Pol. Ann. Med., 2010; 17(1): 91–100.

CASE STUDY

HISTIOCYTIC SARCOMA IMITATING TUMOR OF THE PANCREATIC TAIL – A CASE STUDY

Katarzyna Kozielec[1], Zygmunt Kozielec[2], Tomasz Arłukowicz[1], Marian Sulik[2],

[1] *Department of Gastroenterology, Provincial Specialist Hospital in Olsztyn, Poland*
[2] *Chair of Pathomorphology, Faculty of Medical Sciences, University of Warmia and Mazury in Olsztyn, Poland*

ABSTRACT

Introduction. Histiocytic sarcoma (HS) is a very rare and diagnostically difficult malignant neoplasm arising from dendritic cells and histiocytes. Its microscopic image is not specific, so the diagnosis of HS requires a wide panel of immunohistochemistry tests to exclude tumors with similar morphology, but of completely different origins. While diagnosing HS, cancers, other sarcomas, lymphomas and malignant melanoma should be excluded as well.

Aim. The aim of this paper was to present a case of HS imitating tumor of the pancreatic tail in a 58-year-old woman.

Case study. Intraoperative diagnosis was as follows: solid-cystic tumor of the pancreatic tail region, penetrating into the mesocolon and occluding the colon by pressing against it. Resection of the pathologic mass and tail of the pancreas, as well as total colectomy were performed. On the basis of postoperative histopathologic evaluation of the surgical specimen and a wide immunohistochemical panel, we excluded epithelial and myogenic origins of the tumor. Gastrointestinal stromal tumor, extramedullary myeloid tumor, lymphoma, neural tumors and malignant melanoma were also excluded. Histopathologic and immnohistochemical findings, compared to other authors' findings led us to the diagnosis of histiocytic sarcoma. Complete resection of the tumor was performed, with sufficient margins of the healthy tissues.

Physical examination and imaging performed three months after the surgery revealed features of the local recurrence, infiltration of the back wall of the stomach with a major compression of the gastric lumen. Metastatic foci in regions of left ap-

Corresponding address: Katarzyna Kozielec, Oddział Gastroenterologiczny, Wojewódzki Szpital Specjalistyczny w Olsztynie, ul. Żołnierska 18, 10-561 Olsztyn, Poland; e-mail: kasia-3-kmk@o2.pl

Received 26.04.2010, accepted 19.06.2010

pendages and lower pole of the left kidney, multiple small hypodensic areas in the liver and enlarged paraaortal and mesenterial lymph nodes were also found.
Discussion. HS is a very rare and diagnostically difficult malignant tumor. Microscopic image is non-specific, that is why the diagnosis of HS requires a wide histochemical panel to exclude tumors with similar morphology, but of completely different origins. Analysis of negative immunohistochemical studies, results of: CD68, LCA(CD45), CD4, CD30, CD31, Fascin, CD43, CD15, CD34, and comparing them with the results obtained by other authors led us to the diagnosis of HS. Clinical prognosis is negative and the most frequent course of the disease is aggressive.
Conclusions.
1. Despite the rare prevalence of the tumor, there are numerous, well documented and immunohistochemically confirmed reports of histiocytic sarcoma and its gastrointestinal localization. That is why in a differential diagnosis of gastrointestinal tract-located tumors, sporadically occuring neoplasms should be also taken into account.
2. Diagnosis of HS requires a wide panel of immunohistochemistry tests to exclude tumors with similar morphology, but of completely different origins.
3. Recurrence of a neoplastic process in the described case confirms that, despite a surgical and microscopically total excision of the tumor, HS prognosis is negative and the course of the disease is very aggressive.

