Serum angiotensin converting enzyme: a possible marker in Lennert’s lymphoma?

Data on raised activity of serum angiotensin converting enzyme (SACE) in patients with lymphoma are rare. High SACE activities were reported in Lennert’s lymphoma, and we report on two cases of Lennert’s lymphoma in which SACE was monitored during the course of the disease and in which SACE reflected the activity of the tumour.

A 24 year old man was admitted with sustained fever, malleoloraedema, and generalised weakness. Physical examination showed subicteric sclerae, hepatosplenomegaly, and enlarged peripheral lymph nodes; pancytopenia was noted. A computed tomogram of the abdomen showed enlarged lymph nodes around the aorta, the vena cava, and the arterial ilaeae. Bone and lymph node biopsy specimens confirmed the diagnosis of Lennert’s lymphoma.

The activity of SACE was 863 U/l (normal values 115–491 U/l). Conventional chemotherapy was started; the patient went to partial remission after one course; SACE dropped to 287 U/l. Because of long standing leucopenia, treatment was changed to a less aggressive chemotherapeutic combination. Increased hepatosplenomegaly, fever without, evidence of infection, and pancytopeny recurred. The SACE activity rose to 689 U/l, and the patient remained resistant to further chemotherapy.

A 21 year old man presented initially with renal colic, which an intravenous pyelogram showed to be due to an excluded right kidney. Because of coexisting splenomegaly, a computed tomography scan of the abdomen was performed and an extensive tumoral mass was found. The histological appearances of an abdominal lymph node and the spleen were first thought to be diagnostic of Hodgkin’s disease. The patient developed a mild hypercalcemia.

Combination chemotherapy with two courses of MOPP (mustine-oncovicillin, procarbazine, and procarbazine) and three of ABVD (adriamycin-bleomycin-vincristine-dacarbazine) was begun. After an interruption of three months an intraginal lymph node relapsed and an additional course of MOPP and ABVD was administered.

Because of anorexia, vomiting, fever, icterus, hypercalcemia and anaemia, the patient was referred to our hospital one and a half months later. Progression of abdominal lymphoma was noted on computed tomography and ultrasonography. Liver and bone biopsy specimens showed reactional histioctyosis and a haemophagocytic syndrome in association with malignant lymphoma, mostly closely resembling a T cell type. Lennert’s lymphoma was diagnosed. SACE activity was 1004 U/l. The patient received another course of chemotherapy. After some complications, the patient died of cardiac arrest. The SACE activity was 422 U/l.

Lennert’s lymphoma is a haematological malignancy characterised by the presence of a high content of epitheloid histocytes, T cells, and rare Reed-Sternberg-like cells. In a study of sarcoidosis, Gaucher’s disease, and other granulomatous disorders Lieberman et al found that three out of 11 patients with Lennert’s lymphoma had raised SACE activities.

Reported cases of histiocytic medullary reticulosis have been associated with increased SACE activity. Grönhagen-Riska et al assumed that the increase in SACE activity reflected the monocytic line having reached the macrophage or histioctytic stage. Deremee et al reported two patients with non-Hodgkin’s lymphoma who showed increased SACE activities. These authors suggested a direct role for SACE in the metabolism of vitamin D.

Lennert’s lymphoma is possibly associated with an increase in SACE activity due to epitheloid proliferation. This can be helpful in differentiating Lennert’s lymphoma from other malignancies. Tumour activity can also be evaluated by this marker during and after treatment.

References