Malignant melanoma developed on a congenital melanocytic nevus with lymph node metastasis in a 19-month-old boy

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Although rare, malignant melanoma occurs in children. The risk of degeneration of a congenital melanocytic nevus into a melanoma is approximately 0.7%. We report a case of malignant melanoma that developed on a congenital melanocytic nevus in a 19-month-old boy. The child was treated by surgical resection, superficial parotidectomy, and modified radical neck dissection with adjuvant therapy. The follow-up has been 24 months without metastasis. Early clinical detection, fast histological confirmation, prompt surgery, and adjuvant therapy are the only means to achieve a long survival period for children suffering from malignant melanoma.

Key words: congenital melanocytic nevus, malignant melanoma, pediatric melanoma.

Malignant melanoma is a rare pathology in children. Childhood melanoma has been reported to account for 0.9% to 3% of all pediatric malignancies¹,². Approximately 1%-3% of all newborns have congenital melanocytic nevi of varying size¹-³. About 50%-60% of cutaneous melanomas arising from large congenital melanocytic nevi are diagnosed before age five³-⁷. Warning signs for melanoma include color change, increase in diameter, irregular borders, surface ulceration, and bleeding.

A multidisciplinary approach must be taken in children with melanoma. This should include involvement of the dermatopathologist, plastic surgeon, nuclear medicine specialist, and pediatric oncologist. We present a case of malignant melanoma developed on a congenital melanocytic nevus in a 19-month-old boy and the multidisciplinary management performed at our institution.

Case Report

A baby boy presented at birth with a pigmented, raised skin lesion measuring approximately 1 cm in diameter on the left cheek. There was no history of melanoma in his family and no evidence of melanoma in his mother. At 19 months of age, the lesion persisted and had grown darker. He underwent an excisional biopsy of the lesion. Histologic review showed a malignant melanoma with Breslow thickness of 3 mm, Clark level 4 with negative deep margin. He was referred to our institution, and the clinical exam revealed a healthy boy with an oblique scar measuring 2 cm in diameter on the left cheek. There were no palpable lymph nodes bilaterally. On the morning of the operation, the patient was sent to the Department of Nuclear Medicine for the sentinel lymph mapping. In the operation, a wide local excision was performed with a 1-cm margin, and sentinel lymph node biopsy was performed. Margins were widely clear and the sentinel node was positive for metastatic melanoma (Fig. 1). He underwent staging studies including computed tomography scans of his head, chest, abdomen, and pelvis, as well as a bone scan. None of these tests showed any evidence of metastasis. One month later, left neck lymph node dissection and superficial parotidectomy were performed, yielding 36 lymph nodes with one metastatic disease. His postoperative course was complicated by a facial paralysis that healed spontaneously within two weeks (Fig. 2a, 2b). His final staging, at the age of 21 months, was stage IIIA (T3a, N1a, M0).
The patient was referred to the Department of Pediatric Oncology, and interferon (IFN) alpha 2b treatment was planned. IFN alpha 2b was given subcutaneously five times weekly for the first month and continued as three times per week for 48 weeks. The boy is now four years old and without signs of locoregional or distant metastasis on clinical examination.

Discussion

Malignant melanoma in children can be broken down into five categories according to source: transplacental melanoma, transformation of a giant congenital melanocytic nevus, melanoma on xeroderma pigmentosum, de novo melanoma, and melanoma on a pre-existing nevus.8,9 The most common signs associated with melanoma in childhood are increase in size, bleeding, and a change in color.4,10 The 1 in 20,000 newborns with a giant congenital melanocytic nevus (>20 cm in diameter in adulthood) has a lifetime risk of developing melanoma ranging from 4%-15%. These melanomas typically develop in the first five years of life. The significance of smaller congenital melanocytic nevi (<2 cm in diameter), which occur in approximately 1 in 100 newborns, is less clear, with the rate of malignant transformation reported to somewhere between 0% and 5%, often occurring in adulthood.4,9,11 As such, current recommendations are to excise fully before the age of 10 years, if feasible. However, all concerning lesions should be biopsied as early as possible. Although small-sized congenital melanocytic nevi have slightly lower rates of transformation, our patient’s lesion was approximately 1 cm in size and had malignant transformation without family history.

Due to the high rate of nodal metastasis, several studies recommend sentinel lymph node biopsy to predict the prognosis in children with primary melanoma. This technique permits accurate staging of regional lymph nodes with a low morbidity procedure for tumors greater than 1 mm in thickness.9,12-14 A combination of isotope lymphatic mapping with intraoperative handheld gamma probing and isosulfan blue dye provides sentinel lymph node identification in 95% of cases.15-17 To identify the sentinel node (the first lymph node to receive drainage from a particular site), a colloidal material (most commonly isosulfan blue dye and/or technetium-99 m-labeled sulfur colloid) is injected around the biopsy site. This material drains from the site of injection and...
concentrates in a single or limited number of nodes, presumably having taken the same path that malignant cells would take if they were to spread to regional lymph nodes. At surgery, the colloidal material is identified within the lymph nodes by visual inspection (blue dye) and/or a handheld gamma probe (radionuclide isotope), and these nodes are then removed and carefully examined with serial sectioning and immunohistochemistry. For this patient with positive sentinel lymph node (identified melanoma), a complete lymph node dissection was performed because other lymph nodes may have also contained regional melanoma metastases\textsuperscript{9,12,18,19}. Modified radical neck lymph node dissection carried unknown risk in the development of a 19-month-old baby. However, without other treatment options, an aggressive surgical approach was chosen. Postoperatively, he did not develop any long-term morbidity from the procedure and continued to develop appropriately.

Numerous studies indicate that the natural progression of childhood melanoma is no different from its adult counterpart\textsuperscript{9,20–22}. Because of the rarity of the condition, children with melanoma undergo chemotherapy and immunotherapy modified from adult protocols\textsuperscript{16,23}. The ECOG Trial 1684 demonstrated a significantly improved five-year relapse-free survival rate (37\% vs. 26\%) and overall survival rate (46\% vs. 37\%) in patients with thick (4.0 mm) melanomas and excised nodal disease when treated with high-dose IFN alpha-2b as opposed to those undergoing observation alone\textsuperscript{16,23}. The standard therapy for melanoma in adults remains debated because no therapy has been shown to alter survival in a prospective randomized study. A retrospective analysis has shown the efficacy of sentinel lymph node biopsy and high-dose IFN in pediatric populations\textsuperscript{14}. There is no literature to guide decision-making with regard to adjuvant therapy based on a pediatric population and, because of the scarcity of the disease, there is unlikely to be any soon. However, phase III studies of chemoimmunotherapy using combinations of cisplatin, vinblastine, and dacarbazine with alpha-IFN are most encouraging\textsuperscript{2,9,24,25}. In keeping with our initial aggressive surgical management, we offered our patient only high-dose IFN therapy. In consultation with the Department of Pediatric Oncology, we decided that the potential benefit would likely outweigh the toxicity.

Cases of infantile melanoma are very rare, and only a few have been presented in the literature. Our patient had successful surgical management of metastatic congenital melanoma with wide local excision, parotidectomy and modified radical neck lymph node dissection. There has been no detectable long-term morbidity related to the surgery and adjuvant therapy. This experience supports the application of standard management for melanoma, even in a 19-month-old child.

REFERENCES

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