

## Case Report

# Monoblastic sarcoma- a rare variant of myeloid sarcoma presenting as a supraorbital mass in an infant

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## Introduction

Monoblastic sarcoma is a rare variant of myeloid sarcoma comprising a population of more than 80% monoblasts (1). Presentation is usually in adulthood (2). This is a rare case of monoblastic sarcoma presenting as a supraorbital mass in an infant.

## Case report

### Clinical history

A previously healthy seven-month-old boy presented with a left supra orbital swelling of two months duration. Examination revealed a non tender, firm supra orbital mass measuring 6cm in maximum diameter. Full blood count and blood picture were normal. Magnetic resonance imaging scan of brain and orbital region revealed a well defined, large soft tissue mass in the temporal region with intracranial and orbital extension. There was no extension to temporal lobe or optic nerve.

### Pathological features

Fine needle aspiration (FNA) of the mass revealed cellular smears comprising discohesive, singly scattered monomorphic cells (Figure 1A). The cells had minimally

pleomorphic, round to oval nuclei with fine chromatin, occasional prominent nucleoli and scanty basophilic cytoplasm (Figures 1B and 1C). Occasional apoptotic bodies and abundant mitoses were seen. Based on these findings a diagnosis of a round blue cell tumour was made. The lesion was subsequently excised. Intraoperative crush smears concurred with the FNA diagnosis of a round blue cell tumour (Figure 1D).

Gross examination of the excised specimen revealed a firm whitish lesion measuring 55x40x35mm within fibro-fatty tissue. Microscopic evaluation of the tumour showed sheets, clusters and singly scattered cells (Figure 1E). The tumour showed areas with starry sky (Figure 1F), nested/alveolar (Figure 1G) and single file patterns (Figure 1H). The cytological features were similar to that seen in the FNA with small to medium sized minimally pleomorphic cells with hyperchromatic nuclei and scanty eosinophilic cytoplasm. Nucleoli were inconspicuous in most cells. Scattered cells with eccentric eosinophilic cytoplasm were seen. Mitoses were abundant with atypical figures. The tumour cells infiltrated the adjacent skeletal muscle fibers and bone.

The tumour cells showed strong diffuse cell membrane positivity for leukocyte common antigen (LCA) (Figure 1J) and were negative

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for CD99, desmin, MyoD1 and synaptophysin excluding Ewing sarcoma, rhabdomyosarcoma and neuroblastoma. The tumour cells were negative for CD3, CD20, CD19 and nTdT. The Ki67 proliferative index was >90% (Figure 1K) indicating a high grade, aggressive lesion but excluding a lymphoblastic lymphoma, Burkitt lymphoma and small cell variant of diffuse large B cell lymphoma. Since the tumour

showed a LCA only phenotype, myeloid sarcoma and blastic-natural killer (NK) cell lymphoma were considered in the differential diagnosis. The tumour cells were negative for CD2, CD5, CD7, CD4 and CD34. There was strong membrane positivity for CD56 (Figure 1L) and granular cytoplasmic positivity for CD68 (Figure 1M). Myeloperoxidase (MPO)

