Granulocytic sarcoma of the lacrimal gland

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A 35-year old man presented with acute myeloid leukemia (AML-M2), with 45, X, -Y, t(8;21)(q22;q22) [8] and AML1-ETO fusion.[1] A molecular remission was obtained by 7+3 induction followed by high dose Ara-C and 5+2 consolidations x4. One year later, he presented with left eye proptosis and double vision. Magnetic resonance imaging studies confirmed left lacrimal gland swelling (Figure 1A and B) with no other extramedullary deposits. Visual acuity and fields, as well as fundoscopy and slit lamp examination were normal. An aspirate confirmed leukemia relapse, with concurrent marrow disease despite normal peripheral counts (Figure 1C to E). Staining for CD56 was negative. A lumbar puncture showed no morphological or molecular evidence of leukemia involvement. He was reinduced with idarubicin, ara-C and etoposide x2 and achieved complete resolution of marrow and extramedullary disease with full visual recovery. An allogeneic stem cell transplantation (SCT) was performed from his matched sibling and he remained in remission six months afterwards. Granulocytic sarcomas are rare complications of *de novo* AML, but are associated with t(8;21) and CD56 (neural cellular adhesion molecular: NCAM) expression.[2] Glandular tissues e.g. breast, urogenital tract, gut, as well as the skin and brain are often affected. Involvement of orbital adnexa is not uncommon, especially in young children,[3,4] and granulocytic sarcoma is represented in several ophthalmological case series of lacrimal neoplasms.[5,6] Interestingly, however, synchronous or metachronous spread to the other eye is rare. It should be noted that granulocytic sarcoma after chemotherapy is usually accompanied by marrow relapse, and systemic therapy is usually needed. With the presence of orbital disease, central nervous system prophylaxis may also be prudent. Secondly, despite the dramatic extramedullary relapse, the response to chemotherapy and ultimate prognosis is still good, especially for t(8;21) AML. Furthermore, visual acuity is usually maintained and additional radiotherapy, with its sight-threatening potential, is debatable. Finally, the higher risk of extramedullary failure of AML after SCT may call for careful post-SCT molecular monitoring and CNS prophylaxis in such patients with previous history of tissue involvement.[7]

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Figure 1. **A and B: Transverse and coronal sections of T2 weighted magnetic resonance imaging scan showing contrast enhancing mass occupying the superior lateral aspect of the orbit due to lacrimal gland infiltration. C to E: Needle aspiration showing cluster of blast cells, with strong expression of myeloperoxidase (D) and Sudan Black B (E) (x 400 magnification).**

References

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