ORIGINAL ARTICLE

HAIRY CELL LEUKEMIA: AN AUTOPSY STUDY

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Summary: Autopsy study from 21 patients with hairy cell leukemia was performed. All patients had the expected widespread involvement of the hematopoietic system. Leukemic infiltration of lymph nodes was detected in 12 cases. Liver involvement was present in 19 patients, leukemic infiltration ranged from focal portal and sinusoidal infiltration to massive infiltration that effaced the hepatic architecture. Twelve patients showed leukemic infiltration of the spleen, remaining 9 patients underwent previous splenectomy for massive splenomegaly. We also found leukemic infiltration of kidneys in four cases, two patients showed leukemic involvement of the lungs. The cause of death was related to impaired immunity (sepsis, bronchopneumonia, etc.) in the majority (73 %) of cases.

Key words: *Hairy cell leukemia; Tricholeukemia; Trichocellular leukemia; Autopsy; Leukemic infiltration; Splenectomy*

First large autopsy study of hairy cell leukemia was performed by Bouroncle et al. in 1958 (1). Authors reviewed autopsy findings in 8 of their 26 patients and they described leukemic involvement of the liver, lymph nodes, lungs, kidneys, pancreas, stomach, small intestine, and adrenals. The immediate cause of death was pneumonia in two patients and rupture of the spleen, carcinoma of the colon, acute adrenal insufficiency, septicemia, pulmonary edema, and massive retroperitoneal bleeding in the remaining six patients, respectively. Autopsy findings were further discussed in the review of literature (11,12).

In this report, we provide details of the pathologic findings in 21 patients with hairy cell leukemia as seen at the autopsy.

Materials and methods

Since 1963 we performed 21 autopsies on patients with hairy cell leukemia in The Fingerland Department of Pathology in Hradec Králové, Czech Republic. For this report, microscopic sections from each autopsy were stained with H&E and reviewed. Acid-fast staining was performed in one case (case 14).

Results

The significant autopsy findings for each of the 21 patients are summarized in Table 1.

In our series is 17 men (81 %) and 4 women (19 %), male to female ratio is about 4,25:1, corresponding with the literature (1,2,6). The median age of patients was 49 years (range, 18–81 years).

In general, the most pertinent observations are those related to infectious complications and to the pattern of infiltration by the leukemic cells. Although it was difficult, in some instances, to distinguish between small lymphocytes and hairy cells in tissue in which the infiltrate was scant, in most cases the larger size of the nuclei of the hairy cells, the looser chromatin, and the DBA.44 positivity allowed accurate identification of leukemic infiltrates (8).

Bone marrow

Leukemic infiltrates in bone marrow were present in each patient. Grossly, the marrows were described as pale and firm, redder than usual, and soft. Microscopically, leukemic infiltrates were focal, interstitial, or diffuse. There was a marked reduction in normal hematopoiesis in most patients, especially in those with infectious complications. Megakaryocytes were increased in number, occurred in focal clusters, in case 12, where the marrow showed extensive osteomyelosclerosis.

Lymph nodes

Leukemic involvement was present in 12 cases (57 %). In most cases we found leukemic infiltration in retroperitoneal, perirenal, abdominal, mediastinal, and paratracheal lymph nodes. Microscopically, the lymph node architecture was partially or totally effaced in at least some of the lymph nodes from already mentioned 12 patients. In one patient (case 13) we found diffuse, necrotising leukemic infiltration of the thymus. In case 14 we found generalized specific lymphadenitis, the lymph nodes were enlarged up to 3 cm in

Tab. 1: Autopsy findings.

Case	Leukemic Infiltration				Additional Findings	Cause of Death
	Bone Marrow	Lymph Nodes	Liver (Weight)	Other Sites		
1	+	+	+(3000)	spleen	anemia, ascites, hemorrhagic diathesis	cardiac failure
2	+	-	+(2150)	spleen	pulmonary artery thrombembolism, lung and liver aspergillosis, heart hypertrophy	cardiac failure
3	+	-	+(2670)	-	esophagitis, nodous periarteritis	sepsis
4	+	-	+(2200)	spleen, adrenals	esophagitis, colitis, lung edema	sepsis
5	+	+	+(1770)	-	hemorrhagic diathesis, lung edema	GIT bleeding
6	+	+	+(2070)	-	pancytopenia, esophageal candidiasis, appendicitis, peritonitis, endometritis	sepsis
7	+	+	+(2550)	-	pancytopenia, hemorrhagic diathesis, lung aspergillosis	lung aspergillosis
8	+	-	+(1200)	-	femoral vein thrombosis, bowel infarsation, peritonitis	peritonitis, sepsis
9	+	+	+(2280)	kidneys, adrenals, lungs	hemorrhagic diathesis, bronchopneumonia	sepsis
10	+	+	-(3800)	-	bronchopneumonia	bronchopneumonia
11	+	-	+(1310)	-	arteriosclerosis, heart hypertrophy, lung edema	coronary insufficiency
12	+	+	+(3290)	hypophysis, lungs, spleen	osteomyelosclerosis, pneumonia	sepsis
13	+	-	+(2500)	thymus, kidneys, skin	lung aspergillosis, bronchitis, thromboflebitis	sepsis
14	+	-	+(1940)	spleen	hemorrhagic diathesis, brain hemorrhage, TB lymphadenitis, sacral decubitus	brain hemorrhage
15	+	+	+(4390)	-	esophagitis, enteritis, peritonitis	sepsis
16	+	+	+(3030)	spleen	anemia, pulmonary artery thrombosis	respiratory insufficiency
17	+	-	+(2100)	kidneys	bronchitis, pneumonia, pleuritis	sepsis
18	+	+	+(2510)	kidneys	erysipelas, bronchitis, mediastinal lymphadenitis	sepsis
19	+	+	-(2510)	spleen	DIC, GIT bleeding, bronchopneumonia, hydrothorax, hydropericardium	aspiration
20	+	-	+(2420)	spleen	bronchitis, diffuse alveolar damage, proctocolitis	diffuse alveolar damage
21	+	+	+(1980)	spleen	gastric peptic ulcer, large bowel diverticulitis, bronchopneumonia	sepsis

