Symptomatic trigeminal neuralgia as the first presentation of acute lymphoblastic leukemia

Sir,

It is not unusual for the cranial nerves to be affected early in the course of acute lymphoblastic leukemia (ALL) due to the meningeal infiltration. Here, we report a 19-year-old male with the symptomatic trigeminal neuralgia and right third, fourth and sixth cranial nerve palsy as initial presentation of high-risk T-cell ALL. To the best of our knowledge, symptomatic trigeminal neuralgia as the first presentation of ALL has not been reported earlier.

A 19-year-old Asian male presented with complaints of painful sharp electric shock-like transient spasms on the right hemi-face around the eye and cheek for last 2 months and diplopia for last 15 days. The painful spasms usually lasted for a few seconds to minutes and triggered by light touch and activities such as brushing teeth, talking, and blowing his nose. On examination, right third, fourth and sixth cranial nerve palsy was found. The rest of the general and neurological examination was normal, including trigeminal sensations. Magnetic resonance imaging (MRI) of the brain showed a right parasellar mass, which was hyperintense on T1, isointense on T2 and fluid attenuated inversion recovery sequences; and intensely contrast enhancing on T1 sequence with the gadolinium contrast [Figure 1]. Involvement of right cavernous sinus and encasement of ipsilateral internal carotid artery were also evident from the MRI. Peripheral blood picture and bone marrow examination revealed features consistent with ALL. Cerebrospinal fluid cytology revealed the presence of leukemic blasts. Thyroid function tests, antinuclear factor, serum immunoglobulins, immunoelectrophoresis, and antibodies to acetylcholine receptor were all within normal limits. Immunophenotyping showed cluster of differentiation (CD) 3 and CD5 positivity. In view of positivity for chromosomal translocation t (4; 11) (q21; q23), he was diagnosed as a case of high-risk T-cell ALL, presenting as a parasellar mass (leukemic deposit) with right sided symptomatic trigeminal neuralgia and third, fourth and sixth cranial nerve palsy. For trigeminal neuralgia, the patient was recommended carbamazepine, 800 mg daily. He was given chemotherapy as per ALL-Berlin-Frankfurt-Munster 95 protocol. At first remission, he received 18 gray/10 fractions therapeutic cranial irradiation, followed by 8 gray/4 fractions boost to the right parasellar mass with 3-dimensional conformal radiotherapy technique. After 3 months of treatment completion, there was partial improvement of diplopia and the trigeminal neuralgia was well-controlled.

Cranial nerve palsies as the first presentation of ALL, although uncommon, but have already been reported.[1-3] There have also been some reports of cranial nerve palsies as the first sign of relapse or recurrence in ALL patients.[4,5] Unless specifically treated, the neurological deficit is usually progressive in this situation. We believe that this is the first description of a patient with a systemic lymphoproliferative disorder, namely ALL, presenting with symptomatic trigeminal neuralgia. The cause of the right third, fourth and sixth cranial nerve palsies in our patient was most probably leukemic involvement of the right cavernous sinus, as was
shown by MRI. There was partial improvement of the motor functions of the right eye, even after 3 months of treatment completion. The probable causes might be late presentation with some irreversible ischaemic damage to the cranial nerves due to the pressure effect of the right parasellar leukemic deposit and delay in radiotherapy.

To conclude, symptomatic trigeminal neuralgia and cranial nerve palsies led to the diagnosis of ALL in this patient with minimal systemic clues.

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