Mediterranean BioMedical Journals
International Journal of Medicine and Surgery
2017, Volume 4, Regular Issue, ID 169
DOI: 10.15342/ijms.v4ir.169

CASE REPORT

MYELOID SARCOMA, A RARITY OF THE HEAD AND NECK REGION - A CASE REPORT

Dr Krishnamoorthy Madhusudhan [1], MBBS; Dr Bee Sun Lee [2], MRCP; Dr Sellymiah binti Adzman [3], M.Med; Dr Shahrul bin Hitam [1], M.Med

[1] Department of Ear, Nose, Throat and Head &Neck Surgery, Ampang Hospital, Selangor, Malaysia.
[2] Department of Hematology, Ampang Hospital, Selangor, Malaysia.
[3] Department of Pathology, Serdang Hospital, Selangor, Malaysia.

ABSTRACT
Myeloid sarcoma is a rare disease, where a mass (tumour) of either myeloblasts or immature myeloid cells conglomerate in extramedullary anatomic sites. It may arise de-novo or it may present in association with acute myeloid leukemia. It can also occur in patients with myelodysplastic syndromes, myeloproliferative disorders, and as blast transformation in myeloproliferative neoplasia. Myeloid sarcoma of the head and neck area can pose a diagnostic challenge because of the low frequency of occurrence, and the vast diversity of tumours occurring from multiple lineages in this anatomic region. This can lead to a broad spectrum of various differential diagnoses. Therefore, a high index of suspicion is required when dealing with a possible case of myeloid sarcoma, as it is a time sensitive diagnosis with various diagnostic dilemmas. Here, we report an example, a case of tonsillar myeloid sarcoma. We wish to highlight the investigations that helped us procure this diagnosis in a timely manner.

KEY WORDS: Myeloid sarcoma, full blood picture, diagnosis, tonsils, acute myeloid leukemia.

Corresponding author :
Dr Krishnamoorthy Madhusudhan, ENT Department, Hospital Ampang, Jalan Mewah Utara, Pandan Mewah, 68000 Ampang, Selangor, Malaysia.
Email : kmadhu_87@yahoo.com

INTRODUCTION
Introduction: Myeloid sarcoma (MS) is a neoplasm of immature granulocytes, monocytes, or both, involving any extramedullary site [1]. The term MS has replaced the term granulocytic sarcoma and chloroma which were used in the past [2]. Some common extramedullary sites involved are skin, soft tissues, lymph nodes and the gastrointestinal tract [3]. However, in only 16% of the cases the tumor was found in the head and neck region [2]. There is a predilection for males with the median age of 56 years. MS can be the initial presentation of acute myeloid leukemia or it can arise de-novo; on its own [3]. MS usually occurs in the pediatric population, and rarely occurs in adults [4]. Besides simple blood parameters, MS of head and neck region requires the aid of immunohistochemical staining as a definite diagnostic tool [5].

CASE REPORT
A 73 years old male, was referred to our clinic, with a one-month history of dysphagia and noisy breathing. Initial physical examination revealed audible stertor with bilateral grade-3 tonsillar hypertrophy and multiple shotty cervical lymph nodes. Fibre-optic flexible nasoendoscopy was done, and it revealed hypertrophy of pharyngeal lymphoid tissue at bilateral peritubal areas, palatine tonsils and base of tongue. Tissue biopsies of both palatine tonsils were taken and sent for histopathological examination. Laboratory parameters on this initial presentation showed haemoglobin of 8.3g/dl, hematocrit of 25.2% and an elevated WBC of 91.6K/uL with a

66.2% monocyte preponderance. At this point of time, platelet count was 68K/uL. Patient was advised for admission and for further workup of his deranged hematological parameters, however he refused. He presented again to the hospital after four weeks with complaints of body weakness and lethargy. At this juncture, patient was already in tumor lysis syndrome, and he was now agreeable for admission. During his stay in the ward, a thorough list of investigations was done. Peripheral blood film (Figure 1.) showed circulating myeloblasts and monoblasts.

A bone marrow aspirate and trephine was also performed (Figure 2.) confirming the diagnosis of acute myeloid leukemia.

