

**CASE REPORT**
**ISOLATED MYELOID SARCOMA**
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**ABSTRACT**

Myeloid Sarcoma (MS) is a tumour mass of myeloblasts or immature myeloid cells in an extra-medullary site. Isolated MS is a rare disease found only seen in case reports. Here, we present the case of a 25 years old man with bilateral cervical lymphadenopathy who was diagnosed as MS on the basis of lymph node biopsy and immunohistochemistry. There is no definite consensus on management of MS and needs prospective trials.

**Key Words:** *Extra-medullary hematopoiesis, Myeloblasts, Myeloproliferative disorders*

**INTRODUCTION**

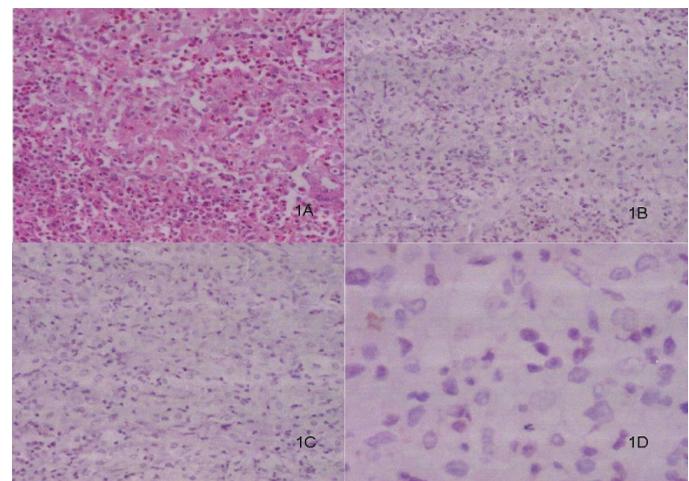
Myeloid sarcoma is a tumour mass of myeloblasts or immature myeloid cells occurring in an extra-medullary site or in a bone. The tumour may occur concurrently with acute or chronic myeloid leukaemia or with other types of myeloproliferative disorders or myelodysplastic syndromes.<sup>1-2</sup> It can also appear as isolated MS which means MS without any evidence of a history of leukemia, MDS, or myeloproliferative neoplasm and a negative bone marrow biopsy. Isolated MS is rare and only seen in limited case reports<sup>3</sup>.

Here we described one of the rare cases of isolated MS happened in the cervical lymph nodes without any signs of leukemia, MDS, and myeloproliferative disorders, and we also shared our experience here during the treatment of the rare case.

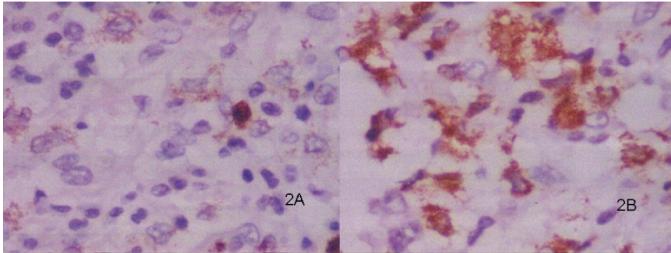
**CASE REPORT**

A 25-year-old man was referred to B. P. Koirala Memorial Cancer Hospital for a complain of bilateral cervical lymph nodes enlargement. A biopsy report from left cervical lymph node tumor tissue brought from the previous hospital showed CD21, CD35, CD23 were negative (Figure1) for the neoplastic cells, but CD68, and myeloperoxidase (MPO) were positive (Figure2). Bone marrow aspirate and biopsy were slightly hypercellular with trilineage hematopoiesis. No blast or immature cells was found in peripheral blood. No evidence of leukemia, myelodysplasia, or any type of myeloproliferative disorder was identified. A diagnosis of nonleukemic MS of the cervical lymph node was made. AML-based induction regimens was given to this patient and the tumor mass shrunk significantly (figure3). Two weeks later the patient got a neutropenia state, G-CSF was used in this patient, and soon the WBC increased to 10000/dl. The tumor mass gradually increased in size. G-CSF

was omitted from his treatment abruptly as the WBC count was continuously increasing. Blood smear and bone marrow aspiration were done in order to exclude the development of AML. The results showed that no evidence of leukemia was found in bone marrow, but there are a lot of immature white blood cells in peripheral blood to with 2% of blast. The second course of induction therapy was given to this patient and the WBC soon decreased and the tumor mass began to shrink. Now this patient is in a good state after the chemotherapy.



