

SOLITARY PLASMOCYTOMA OF THE SPLEEN
WITH MARKED POLYCLONAL INCREASE OF GAMMA G,
NORMALIZED AFTER SPLENECTOMY

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Abstract. A routine medical examination revealed a markedly increased ESR in a 40-year-old man. Further studies showed an enlarged spleen and a marked polyclonal increase of serum globulins. Splenectomy was performed, revealing a solitary plasmocytoma which weighed 1 kg. In the course of the first six weeks following splenectomy the serum proteins became practically normalized.

Primary extramedullary plasmocytoma is relatively rare. In Innes and Newall's (2) material of 188 cases of myelomatosis only four had primary extramedullary plasmocytoma, in no case located to the spleen. At autopsy, however, more than 20% of their patients with myelomatosis had gross extramedullary tumor masses within the abdomen or thoracic cavity, in more than 10% of the cases also involving the spleen.

Dolin and Dewar (1) in 1956 made an extensive review of the literature on primary extramedullary plasmocytomas, 78% of which were located to the upper air passages and oral cavity. Neither among their own nor among previously reported cases was there any instance of primary plasmocytoma in the spleen.

Villa (3) reported on a patient with plasmocytoma of the spleen as well as plasmocytic infiltration of the liver and bone marrow. This patient had marked hypergammaglobulinemia with serum gamma globulins of about 6 g and total proteins of about 10 g per 100 ml. It is unclear whether the increase in gamma globulins was monoclonal or polyclonal (4). The markedly elevated gamma globulins remained unchanged for nine months following splenectomy, but were normalized two years later. Since that time the patient has remained in good health, with normal serum proteins (3, 4). If one accepts Villa's

patient as a case of plasmocytoma in the spleen, the patient most likely had widespread disease at the time of splenectomy. The recovery a year after splenectomy might have been due to the removal of the prevailing splenic involvement.

We have been unable to find reports in the literature of primary plasmocytoma in the spleen without involvement of other organs.

The serum protein changes in solitary plasmocytoma are similar to those of generalized myelomatosis, but as a rule less pronounced, corresponding to the smaller number of plasma cells involved (6). In more than 150 personally examined cases of myelomatosis Waldenström saw no instance of polyclonal increase of immune globulins (6).

CASE REPORT

A man born in 1929 was admitted to the section of Hematology, Rikshospitalet, on February 24, 1969. He had previously been in good health. During the last few months prior to admission, however, he had felt that his usual energy was lacking. A medical examination in January 1969 by his family physician revealed a sedimentation rate of 107 mm and total serum proteins of 9.5 g/100 ml with 4.3 g gamma globulins. The patient was then admitted to hospital for further studies.

Physical examination on admission revealed a pea-sized lymph node on each side of the neck, otherwise no enlargement of lymph nodes. One had the impression of a mass below the left costal margin, but physical examination was difficult because of well developed abdominal muscles. Physical examination was otherwise negative.

Laboratory findings: Nitric acid ring test showed no proteinuria. Erythrocyte sedimentation rate was 100 mm, hemoglobin 13.3 g, erythrocytes 4.7 mill., leukocytes 6,600, platelets 270,000 per mm³.

