Traumatic left shoulder fracture masking aggressive granuloblastic sarcoma in a CML patient

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A 39 year-old man was diagnosed with Philadelphia Positive Chronic Myelogenous Leukemia (CML) in chronic phase in 1989. He was treated with alpha-interferon at a median daily dose of 5 MU because of side effects (flu-like syndrome not controlled by the use of paracetamol) achieving a complete hematological remission but no cytogenetic response. Neither related nor unrelated matched donor was found. In 1997, because of resistence to treatment, alpha-interferon was stopped and changed to hydroxyurea. In April 2000 the patient in chronic phase was enrolled in a perspective study on the use of the newly introduced tyrosine kinase inhibitor imatinib. He received 400 mg/day p.o. thereafter mainteining his hematological response, but never achieving a cytogenetic response.

In June 2002, left shoulder pain ensued; no scheletal or soft tissue lesions were evident at a bone X ray study. In August 2002, the patient was involved in a car accident which resulted in traumatic left humerus fracture which did not require surgical repair.

Because of increasing left arm and shoulder edema suggestive of local thrombosis, sonographic and doppler studies were performed which were normal. In December 2002 MR imaging (Figure 1) of the same shoulder showed, along with signs of the recent traumatic fracture, the presence of a proximal humerus osteolitic lesion associated with extensive substitutive tissue which was biopsied and lead to a diagnosis of granuloblastic sarcoma (Figure 2, Figure 3). Bone marrow aspirate was still consistent with a diagnosis of CML chronic phase.

Imatinib dosage was increased up to 800 mg/day p.o. without response; therefore Cytosin-Arabinoside (500 mg/day i.v. for 4 days) and Dexamethasone (40 mg/day e.v. for 4 days) were administered. Aplasia was complicated by the development of fever and TC imaging highly suggestive of mycotic pulmonary infection.

Left shoulder lesion worsened (Figure 4) even during post-chemotherapy aplasia and therefore local radiotherapy was administered (21 Gy). The patient died in February 2003 with rapidly progressive local disease and mycotic pulmonary infection; however, bone marrow examination was still consistent with a diagnosis of CML chronic phase.

Conclusions

Granuloblastic sarcoma, originating in a bone fracture, is a rare event; this localization was resistant to every kind of therapy and responsible for death, despite the persistent chronic phase of bone marrow.

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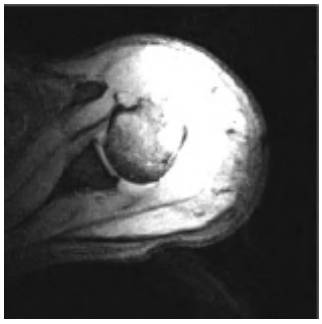

Figure 1. Proximal humerus osteolitic lesion associated with extensive substitutive tissue.

Figure 2. Immuno-hystological assay shows monotypic staining for CD34.

Figure 3. Monomorphic population of large cells with oval, round and distorted nuclei, small nucleoli, scarce cytoplasm. Mitotic figure is present (Haematoxylin and Eosin stain).