Key words: histiocytic sarcoma (HS)

INTRODUCTION

Histiocytic sarcoma (HS) belongs to the group of neoplasms derived from dendritic cells and histiocytes. The WHO classification distinguishes in this group eight types of proliferations of dendritic cells and histiocytes. These are very rare neoplasms, of which HS and interdigitating dendritic cell sarcoma have a negative prognosis and are usually characterised by a very aggressive clinical course [2, 3, 8, 13, 16]. About 10% of patients progress, in a short time, from localised, monoorgan to disseminated, multiorgan variant.

AIM

The aim of this paper was to present a case of HS imitiating tumor of the pancreatic tail in a 58-year-old woman.

CASE STUDY

A 58-year-old woman, non-smoking, denying the use of alcohol, after laparotomy at the age of 19 for a foreign body in a gastrointestinal tract lumen (swallowing of a needle),

complicated by an abscess, was admitted urgently to the Gastroenterology Department of Provincial Specialist Hospital in Olsztyn, in November 2009, for disturbing abdominal symptoms. On admission, the patient complained of constipation, lasting for 6 months, accompanied by a persistent tenesmus, feeling of an incomplete defecation and abdominal pain of a changeable nature. According to a clinical history, in 2003 the abdominal cystic tumor was performed revealing a cyst localized between the pancreatic tail and spleen. Since then, no progression in the imaging studies was observed.

Physical examination revealed paleness, post-surgical scar from the sternum to the pubic symphysis, abdominal obesity, mild systolic, mitral valve murmur, palpational tenderness and a mass of the left lumbar region.

Laboratory studies revealed: normocytic anemia (Hb 10.0 mg/dL), thrombocytosis. Tumor markers (CEA and Ca 19.9) were within normal range.

Colonoscopy findings: tumor, almost entirely occluding the intestinal lumen, 40 cm from anal sphincters (Fig. 1). Specimens for histopathological examinations were taken. Microscopic verification showed only the presence of necrotic-inflammatory masses. Abdominal sonography extended the diagnosis: a solid-cystic tumor, 14×8×9 cm, near the pancreatic tail and adhering to the stomach wall was found (Fig. 2, 3). A solitary, hypogenic lymph node was revealed in the near proximity. Other organs of the abdominal cavity were normal. The physician performing USG suggested an intestinal wall origin of the tumor. Upper gastrointestinal tract endoscopy revealed features of erythematous gastropathy with a negative urease test.

Taking into account clinical signs of subileus, with endoscopically and USG suggested presence of a tumor growth, after surgical consultations the patient was qualified for surgery.