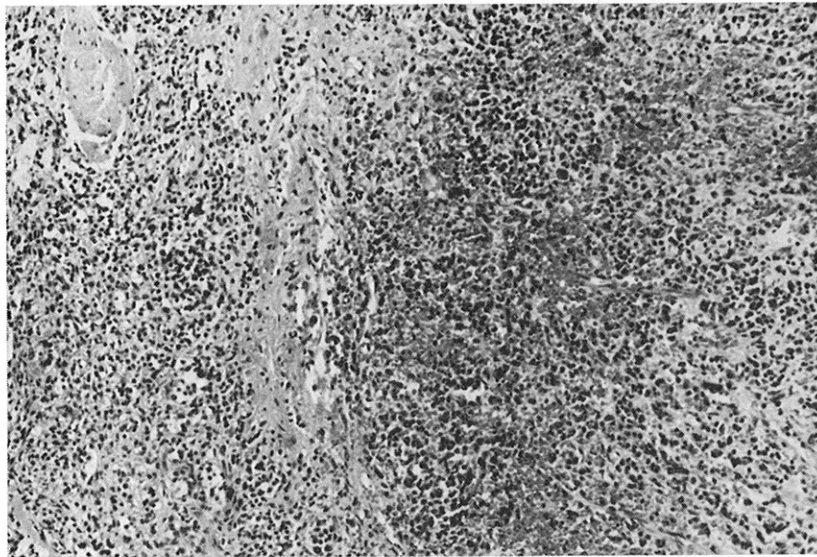


Fig. 1. The tumor tissue (shown on the right) is separated from the remaining splenic tissue by strands of connective tissue. In the tumor tissue plasma cells grow diffusely in a vascular stroma, completely effacing the normal splenic structures. Hematoxylin-eosin. $\times 24$.

Peripheral blood smear showed normal erythrocytes. Sixty per cent of the leukocytes were segmented neutrophil granulocytes, 30% were lymphocytes. Bone marrow smear was slightly hypocellular. The erythropoiesis was normoblastic and amounted to 20% of the nucleated cells in the bone marrow. The granulocytopoiesis was normal in extent and distribution. Mature lymphocytes amounted to 15% of the nucleated cells. There was no increase in plasma cells. The number of megakaryocytes appeared normal.

Bone marrow biopsy showed a cellular marrow with normal distribution of the different cell types and stages.

Biopsy of the enlarged lymph node on the neck

showed chronic lymphadenitis, with only a few scattered plasma cells.

Antistreptolysin titer was 1,280 (normal < 200). Anti-staphylolysin titer 16 (normal 2 or less). Plasma fibrinogen 0.63/100 ml (normal 0.2-0.4). Total serum proteins were 10.1 g/100 ml (normal 6.5-8).

Serum electrophoresis showed a marked polyclonal increase in gamma globulins amounting to 5.2 g (normal 0.9-1.7). Immune electrophoresis showed a marked polyclonal increase in gamma G, quantitated to 5 g/100 ml. No increase in gamma A or in gamma M demonstrated.

Bromsulphalein test (5 mg/kg body weight) showed 1.4% retention in 45 min. Serum glutamic pyruvic

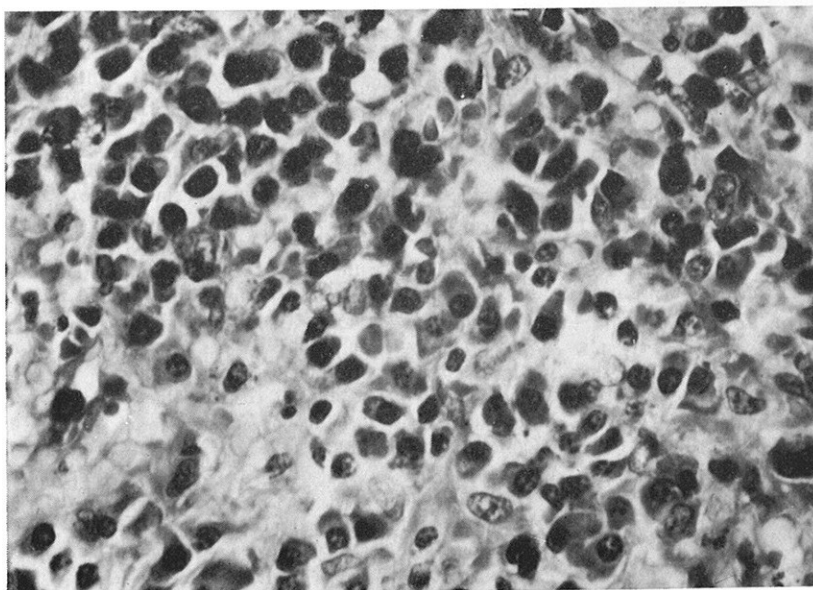


Fig. 2. This high-power micrograph illustrates how the plasma cells in the tumor are growing diffusely, with only a few reticulum cells among them. The plasma cells are in general similar to normal plasma cells, although they vary more in size and configuration. Hematoxylin-eosin, $\times 645$.

