Pediatric Melanoma Rare, With Puzzling Features

Children diagnosed with the disease represent many skin types and have few traditional risk factors.

BY BETSY BATES
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PORTLAND, ORE. — Melanoma in a pediatric patient is a lot like Sasquatch, Dr. Seth Orlow remarked at the annual meeting of the Pacific Northwest Dermatological Society.

It's a very rare thing to see, but if it's around, you surely don't want to miss it.

Drawing from case series large and small and from his own experience, Dr. Orlow painted a puzzling picture of childhood melanoma, from the hodgepodge of clinical features to highly variable survival figures.

Patients younger than 10 years are more likely to be nonwhite, have nodular lesions, present with head/face/neck primaries, and have metastatic disease.

Perhaps the only absolutely clear conclusion in the literature is that melanoma in children is exceedingly rare, even in referral centers seeing a large number of patients with suspicious lesions, said Dr. Orlow, director of pediatric and adolescent dermatology and chair of dermatology at New York University.

A SEER (Surveillance, Epidemiology, and End Results) database analysis of 140,206 cases of melanoma diagnosed between 1973 and 2001 found just 1,255 cases in patients under 20 years old; 204 of these were melanoma in situ, and just 95 occurred in children younger than 10 years of age.

The overall incidence of childhood melanoma rose 2.9% per year. In children younger than 10, the incidence rose 1.4% per year.

Younger children were less likely than older children with melanoma to be in a traditional high-risk category.

They were more likely to be nonwhite, have nodular lesions, present with head/face/neck primaries, and have metastatic disease. Many had a history of other malignancies and may have received radiation therapy and/or chemotherapy for leukemia or another cancer, Dr. Orlow said.

Other studies similarly have found few traditional risk factors, frustrating dermatologists who might hope they can simply raise the red flag for a fair-skinned child with a history of blistering sunburns.

"We feel, with some reason, we can identify adults who are at high risk. We know if you have a family history, you have many atypical nevi, you have a history of blistering sunburns, if you're red-haired and freckled, you're going to have an increased risk of melanoma" as an adult, he said.

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The diagnostic rules used for adults don't apply in children. "In prepubescent melanoma, all bets are off," Dr. Seth Orlow said.

"In prepubescent melanoma, all bets are off," Dr. Orlow said.

Children diagnosed with melanoma represent a rainbow of skin types.

Most of them have no family history of the disease, and many have no precursor lesions.

A series from Milan's Istituto Nazionale Tumori found that 14 of 33 children under age 14 years who were diagnosed with melanoma had amelanotic lesions.

"There's no way 40% of adults' lesions would be amelanotic," he said.

Dr. Orlow's own patients over 16 years of practicing at New York University have included a 16-year-old Jamaican girl with a primary lesion that looked like a keloid on her thigh, a 14-year-old Peruvian girl with a fingertip lesion, a 12-year-old Russian boy from Chernobyl with a lesion on the lower back, and a 12-year-old Ashkenazic girl with a small (less than 1 mm) shoulder lesion.

Another speaker at the meeting, Dr. Joseph Gruss of the University of Washington, Seattle, described a case of metastatic melanoma present at birth in an African American girl with a large scalp lesion.

The bottom line is that pediatric melanoma is a mysterious disease that calls for an open mind.

"These are not the things you're used to seeing in older children and adults. It is a very different disease," Dr. Orlow said.

Survival of melanoma in adulthood is fairly well predicted by lesion characteristics and other factors, but the survival of childhood melanoma is neither well-characterized nor consistent from study to study.

"You'll see wildly different numbers from different centers," he commented.

For example, among 13 cases in children under 17 years old seen at Montreal's Hopital Ste. Justine over a 22-year period, the 5-year survival was 59%.

Among 23 cases referred to Children's Hospital in Boston and reviewed by Dr. Ray Barnhill, a dermatopathologist, survival appeared linked to tumor type (Semin. Diagn. Pathol. 1998;15:189-94).

All five cases he characterized as "small cell melanoma" were fatal. There was no precursor lesion in four of the five, and two of the five were verrucous, Dr. Orlow said.

Of six cases of "adultlike melanoma," two were fatal. Both were on the backs of older children in the series.

One of the three patients with "Spitz-like melanoma" died, but none of the nine with "atypical Spitz tumors" did. It is doubtful that the latter cases were really melanomas at all, Dr. Orlow said.

The Milan series cited a survival rate of 90% in children under age 10 years and 47% in children over 10, in a pattern that did not seem to correlate with the apparent severity of presenting features.

"It makes you wonder if all of the lesions they were calling melanoma really were melanomas," Dr. Orlow said.

The SEER database cited survival rates of 89% and 92% in children under age 10 and aged 10-19, respectively, raising similar questions.

Indeed, a study published in 1996 highlighted the difficulty of interpreting melanoma statistics in children (Int. J. Cancer 1996;68:317-24), he said.

In this review, 42 of 60 "melanoma" lesions diagnosed in children under 16 years old in five Western European countries over a 33-year period were later reclassified as nevi.

The 5-year survival rate for the patients who had true melanoma lesions was 84%. ■

Case Illustrates Elusive Diagnosis

Melanoma in children is rare and often unheralded by a precursor lesion or melanocytic pigmentation, and it can elude diagnosis, Dr. Orlow said.

"The real thing you have to be on the lookout for is rapid and unexpected growth," he emphasized.

He described the case of a boy who first presented at age 2 years with a 2-mm papule on his right ear. The lesion was treated with liquid nitrogen.

The lesion returned and measured 4 mm at age 5, 8 mm at age 6 (when it was biopsied and diagnosed as a Spitz nevus), and 10 mm at age 7, when a recurrence was excised and a second biopsy revealed "Spitz nevus with moderate atypia."

"They were good at measuring it," Dr. Orlow quipped.

The boy was referred to New York University at age 7. A work-up revealed cervical adenopathy, and a lymph node biopsy detected metastatic melanoma.

Although the child went into apparent remission after 9 months of biochemotherapy, a follow-up positron emission tomography scan at age 11 revealed abnormal foci in the liver, bilateral cervical triangle, and right paratracheal areas.

Despite the ominous course of this case study, Dr. Orlow recommends "restraint in biopsying" when a lesion first presents in a child.

After all, he reminded the audience, any patient destined to have 30 nevi in adulthood will be developing new, completely normal lesions at ages 8, 9, and 10.

There are lesions, like the one in this child, that should have been biopsied "much earlier," he said. But there are many more "that show up in patients you wouldn't expect … when reasonable people couldn't have guessed it was anything like a melanoma until they took it out and discovered it was."

An annual examination makes sense for children older than 12 years who have multiple nevi—particularly if they are atypical—and a family history of melanoma, he said.

In children under age 12, though, those factors do not seem to clearly confer elevated risk.

It all gets back to observation, so he is ever on the lookout "for peculiar lesions demonstrating unexpected growth, especially things you can't quite characterize, like a pink, eroded papule that doesn't quite look like a pyogenic granuloma," he said.

Spotting Pediatric Melanoma

- ► Family history may be negative.
- ► Patient may be nonwhite.
- ► Child may have history of other malignancy.
- ► There is usually no precursor lesion.
- ► Amelanotic, nodular lesions are common.
- ► Rapid, unexpected growth is a red flag.

Source: Dr. Orlow