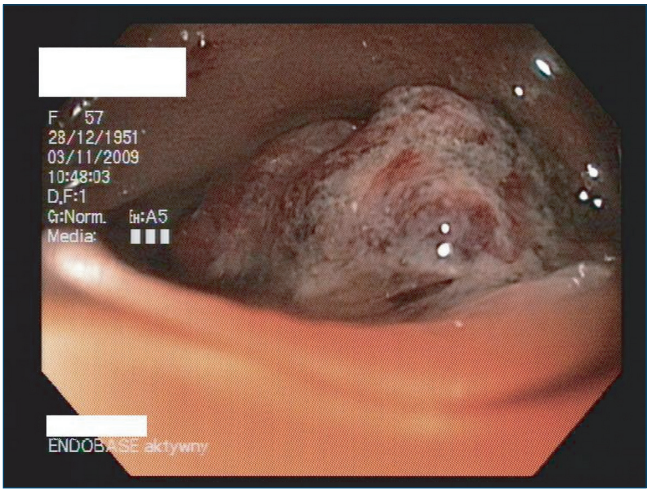

Fig. 1. Colonoscopy. Tumor mass protruding into the colon lumen

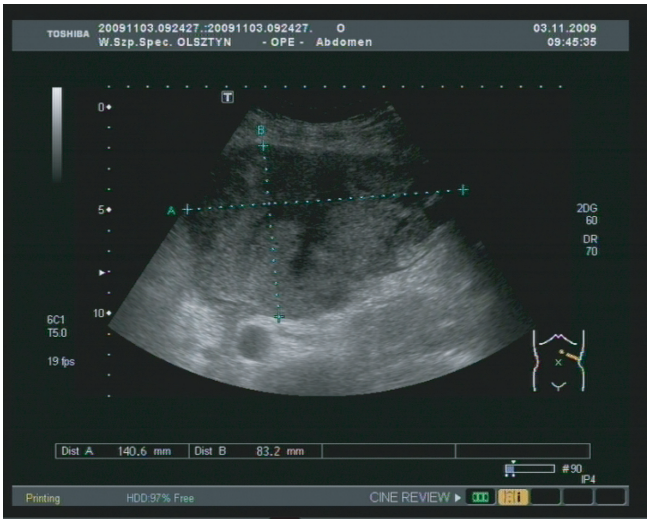

Fig. 2. Abdominal ultrasonography. Tumor-like, cyst-structured lesion in the area of the pancreatic tail

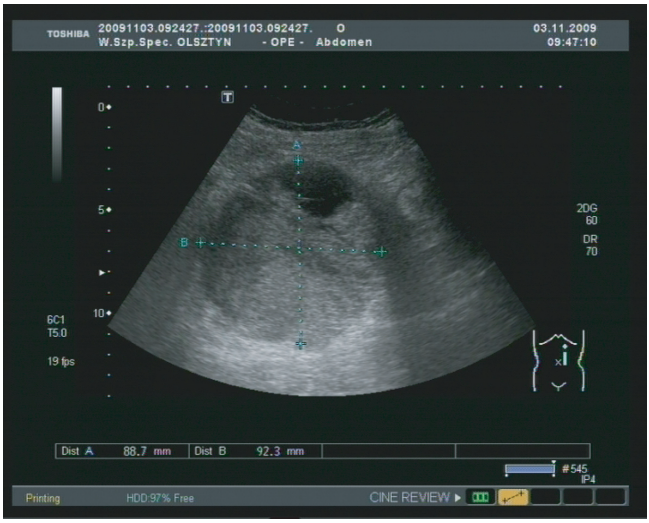

Fig. 3. Abdominal ultrasonography. Tumor-like, cyst-structured lesion in the area of the pancreatic tail

Intraoperative diagnosis was as follows: solid-cystic tumor of the pancreatic tail region, penetrating into the mesocolon and occluding the colon by pressing against it.

Resection of the pathologic mass and tail of the pancreas as well as total colectomy were performed. Morphotic elements were infused. Surgery was not followed by any complications. The patient was released from hospital on the 9th day.

Histopathological evaluation of the surgical specimen, with a wide immunohistochemical panel, led to the diagnosis of HS. Tumor was resected totally, with sufficient microscopic margins of the healthy tissues.

The patient remained under Oncology Outpatient care. A month later USG was performed revealing liquid containing an echogenic mass, 9.5 × 5 × 3 cm, in the left hypochondriac region, and a spleen enlargement.

DISCUSSION

HS is a very rare and diagnostically difficult malignant tumor. Its name was introduced in 1970 by Mathe et al. [10]. Microscopic image is non-specific, that is why the diagnosis of HS requires a wide histochemical panel to exclude tumors with similar morphology, but of completely different origins. While diagnosing HS, cancers, other sarcomas, lymphomas and malignant melanomas should be excluded.

In the described case, the surgical specimen consisted of part of an intestine 25 cm long with a mesocolonic tumor: 15 × 13 × 10 cm, cherry-brownish, infiltrating the intestinal wall. Microscopic examination revealed diffuse tumor infiltration consisting of large, epithelioid cells with acidophilic cytoplasm, with oval to irregular nuclei, often with irregularly folded surface, some of them with clearings containing distinct nucleoli; numerous multinucleated cells were also present (Fig. 4, 5). Part of the tumor showed sarcomatoid areas – spindle cells arranged in storiform pattern (Fig. 6, 7). Immunohistochemical tests excluded: epithelial (CK7–, CK20–, CDX2–), and myogenic origins of tumor (Desmin–, SMA–), gastro-intestinal stromal tumor (GIST) (CD117–), extramedullary myeloid tumors (MPO–), lymphoma (CD3–,CD20–), nervous system derived tumors and melanoma (S100–), metastasis from ovary (CA125–). Analysis of those negative immunohistochemical studies, as well as following results: CD68+, LCA(CD45)+, CD4+, CD30–, CD31+, Fascin–, CD43–, CD15±, CD34–, and comparing them with the results obtained by other authors [4–7, 9, 13, 16] led us to the diagnosis of HS.

