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Case Report Open Access

Myeloid Sarcoma of Orbit in a Case of Acute Myeloid Leukemia

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Abstract

Background: Extramedullary proliferation of cells of myeloid origin is termed as myeloid sarcoma. About 3-8% cases of AML present with myeloid sarcoma.

Methods: A 10 year old child with proptosis was investigated and found to have mass in left supralateral region of left orbit. Enucleation of the eye was done and sent for histopathology along with the mass.

Results: Histopathology showed sheets of round tumor cells positive for CD 43, LCA and C- Kit. Peripheral smear examination showed >20% blasts.

Conclusion: A diagnosis of granulocytic sarcoma in a case of acute myelogenous leukemia was given.

Keywords: Acute myeloid leukemia; Myeloid sarcoma; Orbital involvement

Introduction

Extramedullary proliferation of cells of myeloid origin is termed as myeloid sarcoma. This tumor was first described by Allen Burns [1-3] in 1811. It was green in color, hence was called as chloroma, however, not all of these tumors are usually green [2,3]. Rappaport [4] in 1966 renamed this tumor as myeloid sarcoma [2]. The terminology presently in use is granulocytic sarcoma as by WHO nomenclature. This sarcoma was found to be associated with acute leukemia in 1893 [5]. But it can precede or follow or occur concurrently with myeloid leukaemia which can be acute or chronic. It can even be seen in association with other myeloproliferative disorders and myelodysplastic syndromes. A mean interval of 10.5 months has been observed for the development of leukaemia in non leukemic cases from the time of diagnosis [5]. The sites most commonly involved by this tumor include bones especially skull, orbit, paranasal sinuses, spine, ribs, sacrum and sternum [6]. It can also involve the lymph nodes, skin, breast, ovary, brain, gastrointestinal tract and kidney [3,6,7]. About 3-8% cases of AML present with myeloid sarcoma [2,3,8] especially AML M2 [6]. Other variants are also known to cause extramedullary manifestations but to a lesser extent [6]. In this report of our case, a male child presented with proptosis without any prior history of leukaemia and was diagnosed as a case AML later during the haematological investigations on a peripheral blood smear.

Case Report

A 4 year old male child came to our outpatient department with protrusion of the left eye along with redness and watering since 15 days. One week later, he experienced pain in left eye. However, there was no loss of vision. Right eye was normal. The protrusion was gradually progressive and the patient was not able to close the eye at present. He was irritable and non cooperative. Ophthalmic examination revealed pupils reacting to light, spontaneous movement decreased on left side in all directions, more on lateral side. However, fundus examination could not be done as patient was non cooperative. Motor reflexes were normal. CT scan of the orbit showed an isodense soft tissue mass of 37x24 mm along the superolateral aspect of the left orbit (Figure 1). The retrobulbar tissue, bony walls, muscle and optic nerves were unremarkable. Tumor excision along with the left eye enucleation was done in view of exposure keratitis and shrunken eyeball. Tumor was hard, well circumscribed. Cut section was greyish white and firm to hard in consistency (Figure 2). Imprint or squash smear on staining with Leishman stain revealed isolated, non cohesive, non granular cells, with round to oval nucleus with fine, dispersed chromatin, distinct nuclear membrane, small eosinophilic nucleolus and rim of pale cytoplasm. Few cells showed cytoplasmic eosinophilic granules or nuclear infoldings. Histopathology revealed an infiltrating tumor comprised of sheets of round cells of intermediate to large size with scant cytoplasm having pleomorphic nuclei with conspicuous nucleoli with irregular nuclear membranes and convolutions. Eosinophilic granules were evident in the cytoplasm. Mitotic activity was brisk as were apoptotic bodies (Figure 3). Peripheral blood smear examination of the patient showed more than 20% myeloblasts with Auer rods (Figure 4). These cells were strongly positive for myeloperoxidase (Figure 5). Immunohistochemical analysis revealed strong LCA, CD 43 and C Kit positivity (Figure 6). CD20, CD3, mic 2 and Tdt were negative. However, there was no tumorous involvement of eyeball. Flow

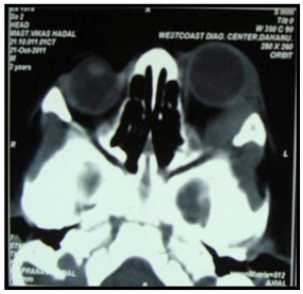

Figure 1: Axial CECT showing tumor occupying the lateral orbital wall and causing proptosis.