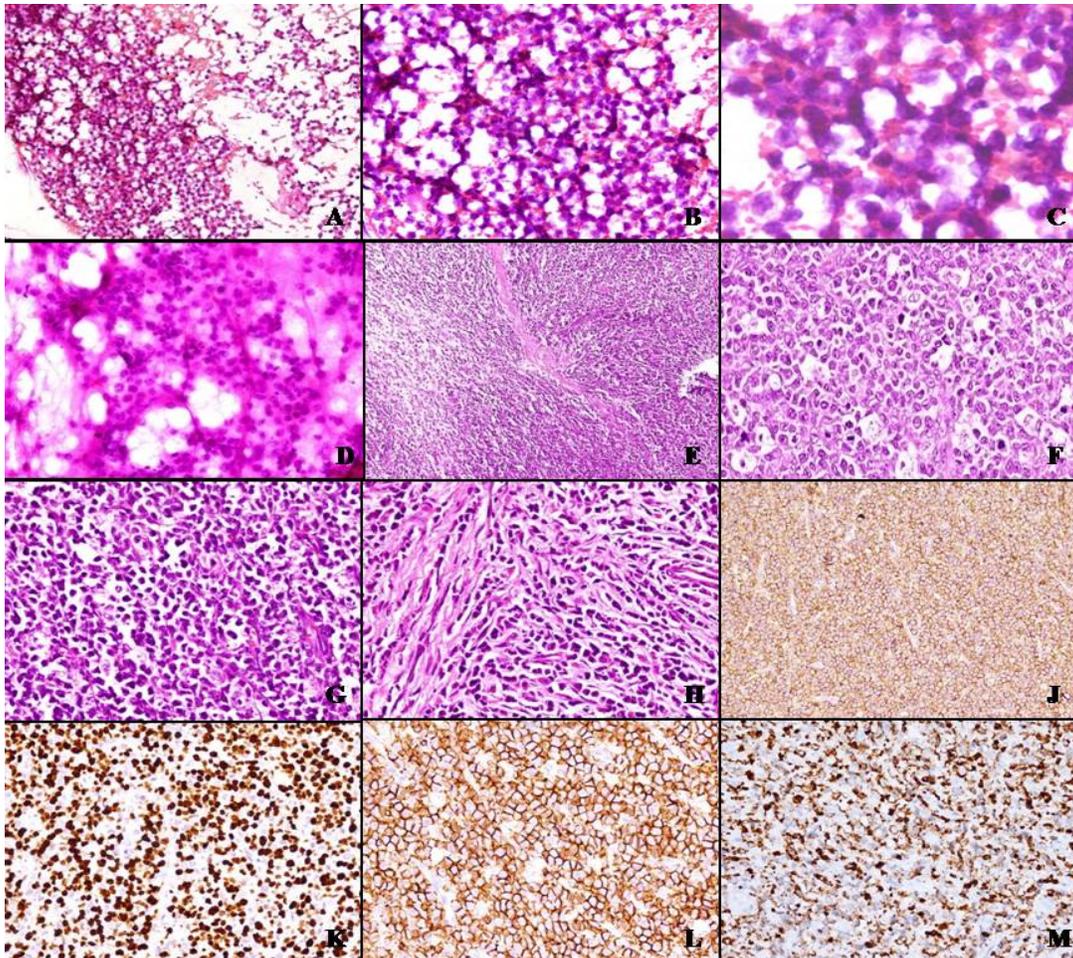


Figure 1. A) FNAC: cellular smears comprising discohesive, singly scattered monomorphic cells (H&E x 100) B)FNAC: cells contained mildly pleomorphic round to oval nuclei with fine chromatin and scanty eosinophilic cytoplasm(H&E x 200) C)FNAC: medium sized cells, with finely granular chromatin, occasional prominent nucleoli and scanty cytoplasm (H&E x 400) D) Crush smear showing similar cytological features as in FNAC (H&E x 100) E) Diffusely infiltrating round blue cells (H&E x 100) F) Starry sky appearance (H&E x 400) G) Alveolar pattern (H&E x 400) H) A single file pattern(H&E x 400) J) Diffuse cell membrane positivity for LCA (LCA x 200) K) High Ki67 proliferative index of around 90% (Ki67 x 200) L) Diffuse cell membrane positivity for CD 56 (CD56 x 200) M) Granular cytoplasmic positivity for CD 68(CD68 x 200)

was negative. CD68 and CD56 positivity with negativity for CD34 and MPO is usually seen in monoblastic sarcoma which is a rare variant of myeloid sarcoma (1). Hence, this tumour was diagnosed as a monoblastic sarcoma.

### Discussion

Myeloid sarcoma is defined as a tumour mass composed of myeloid blasts with or without maturation occurring at an anatomical site other than the bone marrow (3). Myeloid sarcoma can occur *denovo* as was seen in our patient or concurrently with AML, myeloproliferative disorders or myelodysplastic syndrome (4).

There is a male preponderance and a predilection for last decades of the life. Although the presentation at 7 months was unusual in this case, myeloid sarcoma has been reported from one month to 89 years (4). It commonly involves the subperiosteal bone structures of the skull, paranasal sinuses, sternum, ribs, vertebrae and pelvis. However, skin and lymph nodes can also be involved (1).