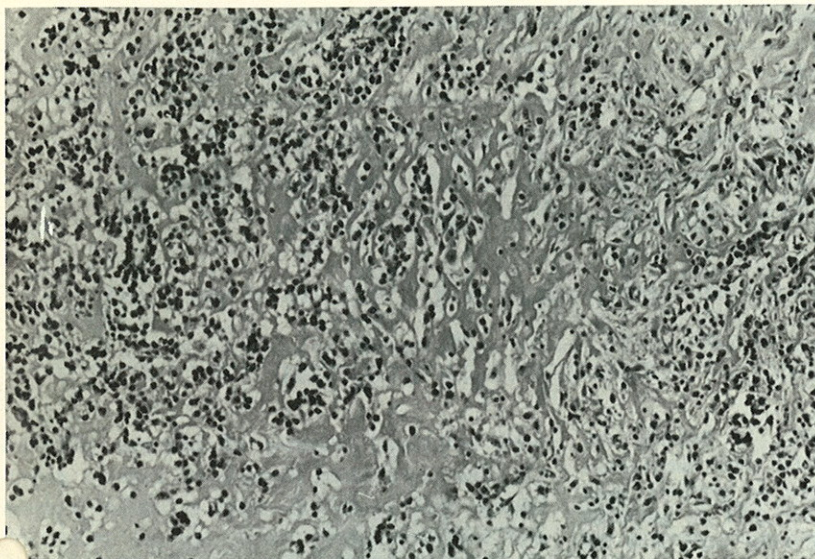


Fig. 3. This picture was taken from an area which macroscopically appeared white and translucent. It shows how plasma cells are lying interspersed in a hyaline and acellular stroma resembling amyloid. Hematoxylin-eosin, $\times 24$.

transaminase (SGPT) was 5 U/l. Serum urea 36 mg, creatinine 0.8 mg/100 ml. Serum calcium 4.1 mEq/l, alkaline phosphatase 29 U/l. Roentgenogram of the chest normal. The tuberculin test was weakly positive. Intravenous pyelography showed normal kidneys and renal pelves. The splenic shadow, however, was seen to be enlarged with several calcifications. Celiacography verified the splenic enlargement. Roentgenograms of the skull, column and pelvis were normal.

The nature of the splenic enlargement remained unknown, and splenectomy was done on March 6, 1969. The spleen weighed 1,030 g. In most places there was a small zone of normal splenic tissue beneath the capsule. The greater part of the spleen was replaced by a soft tumor which was separated from the sur-

rounding splenic tissue by an irregular and thin capsule. The cut surface of the tumor was yellow or brown with scattered areas which had a rubbery consistency and a translucent, white appearance. Microscopically the peripheral splenic tissue showed no pathological changes. It was separated from the tumor tissue by strands of connective tissue (Fig. 1). The tumor itself was very cellular. The majority of these cells were plasma cells, but in some places many reticulum cells and groups of lymphocytes were also found. Some of the plasma cells were binucleated. Apart from that, they looked normal, and few mitoses could be found (Fig. 2). Hemorrhages and groups of hemosiderin-laden macrophages were present in several sections. Corresponding to the white translucent areas were found hyaline masses

