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Original article

Clinicopathological features of myeloid sarcoma: Report of 39 cases and literature review

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ABSTRACT

Objective: To explore the clinicopathological features of myeloid sarcoma (MS).
Methods: We retrospectively analyzed the clinicopathological features of patients with MS and reviewed the relevant literature.
Results: There were 39 patients (20 male and 19 female, with a ratio of 1.1:1) aged 2–62 years (median: 33 years; mean: 33.4 years), with 53.9% patients in the 21–50 years age group. The clinical manifestations varied and were dependent on the lesion location. Immunohistochemistry and special staining showed that the positive rates for myeloperoxidase, CD43, CD34, CD117, CD68, Lysozyme, CD99, and naphthol AS-D chloroacetate esterase were 92.1% (35/38), 91.3% (21/23), 44.8% (13/29), 42.3% (11/26), 61.1% (11/18), 100.0% (5/5), 78.6% (11/14), and 55.6% (10/18), respectively. Histologically, the cells were predominantly arranged diffusely, and the solitary infiltrating cells may show an acinus-like arrangement, while some cells were arranged in linear or "Indian file" pattern. Obvious small nucleoli were seen in most cases. The nuclei were predominantly round or oval, although eccentric, kidney-shaped, and lobulated nuclei were also observed. A varied number of eosinophilic granulocytes were found in most cases. The "starry sky" appearance was observed in a few patients. Among 18 patients, the survival rate was 38.9% and the median survival time was 28 months. MS occurred following allogeneic stem cell transplantation in 5 cases. There were 14 de-novo (45.2%,14/31) and 17 secondary MS (54.8%, 17/31). As for de-novo MS, female rates was higher (64.3% vs. 29.4%), older average years (41.4 vs. 31.1 years), older middle years (47.5 vs.32 years).
Conclusions: The combined application of morphology, immunohistochemistry, and special staining may facilitate the diagnosis of this malignancy. Surgery plus chemotherapy remains the most common treatment for MS. The prognosis of MS was bad.

1. Introduction

Myeloid sarcoma (MS) is an extramedullary proliferation of myeloid blasts. By definition, the infiltrates efface the underlying tissue architecture. Tumor cells lack of specific B- or T-cell markers, and express myeloid or myelomonocytic markers such as CD13, CD33, myeloperoxidase (MPO), CD14, or CD64. MS also expresses CD68, CD117, CD34, lysozyme, CD43, CD163, etc [1]. MS can occur as an isolated lesion but also preceding, concurrently with, or following myeloproliferative disorders [2]. According to the World Health Organization Classification of Tumors of Hematopoietic and Lymphoid Tissues, MS includes granulocytic sarcoma (GS), primitive monocytic sarcoma, and tumor composed of three blood cells (white blood cells, red blood cells, and platelets). Among these three types, GS has the highest proportion (accounting for ~96.3%). Primitive monocytic sarcoma is rare [3], and tumors composed of three types of blood cells are even rarer. GS develops following the extramedullary proliferation and infiltration of primitive or immature granulocytes. The presence of myeloperoxidase in immature granulocytes means that the tumor may become green after air exposure. Thus, it is also known as chloroma. In the current study, we retrospectively analyzed the clinicopathological data of 39 patients with MS to facilitate the diagnosis and treatment of this malignancy.

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Table 1
Disease-onset locations and clinical manifestations in 39 patients.

Locations[a]	n	%	Clinical manifestations
Cervical lymph nodes	7	16.3	Swelling of multiple superficial lymph nodes
Inside and outside vertebral canal	6	14.0	Low back pain accompanied by lower limb pain/numbness/weakness, hypoesthesia, urinary retention, bloating, and constipation
Skin and subcutaneous tissue	4	9.3	Several peanut-sized skin masses in the whole body
Mediastinum	3	7.0	Irritating dry cough
Uterus	3	7.0	Vaginal bleeding, menstrual disorders, and cervical bleeding
Left ovary	2	4.7	Menstrual disorder
Salivary gland	2	4.7	Difficulty in opening mouth; numbness around the mouth
Left testicle	2	4.7	Presented with suspected epididymitis
Gastrointestinal tract	2	4.7	Periumbilical pain, constipation, and incomplete intestinal obstruction
Soft tissue	2	4.7	Pain and numbness, along with correspondingly radiating pain
Nasal passages	1	2.3	Nasal airway obstruction; bloody nasal discharge
Vestibule of larynx	1	2.3	Hoarseness
Right external auditory meatus	1	2.3	Bloody discharge
Retroperitoneal area	1	2.3	Painless mass
Spleen	1	2.3	Remarkably enlarged spleen; traumatic rupture of spleen
Right liver	1	2.3	Hypoechoic nodule
Left kidney	1	2.3	Low back pain/discomfort accompanied by urinary frequency and dysuria
Abdominal cavity	1	2.3	Abdominal mass for over 10 years
Central nervous system	2	4.7	Headaches, head swelling, and vomiting

[a] Occurred at multiple locations in 4 cases.

