Hallucinations Common in Pediatric Lupus

VITALS

Major Finding: Hallucinations are a common finding in children with NPSLE.

Data Source: An observational study of 53 children with juvenile SLE with neuropsychiatric manifestations.

Disclosures: Dr. Lim reported no financial conflicts of interest.

BY KATE JOHNSON

MONTREAL — Pediatric neuropsychiatric systemic lupus erythematosus has several unique manifestations that are not seen in adult patients, and without precise questioning they could easily be missed, reported Dr. Lily Siok Hoon Lim.

Patients can have visual, auditory, and even tactile hallucinations, but about 70% of them have "preservation of insight," meaning they know these experiences are not real, said Dr. Lim, a rheumatologist at the Hospital for Sick Children in Toronto. Because they can distinguish between hallucinations and reality, the children repress their symptoms and do not tell their parents or physicians "because they don't want to be seen as crazy," she said in an interview.

Visual hallucination and distortion are seen in three-quarters of pediatric patients with neuropsychiatric systemic lupus erythematosus (NPSLE) but have not been documented in adults with NPSLE, Dr. Lim said at the annual meeting of the Canadian Rheumatology Association. "They may be looking at a picture frame on the wall, and it might distort and move in and out at them. We find a lot of patients have had mild symptoms for a long time without reporting them. Also a lot of them see bugs, or spiders crawling towards them, and that is very frightening," she said.

In an observational study which she presented as a poster at the meeting, Dr. Lim and her colleagues followed a cohort of children with NPSLE at a single center between August 1985 and December 2008. Of a total of 447 children with juvenile SLE, 53 (12%) children and adolescents (46 female) exhibited secondary psychiatric manifestations and cognitive dysfunction. Half of the subjects had psychiatric manifestations at first presentation of JSLE and 77% exhibited them within a year of diagnosis. The median age of diagnosis with psychiatric illness was 15.9 years and the median duration of psychiatric symptoms prior to diagnosis was 60 days.

Clinical and laboratory measures, imaging features, and treatment regimens were collected using standardized assessment forms, and all patients were evaluated by an experienced psychiatrist.

The clinical features of lupus-related psychiatric disease were identified and classified according to American College of Rheumatology nomenclature for adult disease (Arthritis Rheum. 1999;42:599-608), with the exception of cognitive dysfunction. "Cognitive dysfunction is controversial in lupus because if you take a whole population of lupus patients and systematically study them with neurocognitive tests, 60% of them will have something, but it's subclinical; it doesn't affect how they function," said Dr. Lim.

For this study, the investigators developed a definition of pediatric cognitive dysfunction that included patient- or parent-reported memory or attention deficit affecting academic performance. Specifically, a patient needed to fulfill the following three criteria: self-reported or observed problems with concentration or memory; significant impairment of the patient's academic performance, as indicated by a significant drop in grades; and improvement following treatment.

Using this definition, the study revealed that all patients had significant cognitive dysfunction, said Dr. Lim. "What's special is that our patients had actually reported these problems. For example, their short-term memory was bad; they couldn't remember what they ate for breakfast, or what their homework was. They also couldn't learn new stuff at school, they had word-finding difficulties, and they were also not doing well in school. So you may have had an A student going down to C."

She said that 85% of the subjects had concentration difficulties, 77% had memory deficits, 23% had psychomotor slowing, and 21% had decreased comprehension. Two patients also had prominent depressive features.

In addition, 75% of the subjects also had psychosis with hallucinations. In 83% the hallucinations were auditory, 75% were visual, and 20% were tactile. Visual distortion also was reported in 38% of this psychosis subset, she said.

In all, 42 of the 53 patients underwent magnetic resonance brain imaging, of whom 45% had normal results, 29% had cerebral atrophy only, and 17% had nonspecific white matter changes only. Of the 53 patients, 28 underwent lumbar puncture, of whom 64% had normal results, 29% had elevated total protein, and 7% had an elevated white cell count.

Prednisone was started in all patients and increased according to standard protocol. In addition, all but three patients required second-line immunosuppressant therapy (85% with azathioprine, 55% with cyclophosphamide, and 28% with mycophenolate).

"What we're finding is that even among second-line immunosuppressants, cyclophosphamide is turning out to be something that is very useful," commented Dr. Lim. "When we start patients on azathioprine because their symptoms are mainly cognitive, or they have only mild psychotic symptoms, we find that a third actually need to be switched over." Of the patients with psychosis, 60% (n = 24) also required antipsychotic therapy.

The investigators were able to collect data on response to therapy for some of the patients: Six relapsed and 25 went on to remission (although 3 of these eventually relapsed).

Response was defined as the absence of psychiatric symptoms, no antipsychotic medication, and prednisone at less than 50% of the peak dose for at least 3 months. Remission was defined as absence of psychiatric symptoms, no antipsychotic medication, prednisone at 10 mg or less a day, or 0.2 mg/kg per day or less for at least 3 months. And relapse was defined as a recurrence of psychiatric symptoms, a requirement of at least a 50% increase in prednisone dose, or a change in second-line immunosuppressive agents (not due to adverse effects), or the addition of antipsychotic medication. There were 14 nonresponders. "The nonresponse may be because they presented in adolescence, and the follow-up was short before they transferred to an adult clinic," she said. ■