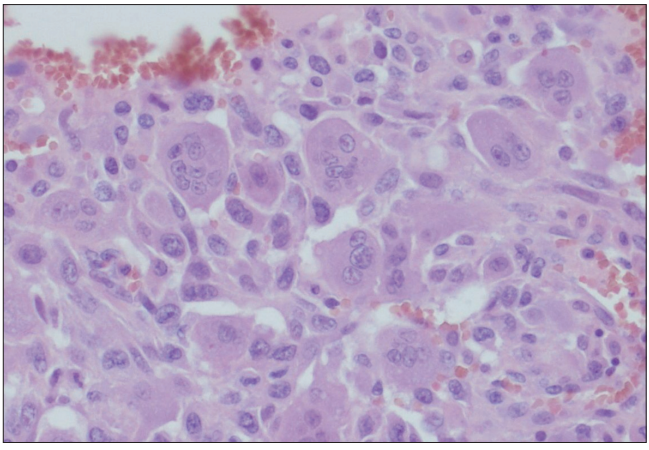

Fig. 4. HS. Multinucleated giant cells [HE 200×]

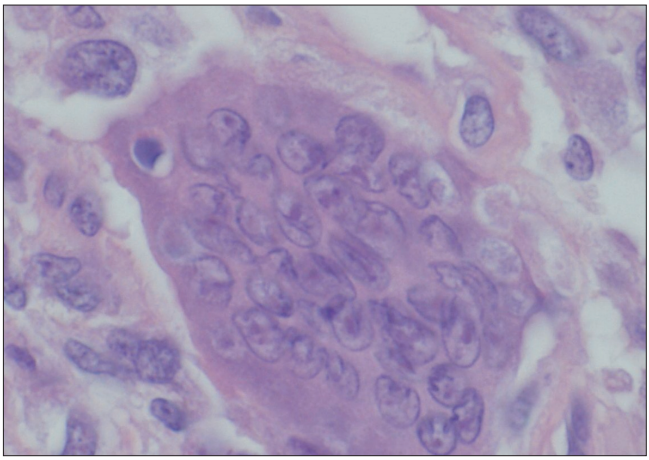

Fig. 5. HS. Multinucleated giant cell [HE 600×]

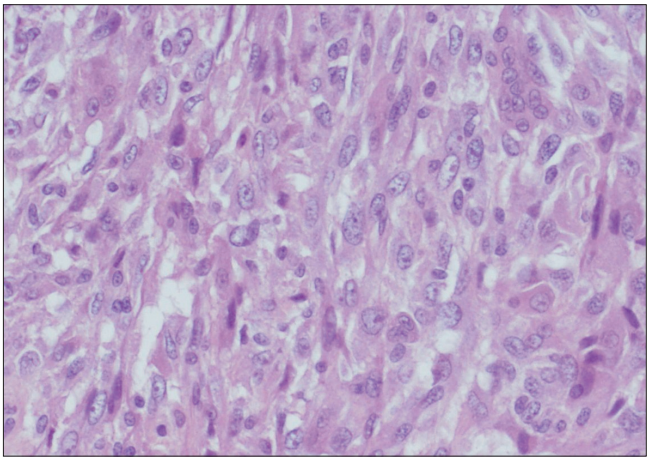

Fig. 6. HS. Sarcomatoid architecture of the tumor [HE 200×]

