CLINICAL REPORT

Invasive Fungal Infection in Acute Myeloid Leukemia Associated with Myeloid Sarcoma of the Sinonasal Cavity: A Rare Case

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Abstract Myeloid sarcoma is a rare malignant extramedullary neoplasm of myeloid precursor cells. The majority of these cases occur in patients with known leukemia or those who eventually develop the disease. It can occur in various sites but sinonasal cavity involvement along with invasive fungal infection is exceedingly rare.

Keywords Myeloid sarcoma · Sinonasal cavity · Acute myeloid leukemia · Aspergillus fungus

Introduction

Myeloid sarcoma (MS) is a tumour mass of myeloblasts with or without maturation occurring outside the bone marrow. These rare extramedullary myeloid tumours may occur *denovo*, or coexist with or follow the presentation of Acute Myeloid Leukemia (AML). MS can also arise as blastic transformation of an underlying chronic myeloproliferative disease or Myelodysplastic Syndrome [1].

The chemotherapeutic agents used in these haematological malignancies produce neutropenia, thus predisposing to invasive fungal infections [2].

Co-existence of MS and invasive fungal sinusitis in the sinonasal cavity is exceedingly rare and can present a diagnostic challenge, as either one could be masked by the other due to their similar clinical features. To the best of our knowledge, only one such case has been reported in the literature [3].

We report a case of concurrent sinonasal myeloid sarcoma and acute fulminant invasive fungal sinusitis in a patient of acute myeloid leukaemia as an aftermath of chemotherapy.

Case Report

A 58 year old lady was diagnosed with Acute myeloid Leukemia (M0 according to French-American-British classification) one month back. She had completed first cycle of chemotherapy, when she presented with a blackish discoloration of tip of the nose with no symptoms of nasal obstruction. Nasal endoscopy revealed a completely eroded right middle turbinate with debris in the right nasal cavity. Computed Tomography of paranasal sinuses (PNS) showed a soft tissue density mass in maxillary, frontal, ethmoid sinuses and nasal cavity on the right side with erosion of medial and posterior-lateral wall of right maxillary sinus (Fig. 1). Tissue debrided from right nasal cavity and right maxillary sinus concretions revealed a bimodal histology of extensive necrosis with fungal hyphae and sheets of immature blasts. Hyphae of *Aspergillus* was confirmed by Periodic Acid Stain (Fig. 2) and later on by growth (*Aspergillus flavus*) in Sabourad's dextrose agar culture medium. Blast cells were positive for IHC marker CD-34.

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Fig. 1 CT PNS (Paranasal Sinus) showing soft tissue density mass of right maxillary sinus and right nasal cavity with erosion of medial and posterio-lateral wall

Fig. 2 Colonies of PAS (Periodic Acid Schiff) positive septate acute angle branching fungal hyphae ($\times 400$)

Discussion

Myeloid sarcoma was first described by the British physician A. Burns in 1811. King coined the term chloroma in 1853 as these tumors often have a green tint due to the presence of myeloperoxidase (from the Greek “chloros” meaning green). Dock later established the relationship of MS with acute leukemia. Turk et al reported the first case of myelocytic leukemia associated with chloroma in 1903.

As 30% of these tumors can be white, gray or brown rather than green, the more correct term ‘granulocytic sarcoma’ was proposed by Rappaport in 1967.

Incidence of MS is 1–9% of patients with AML. It is more common in children and young adults with no difference between sexes [4].

Approximately 70% of MS occur in patients with a known myeloproliferative syndrome or acute non-lymphocytic leukemia, the other 30% have no underlying disease at the time of diagnosis (proleukemic phase). Ninety percent of these patients develop acute leukemia within a few months [5].

Pantanowitz et al. [6] in 2005 reported that MS could occur in any one of three clinical settings: (1) In patients who have a history of AML, during active disease or a recurrence; (2) in patients with chronic myeloproliferative disorders (MPD), who are at increased risk of blast transformation or (3) in patients with no history of hematologic disease, although it commonly predates the development of leukemia, often within one year.

While MS can be solitary or multifocal, most cases manifest as isolated single lesions [7]. The most common sites of extramedullary involvement of AML include the skin (leukemia cutis), lymph node, and bone, but other sites, including the female reproductive tract, breast, gut, and testis have been reported. In the head and neck region, MS accounts for 12–48% of the cases and orbits are commonly involved [8]. It is believed that MS begins in the bone marrow and reaches the subperiosteum through haversian channels and then spread to other parts of the body. Involvement of sinonasal cavity is exceedingly rare with only a few reported cases [3].

Patients with acute leukemia are at risk for invasive aspergillosis as a consequence of intensive chemotherapy, which produces prolonged neutropenia. The lung is the most common site of aspergillus infection. Invasive aspergillus rhinosinusitis is an important variant, since it may herald or be coincident with pulmonary disease and often indicates a poor prognosis [2]. The combined destructive effect of MS and invasive aspergillus infection may lead to rapid progression and death as in our case.

MS may be misdiagnosed since it can mimic malignant lymphoma or an undifferentiated carcinoma. Goteri et al. [9] emphasized a clinical diagnostic suspicion of myeloid sarcoma for accurate diagnosis whenever an extramedullary mass appears in a patient with a hematological disorder. An Immunohistochemical panel consisting of MPO, CD43, CD117, CD34, CD68, CD3, CD20 can successfully identify majority of MS. In addition, flow cytometry and cytogenetics help to improve the likelihood of correct definitive diagnosis.

Conclusion

Myeloid sarcoma of the head and neck region can pose diagnostic challenges because of the low frequency of occurrence of myeloid sarcoma and the potential for tumors of almost any lineage to occur in the head and neck [10]. Coexistence of sinonasal myeloid sarcoma and acute fulminant invasive fungal sinusitis poses an urgent diagnostic and management challenge as both are equally fatal and require different treatment.

Compliance with Ethical Standards

Conflicts of interest All authors declare that they have no conflict of interest.

Human and Animal Rights This article does not contain any studies with human participants or animals performed by any of the authors.

Informed Consent Informed consent for publication was obtained from the family member of the subject included in the case.

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