**Figure 1: Results from the pathology study showed that the lymph node was infiltrated by tumor cells (1A 400×) and Immunohistochemical staining showed that the tumor cells were negative for lymphocyte marker like CD21 (1B 400×), CD23 (1C 400×) and CD35(1D 1000×) was also negative for the tumor cells**



**Figure 2: Results from Immunohistochemical staining showed that MPO (2A 1000×) and CD68 (2B 1000×) were both positive for the tumor cells which indicated that the tumor cells are myeloid origin.**



**Figure 3: The tumor size shrunk greatly after chemotherapy which showed that DA regimen is effective for this patient**

## DISCUSSION

Myeloid sarcoma (MS) is a kind of extramedullary hematopoiesis out of bone marrow. The main character of MS is an extramedullary proliferation of blasts of one or more of the myeloid lineages in the tissue in which it is found. The normal architecture of the tissue in which it is found is normally disrupted. It has also been addressed as granulocytic sarcoma, myeloblastoma and extramedullary myeloid cell tumor<sup>4</sup>. Isolated MS is a special type of MS which is often misdiagnosed with lymphoma especially when it happens in lymph node<sup>5-6</sup>. Our case belongs to one of such rare cases since it grew in the cervical lymph nodes and we did not find any evidence of previous history of AML or evidence of AML in his bone marrow.

For the diagnosis of MS, obtaining tissue sample is always recommended instead of a fine needle aspiration if the risks of biopsy are reasonable. The morphologic appearance of MS on H & E is decided by the degree of myeloid differentiation. It typically consists of a diffuse and infiltrative population of myeloblasts and granulocytic cell components. The malignant cells are typically large with abundant cytoplasm and large nuclei. Leder stain can help to confirm the myelocytic differentiation which is very helpful in establishing the diagnosis in the isolated MS<sup>3</sup>.

Immunohistochemistry is very important for establishing the diagnosis of MS since it can differentiate between myeloid and nonmyeloid cells. Monoclonal antibodies to MPO and lysozyme is positive in myeloid cells<sup>7</sup>. MPO is very often positive in the malignant cells of MS and it is a quick way for establishing the diagnosis and ruling out other tumors. CD68-KP1 is also a useful marker. It is the most commonly expressed marker followed by MPO<sup>8-9</sup>. In our case, the tumor tissue grew in the cervical lymph node, and there was neither a previous history of AML nor evidence of a myeloid malignancy in the patient bone marrow. This kind of case is very easily misdiagnosed with lymphoma, but the immunohistochemistry showed that the MPO and CD68 positive cells consisted of most of the tissue mass in the lymph node which indicated this was a case of MS instead of a case of lymphoma.

There is no consensus on the treatment of either isolated MS or MS presenting concomitantly with AML. Randomized prospective trials are lacking. The current recommended treatment regimen in patients presenting with MS is conventional AML-type chemotherapeutic protocols<sup>10</sup>. But according to the report of Yamauchi and Yasuda cytarabine may be an important agent in the treatment of MS since they found a prolonged DFS in the patients treated with cytarabine contained regimen compare with those who were initially misdiagnosed and treated with agents used to treat lymphoma, sarcoma or multiple myeloma<sup>11</sup>. Due to a higher rate of progression to AML in isolated MS patients, localized treatment like radiotherapy and surgical treatment is not recommended<sup>3</sup>. In our case, a regimen of DA for AML was given to this patient for the induction treatment, and the tumor mass shrunk greatly after the induction therapy which indicated that the regimen was effective to the patient. But during the bone marrow suppression the use of G-CSF greatly increased the WBC in peripheral blood, and the immature myeloid cells including certain ratio of blast appear in the peripheral blood. Since MS can easily develop into AML we examined the patient's bone marrow, but the bone marrow harvested at the same time showed even lower percentage of blast than in the peripheral blood. This indicated that the blasts and immature myeloid cells were not due to the mobilization function of the G-CSF from bone marrow. Bringing the fact that after the use of G-CSF, the WBC increased greatly along with the increasing of tumor size, we assumed that the immature myeloid cells may be mobilized by G-CSF from the tumor mass. This made us to think that the G-CSF may stimulate the progression of the tumor cells of MS. So according to this case we strongly recommend that G-CSF should not be used in MS unless there is a severe infection during the neutropenia state, in

order to avoid the tumor cells being driven into cell cycle and growing more faster than before the treatment.

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