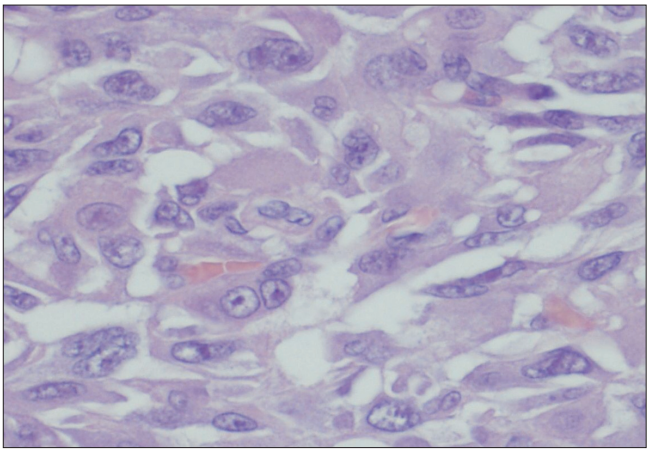

Fig. 7. HS. Sarcomatoid architecture of the tumor [HE 200×]

The patient was hospitalized again in March 2010 with symptoms of gradually increasing weakness, headache and dizziness, lack of appetite, meteorism, nausea, and dark stools present.

In laboratory tests severe anemia and increased levels of glucose were observed.

Endoscopic examination of the upper gastrointestinal tract revealed infiltration of the posterior wall of the stomach, with the presence of external pressure severly occluding gastric lumen. Mucous membrane of a changed surface was fragile and easily bleeding.

Cystic tumor imaging of abdominal cavity organs showed features of tumor recurrence with dissemination and partial occlusion of intestinal lumen – in the anatomical region of pancreas head and tail a mass, 12×14×15 cm, was found. It was pressing upon the stomach, reaching a pre-renal fascia (of a left kidney) with its focal infiltration, dislocating duodenum and jejunum loops. A similar mass, about 6×7×9 cm, was observed on the level of umbilicus, posteriorly from modelated, dislocated anteriorly jejunal loop, with the intestinal wall infiltration. Smaller foci were present in the region of left appendages on the level of lower left kidney pole (Fig. 8). Multiple hypodense foci, up to 4.5 cm, in the liver (Fig. 9) and multiple enlarged paraaortal and mesenteric lymph nodes were also found.

