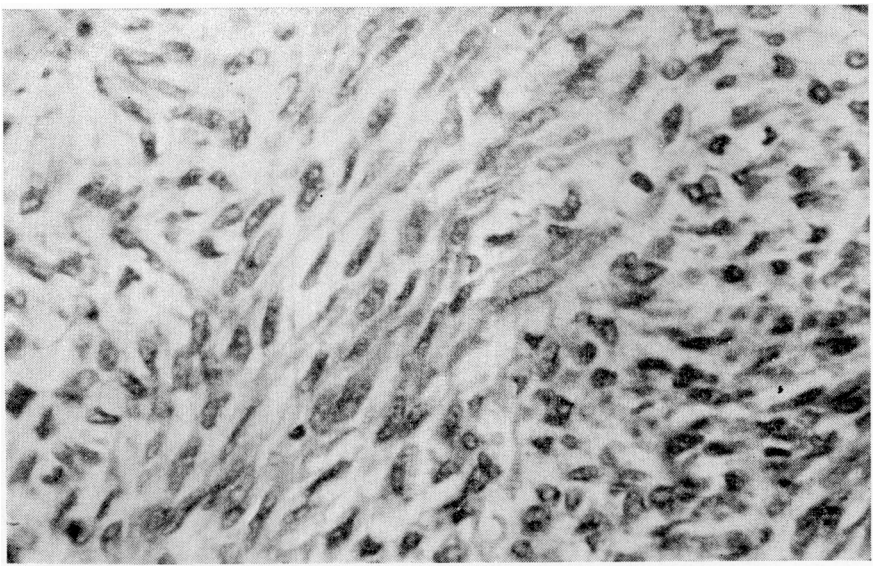

FIG. 5. Cellular spindle cell sarcoma with whorled pattern. Cells variable. Stroma stains brown in Masson stain indicating probable muscle origin. Original magnification *500* ×.

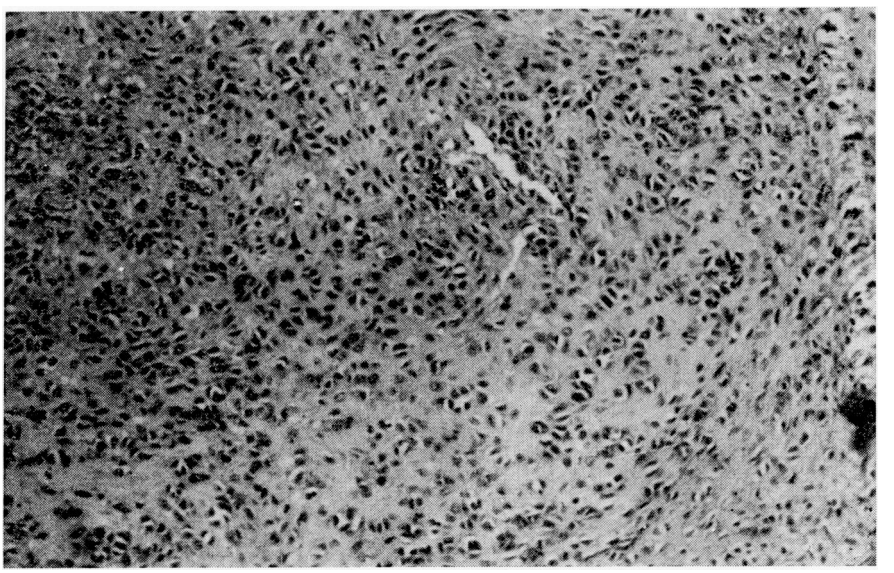

FIG. 6. Area resembling cartilage. Note spaces about many nuclei. The abundant interstitial material is somewhat basophilic in H & E. Original magnification *156* ×.

ml., direct bilirubin 14.4 mg/100 ml. Urinalysis was within normal limits. Chest film was interpreted as showing cardiac enlargement and bilateral pleural effusion. ECG was read as flattened T-waves and sinus tachycardia with second degree AV block. After treatment for congestive heart failure with apparent improvement she was found dead in bed the next morning.

Gross autopsy

There were 500 ml. of ascitic fluid, 100 ml. of right pleural fluid and 2,000 ml. of left pleural fluid. There was extensive tumor in the abdomen with multiple gray-white 3 to 15 mm. nodules scattered over the peritoneum, omentum and surface of the intestine. The liver weighed 1,540 grams and was about 70% replaced by umbilicated malignant gray-white nodules up to 4 cm. in diameter, typical of metastatic disease. A nodule about 4 cm. in size involved the tail of the pancreas. The ovaries were atrophic and free of tumor. The heart was large (500 grams), dilated in all chambers and contained organizing mural thrombi in the left ventricle. There was gross scarring of the ventricular muscle. The coronary arteries were markedly sclerotic and narrowed, and the aorta was also severely sclerotic. In the left lower lobe of the lung was an arterial embolus and an area of hemorrhagic pulmonary infarction distal to the embolus.