2. Materials and methods

We retrospectively analyzed the clinicopathological data of 39 patients with MS who were diagnosed in the Department of Pathology of the Chinese PLA General Hospital from October 2005 to April 2015. The patients were aged 2–62 years (median: 33 years; mean: 33.4 years): 10 (25.6%) were aged ≤20 years, 21 (53.9%) 21–50 years, and 8 (20.5%) >50years. This means that most patients were middle-aged. There were 20 men and 19 women, yielding a male/female ratio of 1.1: 1.

Most tissue specimens for pathological examinations were gray–white, gray–red, gray-yellow, or gray–brown; in one case the tissue was pale green during intraoperative exploration but turned gray–white after fixation. The specimens were fixed in 10% neutral-buffer formalin solution, embedded in paraffin wax, sectioned to 4 μm, and stained with hematoxylin and eosin (HE). A tumor of hematopoietic and lymphoid tissue was suspected, and immunohistochemistry and special histochemistry staining were then performed. Some patients also received genetic testing.

Immunohistochemistry was performed using an EnVision (Dako) two-step visualization method. After high temperature and high pressure citrate buffer retrieval, sections were developed with diaminobenzidine (DAB) and counterstained with hematoxylin.

During naphthol ASD chloroacetate esterase (NAS-DCE) staining, the deparaffinized sections were put into the incubation solution and incubated at room temperature for 30 min. After the slides were rinsed with running water for 1 min and then with distilled water again, nuclei were counterstained with 2% methyl green. The sections were gently washed with running water and air-dried, then maked be transparent with xylene, finally mounted. The incubation solution was prepared as follows: 0.2 ml 6-azo-pararosaniline was slowly added to 35 ml 0.067 M phosphate buffer, with gentle shaking. When it was well mixed, 1 ml NAS-DCE was added, with increasingly strong vibration and filtrated following formation of pink precipitation.

SPSS17.0 software package was used for statistical analysis. Univariate survival analysis was carried out according to Kaplan-Meier, whereas differences in survival curves were assessed with the log-Rank test.

3. Results

3.1. Clinical data

The disease-onset locations are shown in Table 1. The clinical manifestations were diverse and related to the location, where they caused symptoms of oppression and blockage (Table 1). The non-specific symptoms included anemia, fatigue, dizziness, poor appetite, fever, and swollen superficial lymph nodes.

MS occurred concurrently with acute myeloid leukemia (AML) in 8 cases and chronic myeloid leukemia (CML) in 1. There were 7 cases of MS which occurred following AML; In one of these 7 cases, GS was initially diagnosed in several bones, which was treated with routine chemotherapy for acute non-lymphocytic leukemia. One year after completion of chemotherapy, the patient had a relapse, which was diagnosed as acute promyelocytic leukemia. Haploidentical hematopoietic stem cell transplantation (HSCT) was performed. Eleven months later, the patient developed extramedullary recurrence, which included GS in the central nervous system and in the external auditory canal, followed by intramedullary recurrence. In 5 cases, MS occurred following allogeneic HSCT (allo-HSCT) within 11 months to 11 years. Transplantation was performed because of leukemia in 4 cases and myelodysplastic syndrome (MDS) in 1 case with the longest interval following allo-HSCT.

There were 14 cases de-novo MS (45.2%, 14/31) and 17 secondary(54.8%, 17/31) excepting the other 8 outpatients without medical history (Table 2). In 14 de-novo MS, the patients were aged 7–63 years (median:47.5 years; mean: 41.4 years). There were 5 men (35.7%, 5/14) and 9 women (64.3%, 9/14), yielding a male/female ratio 1:1.8. In 17 secondary MS, the patients were aged 10–62 years (median 32 years; mean: 31.1 years). There were 12 men (70.6%, 12/17) and 5 women (29.4%, 5/17), yielding a male/female ratio 2.4:1. As for de-novo MS, female rates was higher (64.3% vs. 29.4%), older average years (41.4 vs. 31.1 years), older middle years (47.5 vs. 32 years).

3.2. Pathological morphology

Histologically, the cells were mainly arranged diffusely with poorly defined borders, and the solitary infiltrating cells may show