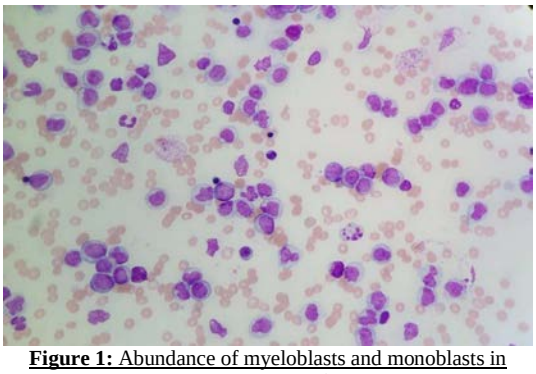

Figure 1: Abundance of myeloblasts and monoblasts in peripheral blood film.

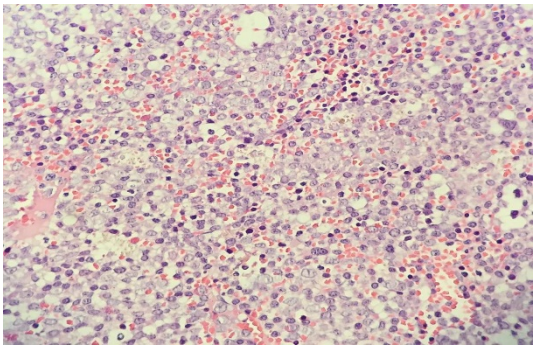

Figure 2: Bone marrow aspirate and trephine showing markedly reduced trilineage hematopoiesis replaced by predominantly blasts population of cells

The initial biopsies taken from both tonsils which were sent for histopathological examination showed loss of normal lymphoid architecture. There was marked infiltration of irregular clusters and singly distributed large atypical mononuclear cells mainly in the interfollicular areas. These cells were either having large irregular eccentric nuclei or large folded vesicular nuclei (Figure 3.) with fine chromatin or conspicuous to prominent nucleoli.

A series of immunohistochemical stains showed that these malignant cells were positive for myeloperoxidase (Figure 4.).

These cells were also positive for CD31, CD 33, KP1 and CD34 (in 5% of the cells); and negative for CD20, CD3, CD123, and CD 117. Upon establishing the diagnosis with a multidisciplinary approach, the patient and his relatives were counselled regarding treatment options and risks of disease propagation. The patient and family members opted for treatment with palliative chemotherapy, bearing in mind his advanced age. He succumbed shortly after commencement of treatment due to the rapid progression of the disease.

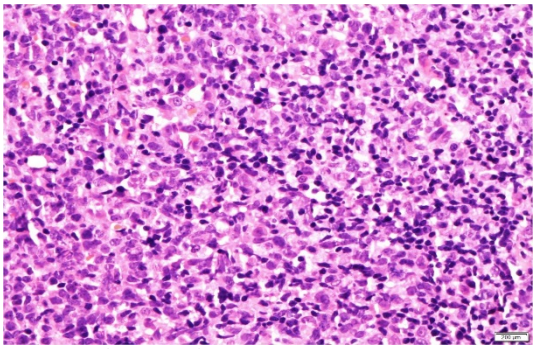

Figure 3: Infiltration of neoplastic cells under hemotoxylin and eosin examination with microscope(x100 magnification) showing irregular eccentric nuclei.

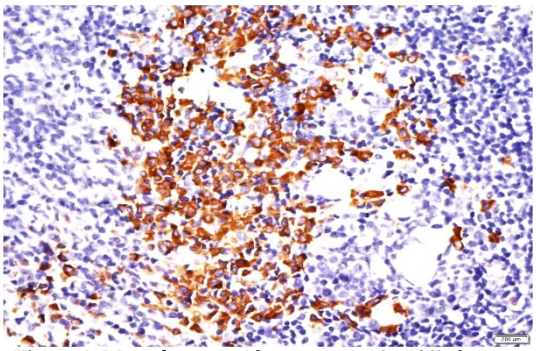

Figure 4: Magnification under microsope (x 100) showing malignant cells positive for myeloperoxidase.