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cytometry confirmed the myeloid origin of tumor cells. Literature shows few cases where cytogenetic analysis of AML patients with myeloid sarcoma is done. It is seen that t (8;21) associated with AML type 2 are more commonly with granulocytic sarcoma than others. The incidence of 4.5-38% is represented with t (8;21) (q22;q22) with development of granulocytic sarcoma [7]. Also 11q23/MLL rearrangements and occurrence of granulocytic sarcoma in body cavities has been suggested [6]. However, cytogenetic studies could not be done in our case. A diagnosis of acute myeloid leukaemia with myeloid sarcoma was given. The patient was referred for chemotherapy.

Discussion

Myeloid sarcoma, previously known as granulocytic sarcoma [9] is predominantly a disease of childhood or young adults [10,11] though a wide age range from infancy to 61 years of age is observed with mean of 7 years. Bone and soft tissues involvement is predominant [10]. This is the most common presentation as highlighted by various studies done previously [2,3,10]. Granulocytic sarcoma has been reported in association of AML with an incidence of 2.5-8%. Subperiosteal bone involvement in vertebrae, sternum, orbit and cranium is observed with orbital involvement in about 3% [3,6]. Lateral wall of orbit is more commonly affected than medial wall. However, the clinical presentation

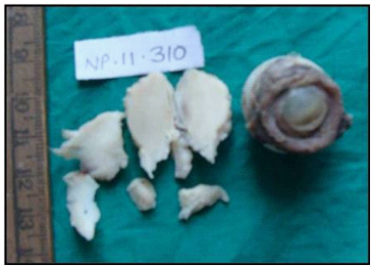

Figure 2: The tumor was firm, greyish white in color. Enucleated eyeball was also received.

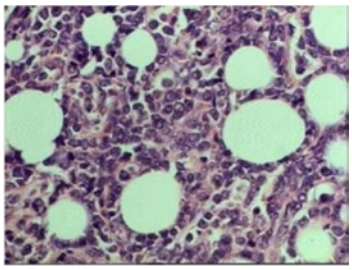

Figure 3: Sheets of tumor cells having large, round to convoluted nucleus were present (H&E, 20x).

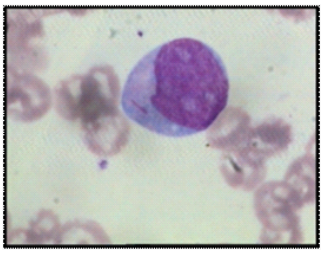

Figure 4: Peripheral blood smear showing blast cells with Auer rods (Leishman stain, 100x).

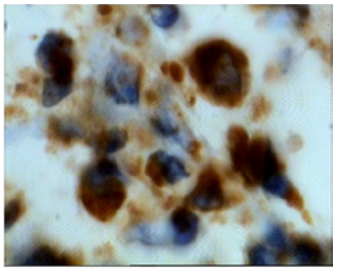

Figure 5: Strong myeloperoxidase staining in myeloblasts.

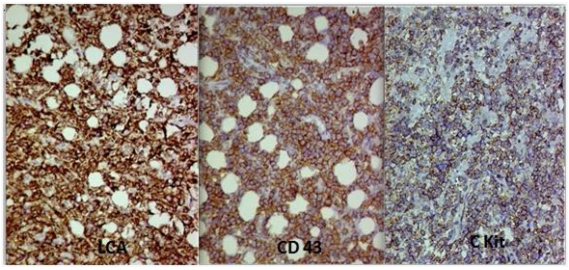

Figure 6: Immunohistochemistry showed strong LCA, CD43 and c kit positivity.