diameter. Microscopically, extensive caseous necrosis was found with numerous acid-fast bacilli. Cultures of lymph nodes confirmed the presence of Mycobacterium kansasii.

Liver

The weight of the livers ranged from 1200 to 4390 g (median, 2420 g). Microscopically, 19 liver specimens (90 %) contained portal and sinusoidal infiltrates of hairy cells, remaining 2 livers did not contain leukemic cells (case 10 and 19).

Spleen

The median spleen weight was 1225 g (range, 220–4100 g). In 9 patients splenectomy was provided for splenomegaly. In remaining 12 patients, 10 cases showed massive, diffuse leukemic infiltration with blood-filled pseudosinuses (10), 1 case showed minimal leukemic infiltration, spleen weight was 220 g (case 2). In case 11 we demonstrated only mild enlargement of spleen weight (320 g) but histologically we were not able to demonstrate leukemic cells because the spleen showed extensive degree of autolysis.

Other organs

Kidneys

The median kidney weight was 180 g (range, 140–305 g). Leukemic infiltration was present in 4 cases (19 %), leukemic infiltrates were discrete, mainly localized around

vessels. The renal weight did not correspond with the density of leukemic infiltration. No evidence of amyloidosis was seen in any case, and Congo red stain showed no reaction in the four cases examined.

Respiratory system

Aside from the organs of the hematopoietic system that were consistently involved with the leukemic process, the pulmonary system demonstrated the most severe pathologic changes, due primarily to infectious complications. Grossly, the lungs were variably described as showing consolidation, edema, and hemorrhage. Microscopically, lung specimens of one of the patients showed disperse, disseminated leukemic cells in the interalveolar septa (case 9); one patient (case 12) showed focal, dense accumulations of hairy leukemic cells.

Suprarenal glands

Adrenals were affected in two patients, leukemic infiltrates were either disperse (case 4), or extensive and diffuse (case 9). There were no hormonal changes noticed.

Pituitary gland was affected only in case 12.

Cause of death

The immediate cause of death as reported in the final pathologic diagnosis was either sepsis or complications of bronchopneumonia in 15 patients, cardiac failure in 3 cases, GIT bleeding in 1, brain hemorrhage in 1, and respiratory insufficiency (thrombosis of pulmonary artery) in 1.

Discussion

In this autopsy series, all patients had leukemic infiltration of bone marrow. The infiltrate was diffuse, interstitial, or focal. These findings correspond with consequent infectious complications, namely sepsis. The hematopoiesis in these cases was extremely reduced, six patients showed extensive hemorrhagic diathesis, three patients had clinical symptoms of anemia or pancytopenia. Other diseases related to reduced immunity include colitis (two cases), peritonitis (2 cases), esophagitis (in the first case non-specific, in the second case esophageal candidiasis). Mycotic infectious further included lung (case 7), and lung + liver aspergillosis (case 2).

The severity of bone marrow infiltration did not correspond with possible previous splenectomy which was performed in 9 cases.

The findings of significant leukemic infiltration of lymph nodes was somewhat unexpected. Hairy cell leukemia is a disorder associated with massive splenomegaly, but peripheral lymphadenopathy is found in only 5 % to 25 % of patients at the time of diagnosis (4,5,7,9). In this series, 57 % of the patients were found at the time of autopsy to have lymph nodes hairy leukemic infiltration in the retroperitoneal, perirenal, abdominal, and mediastinal regions. The same anatomic sites were found in one of previous studies (3).

The majority of studied cases had leukemic affection of the liver (90 %), which ranged from focal portal and sinusoidal infiltration to massive infiltration that effaced the hepatic architecture. There was no correlation between the extent of hepatic involvement and the numbers of circulating leukemic cells, an observation also made previously by Yam et al. (13).

The affection of thymus and pituitary gland is very rare, not mentioned in the literature.

On the basis of this study, we believe that autopsy information can provide valuable data for clinical-pathologic correlation in patients with hairy cell leukemia.

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