DISCUSSION

Myeloid sarcoma (MS) was previously referred to as granulocytic sarcoma and also by the name chloroma meaning ‘green tumor’ [3] .King in 1853 coined the term chloroma due to its frequent green color[4].This macroscopic green appearance is caused by the production of myeloperoxidase in many of these tumors[3]. MS is a tumoral mass of either myeloblasts or immature myeloid cells in extramedullary sites. Besides the common locations mentioned above, MS may involve any organ / system in the body [5]. In this case report, the tonsils were involved which is a rarity [6]. Diagnosing myeloid sarcoma of the head and neck region poses a big challenge, because of the low frequency of occurrence and the potential for almost any lineage of tumour to occur in this region. MS can be mistaken on histology for non-Hodgkin’s lymphoma, small round cell tumors such as neuroblastoma or undifferentiated carcinoma [4]. As such, a high degree of suspicion is needed when dealing with a probable case of myeloid sarcoma [5]. The clinical diagnosis of MS is no doubt difficult, and definitive diagnosis warrants the use of immunohistochemical analyses [2]. An example of a specific immunohistochemical stain, which is a useful tool in clinching the diagnosis of myeloid sarcoma is myeloperoxidase[3]. Imaging studies like Magnetic Resonance Imaging (MRI) scans of MS exhibit low signal intensity on T2-weighted images, which helps to rule out inflammatory lesions [2]. However, it is worth taking note that a simple and relatively fast full blood count, is of paramount importance to narrow down the diagnostic process during the crucial initial stages of presentation. The prognosis of patients with MS is poor, and related to the clinical course of AML. The older the age, the more significant the adverse prognostic factor. [2]

CONCLUSION

Detailed immunohistochemical analyses is required for a definitive diagnosis of myeloid sarcoma. Nonetheless, a full blood count is an indispensable initial investigation which can help in guiding the crucial initial stages of diagnostics. An early diagnosis will save valuable and precious time; thus, treatment can be commenced immediately.

AUTHORS' CONTRIBUTIONS

The participation of each author corresponds to the criteria of authorship and contributorship emphasized in the Recommendations for the Conduct, Reporting, Editing, and Publication of Scholarly work in Medical Journals of the International Committee of Medical Journal Editors. Indeed, all the authors have actively participated in the redaction, the revision of the manuscript and provided approval for this final revised version.

PATIENT CONSENT

Written informed consent was obtained from the patient for publication of this case report.

COMPETING INTERESTS

The authors declare no competing interests.

REFERENCES

[1] Alexiev, B.A., Wang, W., Ning, Y., Chumsri, S., Gojo, I., Rodgers, W.H., Stass, S.A. and Zhao, X.F., 2007. Myeloid sarcomas: a histologic, immunohistochemical, and cytogenetic study. Diagnostic pathology, 2(1), p.42.

[2] Papamanthos, M.K., Kolokotronis, A.E., Skulakis, H.E., Fericean, A.M.A., Zorba, M.T. and Matiakis, A.T., 2010. Acute myeloid leukaemia diagnosed by intra-oral myeloid sarcoma. A case report. Head and neck pathology, 4(2), pp.132-135.

[3] Kaygusuz, G., Kankaya, D., Ekinci, C., Topçuoğlu, P. and Kuzu, I., 2015. Myeloid sarcomas: a clinicopathologic study of 20 cases. Turkish Journal of Hematology, 32(1), p.35.

[4] Payne, C., Olivero, W.C., Wang, B., Moon, S.J., Farahvar, A., Chen, E. and Wang, H., 2014. Myeloid sarcoma: a rare case of an orbital mass mimicking orbital pseudotumor requiring neurosurgical intervention. Case reports in neurological medicine, 2014.

[5] Zhou, J., Bell, D. and Medeiros, L.J., 2013. Myeloid sarcoma of the head and neck region. Archives of Pathology and Laboratory Medicine, 137(11), pp.1560-1568.

[6] Krause, J.R., 1979. Granulocytic sarcoma preceding acute leukemia. Cancer, 44(3), pp.1017-21.