of the patient may be variable. Unilateral or bilateral proptosis is the commonest sign of presentation with orbital involvement [10]. In our case it was unilateral presentation. Males are commonly affected than females [6]. Involvement can occur either in a known patient of AML or else AML is detected later or during the course of the disease. In our case, the patient presented with exophthalmos and was diagnosed with AML during the routine examination of peripheral blood smear. The diagnosis is however challenging when the patient presents without any prior history of haematological malignancy. The differential diagnosis to be considered may include lymphomas, neuroblastomas, rhabdomyosarcomas and other round cell tumors [2,12]. Radiological investigations including CT and MRI are helpful in localisation of the tumor. However, the most confirmatory of the diagnosis is imprint smear and histopathology. The importance of peripheral smear examination has been stressed upon by many authors [3,12]. Imprint or squash smear aid in identification of the cells of hematopoietic origin with some cells showing eosinophilic granules and nuclear infoldings. Leukemic smear along with ancillary investigations like cytochemistry and flow cytometry aid in the diagnosis in most of the cases. Myeloperoxidase stain helps in identifying origin of the cells along with Sudan Black B. Histopathology of the tumor reveals sheets of round cells. Myeloid cell markers like LCA, CD 43 and C kit are expressed supporting the diagnosis of granulocytic sarcoma as in our case. Cytogenetic studies unfortunately could not be done in our case. We were unable to get material for cytogenetic studies.

Conclusion

Granulocytic sarcoma is an uncommon malignant neoplasm associated with myeloid leukaemia having poorer prognosis than myeloid leukaemia only. Therefore, careful history and accurate investigations accompanied by high index of suspicion are needed for proper and timely management of the patient.

References

1. Burns A (1811) Observation on the surgical anatomy of the head and neck. Thomas Royce & Co, Edinburg, United Kingdom.

2. Stockl FA, Dolmetsch AM, Saornil MA, Font RL, Burnier MN Jr (1997) Orbital granulocytic sarcoma. Br J Ophthalmol 81: 1084-1088.
3. Murthy R, Vemuganti GK, Honavar SG, Naik M, Reddy V (2009) Extramedullary leukemia in children presenting with proptosis. J Hematol Oncol 2: 4.
4. Rappaport H (1966) Tumours of the hematopoietic system. Atlas of tumour pathology. Armed Forces Institute of Pathology, USA.
5. Neiman RS, Barcos M, Berard C, Bonner H, Mann R, et al. (1981) Granulocytic sarcoma: a clinicopathologic study of 61 biopsied cases. Cancer 48: 1426-1437.
6. Alkatan H, Chaudhry I (2008) Myeloid sarcoma of the orbit. Ann Saudi Med 28: 461-465.
7. Aalia R Sufi, Tejit Singh (2011) Granulocytic Sarcoma as the First Sign of Acute Leukemia in Childhood. Online J Health Allied Scs 10: 9.
8. Morel F, Herry A, Le Bris MJ, Le Calvez G, Marion V, et al. (2002) Isolated granulocytic sarcoma followed by acute myelogenous leukemia type FAB-M2 associated with inversion 16 and trisomies 9 and 22. Leukemia 16: 2458-2459.
9. Vardiman JW, Thiele J, Arber DA, Brunning RD, Borowitz MJ, et al. (2009) The 2008 revision of the World Health Organization (WHO) classification of myeloid neoplasms and acute leukemia: rationale and important changes. Blood 114: 937-951.
10. Cavdar AO, Arcasoy A, Babacan E, Gozdasoglu S, Topuz U, et al. (1978) Ocular granulocytic sarcoma (chloroma) with acute myelomonocytic leukemia in Turkish children. Cancer 41: 1606-1609.
11. Lee SG, Park TS, Cheong JW, Yang WI, Song J, et al. (2008) Preceding orbital granulocytic sarcoma in an adult patient with acute myelogenous leukemia with t(8;21): a case study and review of literature. Cancer Genet Cytogenet 185: 51-54.
12. Anita Sethi, Supriyo Ghose, Sumeet Gujral, Paresh Jain, Rajive Kumar (2001) Childhood proptosis: The invaluable, though often overlooked peripheral blood smear. Indian J Ophthalmol 49: 121-123.