Microscopically the liver metastases were predominantly papillary carcinoma with smaller areas of spindle cell sarcoma, both entirely similar to what was present in the original splenic tumor. The pancreatic nodule contained both elements, with the sarcoma predominating. There were microscopic foci in the lung entirely of adenocarcinomatous configuration. The ovaries were not involved.

Death was believed to have resulted primarily from arteriosclerotic heart disease with the pulmonary embolus playing a large role. The metastatic tumor was considered to be a major contributing factor. The jaundice was attributed to the liver metastases.

DISCUSSION

In 1923 Smith and Rusk[1] classified malignant splenic tumors according to the three types of tissue that give rise to tumor. These were:

1. The capsular trabecular framework yielding spindle cell sarcoma.
2. The lymphoid components from which lymphosarcoma arises and
3. The vascular structures producing angiosarcoma.

They found 102 cases of all types of primary malignant neoplasms and added two of their own. Gordon and Paley[2] in 1951 accepted 189 cases of which 42 were lymphosarcoma. By 1965[3] 198 cases had been described.

The spleen is obviously not a common source of primary malignancy nor

is it a fertile soil for metastases although these do occur as frequently as in the heart or kidney.[4] It certainly would seem an unlikely site for origin of teratoma. Willis[5] defines a teratoma as "a true tumor or neoplasm composed of multiple tissues of kinds foreign to the part in which it arises. In this they differ from mixed tumor peculiar to particular regions." Willis is not generally enthusiastic about the concept of carcinosarcoma, which he considers a malignant tumor consisting of two independent components such as spindle cell sarcoma combined with either adenocarcinoma or squamous cell carcinoma. He believes most so-called carcinosarcomas represent pleomorphic carcinoma.

In the present case, one of the elements, the mucous-secreting papillary carcinoma, is entirely exotic to the spleen. The other two, the spindle cell and cartilaginous cell types, could be derived from splenic stroma by metaplasia. However, we prefer to consider this a true teratoma with multiple cell elements.

Benign nonparasitic cysts of the spleen are well known.[6] Pertinent to the present case are those of epidermoid, endothelial or mesothelial types. The epidermoid cysts have a wall of dense collagenous tissue and are lined by stratified squamous epithelium. Only two of these have allegedly contained enough other elements to be called dermoids but Rappaport[6] does not accept these as definite dermoids. The origin of these epidermoid cysts is obscure. Among the possible modes of origin proposed are trauma, pregnancy, infection, herniation of splenic tissue, preexisting lymphangiomata, misplaced Wolffian body remnants and autochthonous formation.[7] The simple endothelial and mesothelial lined cysts often contain cholesterol clefts and calcific deposits in hyalinized fibrous tissue walls. Although their origin is uncertain it is not nearly as puzzling as that of epidermoid cysts, because similar cysts lined by endothelium or mesothelium are not uncommon in other abdominal viscera or in the peritoneal cavity generally.

None of these benign cysts seem to provide a ready solution to the problem of origin of the tumor in our case. They have not been noted to give rise to malignancy in other cases, nor have any been described as having mucous-secreting lining cells, nor a papillary architecture nor tissue resembling primitive cartilage. Furthermore our case did not contain squamous epithelium in any of the 36 sections studied.

We considered one other possible source of origin to be the gonadal element of a splenic-gonadal fusion,[8] because the ovary is a favorite site of origin of teratoma. However, splenic gonadal fusion is basically a disease of men (27 of the 30 reported cases). In addition, in the three female cases there was a direct connection between splenic and ovarian parenchyma. In our case there is no such connection and the ovaries appeared normal.

SUMMARY

A cystic malignant teratoma of the spleen containing adenocarcinoma, spindle cell sarcoma and primitive cartilage was removed from a 69 year old woman. She died 4 months later chiefly of arteriosclerotic heart disease. Metastatic lesions from the splenic tumor were found in the abdominal cavity, liver, pancreas and lungs, and were composed of adenocarcinomatous and spindle cell sarcomatous elements. This is believed to be a unique case. The origin of a teratoma within the spleen is puzzling. We suggest that it arose from an embryonal rest.

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