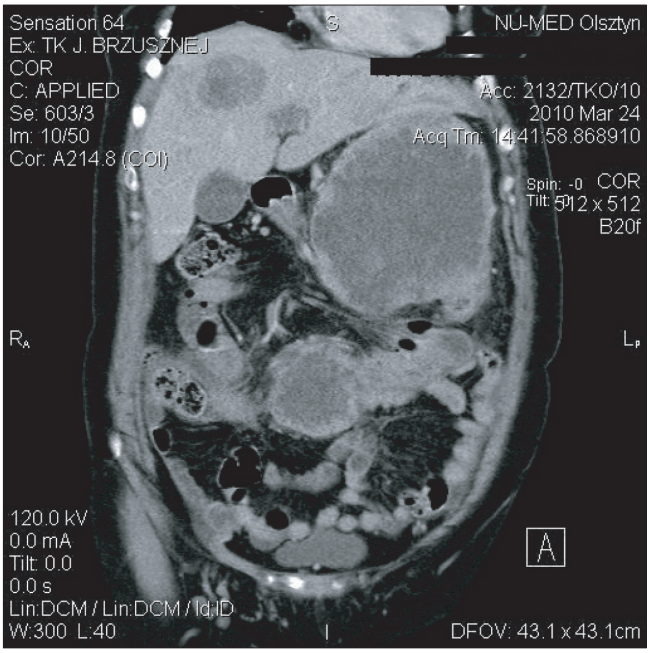

Fig. 8. Abdominal CT scan. Tumor masses in multiple localizations

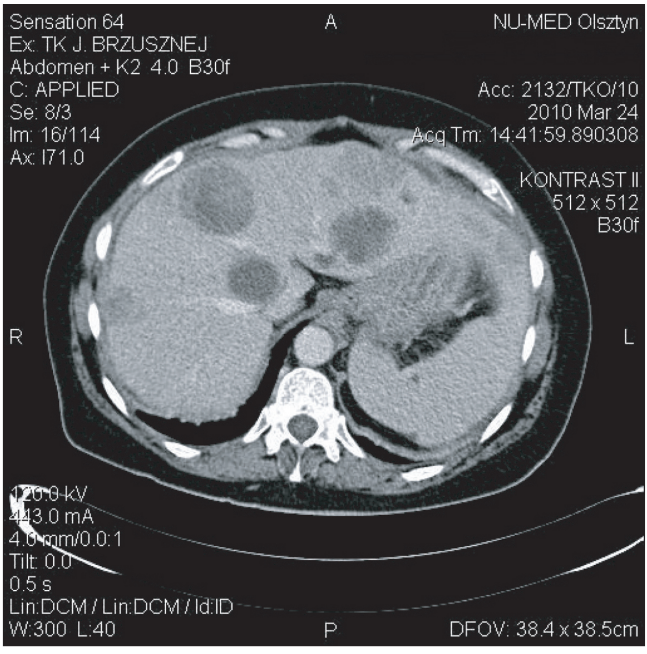

Fig. 9. Abdominal CT scan (liver). Multiple hypodensic areas

After oncology consultations, the patient was qualified for symptomatic treatment. Antalgic, spasmolytics and anti-emetic drugs were administered. After normalization of blood morphotic elements, the patient was released from hospital for further hospice, palliative treatment.

Despite the rare occurrence of histiocytic sarcoma, there are quite a few well documented and immunohistochemically confirmed cases of this tumor, including its gastrointestinal localization [1, 2, 11, 12, 15]. Clinical prognosis is negative and the most frequent course of the disease is aggressive [2, 3, 8, 13, 14, 17].

CONCLUSIONS

1. Despite the rare occurrence of histiocytic sarcoma, there are quite a few well documented and immunohistochemically confirmed cases of this tumor, including its gastrointestinal localization. That is why in a differential diagnosis of gastrointestinal tract-located tumors, sporadically occuring tumors should be also taken into consideration.
2. Diagnosis of HS requires a wide immunohistochemical panel to exclude tumors with similar morphology but of completely different origins.
3. Recurrence of a neoplastic process in the described case confirms that, despite a surgical and microscopically total excision of the tumor, HS has a negative prognosis and its course is very aggressive.

REFERENCES

1. Alvaro T., Bosch R., Salvadó M. T., Piris M. A.: *True histiocytic lymphoma of the stomach associated with low-grade B-cell mucosa-associated lymphoid tissue (MALT)-type lymphoma.* Am. J. Surg. Pathol., 1996; 20: 1406–1411.
2. Copie-Bergman C., Wotherspoon A. C., Norton A. J., Diss T. C. Isaacson P. G.: *True histiocytic lymphoma. A morphologic, immunohistochemical, and molecular genetic study of 13 cases.* Am. J. Surg. Pathol., 1998; 22: 1386–1392.
3. Gonzalez C. L., Jaffe E.S.: *The histiocytoses: clinical presentation and differential diagnosis.* Oncology, 1990; 4: 47–60.
4. Hornick J. L., Jaffe E. S., Fletcher C. D.: *Histiocytic sarcoma: a study of 13 extranodal cases* [abstract]. Modern. Pathol., 2004; 1 (Suppl 1):16A.
5. Hornick, J.L., Jaffe E.S., Fletcher C.D.: *Extranodal histiocytic sarcoma: clinicopathologic analysis of 14 cases of a rare epithelioid malignancy*. Am. J. Surg. Pathol., 2004; 28 (9): 1133–1144.
6. Isaacson P., Wright D. H., Jones D. B.: *Malignant lymphoma of true histiocytic (monocyte/macrophage) origin.* Cancer, 1983; 51 (1): 80–91.
7. Jaffe E. S., Harris N. L., Stein H., Vardiman J. W. (eds.): *WHO classification of tumors. Pathology and genetics of tumours of haematopoietic and lymphoid tissues.* IARC Press, Lyon 2001; 273–290.
8. Jaffe E. S.: *Malignant histocytosis and true histiocytic lymphoma*. In: Jaffe E. S. (ed.): *Surgical Pathology of Lymph and Related Organs.* Saunders, Philadelphia 1995: 560–593.
9. Lauritzen A. F., Delsol G., Hansen N. E., Horn T., Ersboll J., Hou-Jensen K., Ralfkiaer E.: *Histiocytic sarcomas and monoblastic leukemias: a clinical, histologic, and immunophenotypical study.* Am. J. Clin. Pathol., 1994; 102 (1): 45–54.