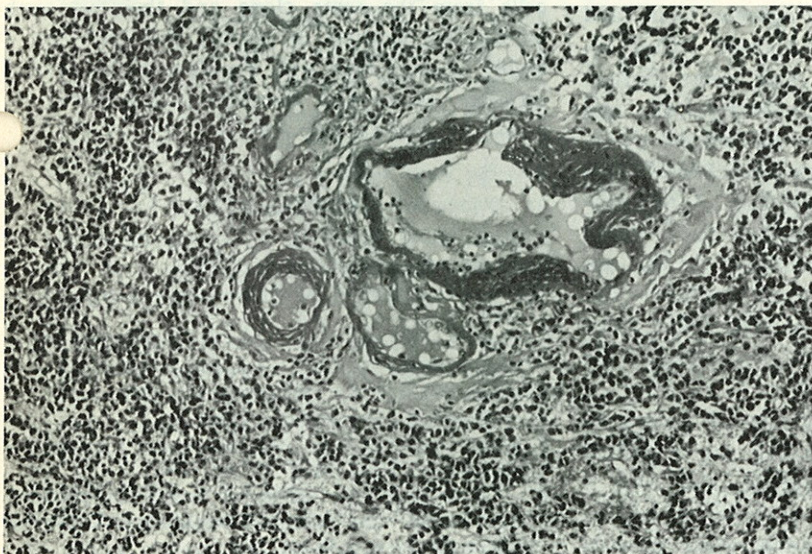


Fig. 4. After staining with methyl violet many vessel walls showed metachromasia, as seen in the center of this picture. The vessels are surrounded by tumor tissue. Methyl violet, $\times 24$.

Table I. Serum proteins and some other laboratory values before and after splenectomy

	Jan.	Feb.	Mar.	Mar.	Mar.	Apr.	May	Jun.	Aug.	Oct.
Date	14	25	6	19	24	22	6	18	6	7
ESR (mm)	107	100		55	55	12	5	2	2	2
Total proteins (g/100 ml)	9.5	10.1		7.5	6.5	7.9	7.6	7.4	7.4	7.5
Gamma globulins (g/100 ml)	4.3	5.2		2.4	1.9	2.3	2.2	2.3	2.1	2.0
Antistreptolysin titer		1280			320	240	200			240
Antistaphylolysin titer		16			4	2	2			1
Fibrinogen (g/100 ml)		0.63			0.63	0.41				0.43
Hemoglobin (g/100 ml)		13.3		12.7	12.1	15.1	15	15.6	16.7	16.0
Serum iron (μ g/100 ml)		35			40	50				90
TIBC (μ g/100 ml)		240			280	330				312

with groups of plasma cells (Fig. 3). Staining with Congo red showed only a doubtful green birefringence when these areas were examined in polarized light. Similarly, staining with Thioflavine gave an unspecific silvery blue fluorescence. On the other hand, both the hyaline masses and many of the vessels revealed a typical metachromasia after staining with methyl violet (Fig. 4). These findings suggested that the hyaline substance could be amyloid, or paraamyloid, deposits.

The patient developed a transient paralytic ileus post-operatively, but afterwards made an uneventful recovery.

In the course of the first six weeks following splenectomy, the erythrocyte sedimentation rate, serum protein changes, antistreptolysin and antistaphylolysin titers all became practically normalized (Table I).

DISCUSSION

Roentgen celiacography as well as cross-sectioning of the removed splenic tumor showed a small rim of normal spleen at the upper end of the grapefruit-sized tumor. The tumor was sharply delineated from the normal splenic tissue and was quite solid. Microscopy showed diffuse infiltration of plasma cells, with some fibrous bands, scattered areas of paraamyloidosis, and complete extinction of all normal splenic tissue. There were no features suggesting Boeck's sarcoid, tuberculosis or other infection. A non-neoplastic plasma cell granuloma like those found in the conjunctiva, and occasionally in the upper air passages, is considered unlikely. The histological examination leaves little doubt about the diagnosis of plasmocytoma originating in the spleen.

The typical polyclonal increase of gamma G in our patient is highly unusual for a patient with plasmocytoma. Our patient also had a moderate increase in antistreptolysin as well as antistaphylolysin titer, like many other patients

with polyclonal increase of immune globulins (5). The pronounced increase of immune globulins, as well as the elevation of antistreptolysin and antistaphylolysin titres, became practically normalized within six weeks following splenectomy. There can therefore be little doubt about the spleen being the site of the abnormal gamma G production.

The polyclonal increase of immune globulins in our patient with solitary plasmocytoma does not fit with the well established concept of myelomatosis as a neoplastic proliferation of a single clone of plasma cells. Our patient resembles those rare cases of lymphoproliferative disorders with polyclonal elevation of immune globulins (5).

One might consider the possibility of two different types of neoplastic proliferation of plasma cells and lymphocytes: the usual type having all cells from the same clone and therefore with monoclonal increase in gamma globulins; and a highly unusual type with proliferation of several different clones and therefore a polyclonal increase in gamma globulins.

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