The cells of myeloid sarcoma are medium sized, have granular cytoplasm and are positive for CD43, CD45, lysozyme, MPO, CD117, CD99, CD68, CD56, CD34 and CD4 (5). CD43 and lysozyme have been shown to be the most sensitive markers (1).

Differentiating myeloid sarcoma from blastic NK cell lymphoma can be difficult. The young age of the patient and clinical presentation were not in favour of a blastic NK cell lymphoma which usually occurs in the older age group and is associated with skin manifestations(6). The lack of cytoplasmic granularity was unusual for myeloid sarcoma. Mitotic figures are generally not abundant in blastic NK cell lymphoma but were numerous in this case. Myeloid sarcoma and blastic NK cell lymphoma both comprise medium sized cells that express CD68 and CD56 (5-7). Consistent negativity for Myeloperoxidase (MPO) and monocytic butyrate esterase in blastic NK cell lymphoma, is important in

differentiating it from myeloid sarcoma (6). In this case the tumour cells were negative for CD2, CD5, CD7, MPO and CD34. Furthermore, CD4, which is usually positive in blastic NK cell lymphoma, was negative in the present case (6). Usually CD34 and MPO are positive in myeloid sarcoma. However, CD68 and CD56 positivity with negativity for CD34 and MPO is usually seen in monoblastic sarcoma, a rare entity of myeloid sarcoma (1,3). In monoblastic sarcoma the cells are weakly positive or negative for CD4 and Ki67 proliferative index is 50-95% (8,9) as seen in this case.

Monoblastic sarcoma is a rare variant of myeloid sarcoma accounting for 3-20% of myeloid sarcoma (8). It is composed of monoblasts which comprise >80% of tumour cells. Monoblastic sarcoma may occur at any age, but it is more common in young individuals than myeloid sarcoma (1). Like myeloid sarcoma, monoblastic sarcoma can occur *denovo* or concurrently with AML, myeloproliferative disorders or myelodysplastic syndrome. *De novo* myeloid sarcoma is often associated with monoblastic differentiation (1). Considering the young age of this patient it is important to exclude coexisting AML. However, the blood picture of this patient did not reveal evidence of a leukaemia. Unfortunately he succumbed before a bone marrow biopsy could be performed.

Myeloid sarcomas, especially in the paediatric age group show t(8;21) (q22;q22) (3). However, studies have shown that *de novo* myeloid sarcomas often show normal karyotype (1, 2). Performing cytogenetic examination for myeloid sarcoma can be difficult if there is no bone marrow or peripheral blood involvement and is therefore not recommended as a routine procedure especially for *de novo* cases (2). Genetic studies were not performed in this case.

Studies on the effects of clinical presentation (isolated myeloid sarcoma vs myeloid sarcoma concurrent or following

AML) on prognosis have been inconsistent, with some showing better outcomes in isolated myeloid sarcoma and others showing no difference in outcome between the two groups (4,9). The current treatment recommendation for isolated myeloid sarcoma and myeloid sarcoma occurring in AML patients is AML-type chemotherapy (9). Haematopoietic stem cell transplantation has also been shown to be associated with better overall survival in patients presenting with myeloid sarcoma with or without concomitant AML (10). Early diagnosis of monoblastic sarcoma is vital since early induction of chemotherapy helps to prevent the spread of the disease to other organs and improves survival (8). However, long term success rates of therapy are very low, and the mortality associated with therapy is high (2). This baby died two months after the surgery.

In conclusion, the very young age of our patient and presentation as a supra orbital soft tissue mass with intracranial extension was unusual for monoblastic sarcoma. The tumour comprised medium sized cells with moderate to scanty cytoplasm, occasional prominent nucleoli and inconspicuous cytoplasmic granularity making it difficult to differentiate from other round cell tumours. The LCA only immune profile led us to consider myeloid sarcoma in the differential diagnosis and guided towards reaching the diagnosis of the rare monoblastic sarcoma, a variant of myeloid sarcoma.

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