10. Mathé G., Gerard-Marchant R., Texier J. L., Schlumberger J. R., Berumen L., Paintrand M.: *The two varieties of lymphoid tissue 'reticulosarcomas', histiocytic and histioblastic types.* Br. J. Cancer, 1970; 24 (4): 687–695.
11. Miettinen M., Fletcher C. D., Lasota J.: *True histiocytic lymphoma of small intestine: an analysis of two S-100 protein-positive cases with features of interdigitating reticulum cell sarcoma.* Am. J. Clin. Pathol., 1993; 100 (3): 285–292.
12. Milchgrub S., Kamel O. W., Wiley E., Vuitch F., Cleary M. L., Warnke R. A.: *Malignant histiocytic neoplasms of the small intestine.* Am. J. Surg. Pathol. 1992; 16 (1): 11–20.
13. Pileri S. A., Grogan T. M., Harris N. L., Banks P., Campo E., Chan J. K., Favera R. D., Delsol G., de Wolf-Peeters C., Falini B., Gascoyne R. D., Gaulard P., Gatter K. C., Isaacson P. G., Jaffe E. S., Kluin P., Knowles D. M., Mason D. Y., Mori S., Müller-Hermelink H. K., Piris M. A., Ralfkiaer E., Stein H., Su I. J., Warnke R. A., Weiss L.-M.: *Tumours of histiocytes and accessory dendritic cells: an immunohistochemical approach to classification from the International Lymphoma Study Group based on 61 cases.* Histopathology, 2002; 41 (1): 1–29.
14. Ralfkiaer E., Delsol G., O'Connor N. T., Brandtzaeg P., Brousset P., Vejlsgaard G. L., Mason D. Y.: *Malignant lymphomas of true histiocytic origin. A clinical, histological, immunophenotypic and genotypic study*. J. Pathol., 1990; 160 (1): 9–17.
15. Seo I. S., Henley J. D., Min K. W., Yum M. N.: *True histiocytic lymphoma of the esophagus in an HIV-positive patient: an ultrastructural study.* Ultrastruct. Pathol., 1999; 23: 333–339.
16. Turner R. R., Wood G. S., Beckstead J. H., Colby T. V., Horning S. J., Warnke R. A.: *Histiocytic malignancies: morphologic, immunologic, and enzymatic heterogeneity.* Am. J. Surg. Pathol., 1984; 8 (7): 485–500.
17. van der Valk P., te Velde J., Jansen J., Ruiter D. J., Spaander P. J., Cornelisse C. J., Meijer C. J.: *Malignant lymphoma of true histiocytic origin: histiocytic sarcoma. A morphological, ultrastructural, immunological, cytochemical and clinical study of 10 cases.* Virchows Arch. A. Pathol. Anat. Histol., 1981; 391: 249–265.