Pediatric Surgical Image

Non-pigmented melanoma with nodal metastases masquerading as pyogenic granuloma in a 1-year old

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Malignant melanomas are the most common skin cancer in the pediatric population. Melanoma incidence is extremely low in infants, and metastatic disease is even less common. We present the case of an 11-month-old girl who presented with a non-pigmented lesion that progressed to an ulcerated lesion. Pathology was found to be Spitzoid melanoma of 7.6-mm thickness. Micrometastases were found on examination of the sentinel lymph node. The family chose expectant observation following the excision procedure. A pediatric melanoma registry may be helpful in developing future analyses of incidence in this specialized population.

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Malignant melanomas are the most common skin cancer in childhood and adolescence, representing between 1% and 3% of all pediatric malignancies in the United States [1]. In babies, however, melanomas are extremely rare. For this reason, large-scale incidence and survival data for this subpopulation is scarce. When a melanoma is diagnosed in an infant, especially on the face, special precautions must be taken for an excision appropriate to the depth of the lesion, while ensuring a cosmetically acceptable result. A deep tumor is coupled with the additional burden of sentinel lymph node (SLN) biopsy. These procedures must be performed with heightened care due to the location and size of the patient. Provisions, including the use of gamma imaging, are available to minimize morbidity. In these complex cases, all decisions must be made with the involvement of a multidisciplinary team. In this report, we present the case of an infant found to have a melanoma of the face with lymph node metastases.

1. Case report

A healthy 15-month-old girl presented to our plastic surgery clinic for a lesion on her right face. Initially noted at 10-months of age and misdiagnosed elsewhere as a pyogenic granuloma, the skin-colored lesion had evolved into a rubbery, erythematous papule over the following month (Fig. 1). It began to ulcerate and bleed intermittently. An excisional biopsy was recommended (Fig. 2). Pathology revealed malignant melanoma, Spitzoid type, Breslow thickness 7.6-mm, Clark's level V with a positive deep margin and no perineural or angiolymphatic invasion. A proliferation of pleomorphic spindle cells was noted with abundant mitoses (10/mm²). Immunohistochemical stains demonstrated diffuse expressions of tyrosinase, s-100, MART-1, Melan A, vimentin and variable staining of CD-99. The proliferation index (MB-1 labeling) was 40 percent. Clear cell sarcoma was excluded with a negative EWSR1 result.

A lymphatic mapping study, involving preoperative lymphoscintigraphy (with perilesional, intradermal injection of 4×125 microCi Tc-99 m sulfur colloid), intraoperative gamma camera imaging and probe survey (Sentinella™; Oncovision, Valencia, Spain), and intraoperative vital dye (with perilesional, intradermal injection of 4×0.25 cc lymphazurin blue), was performed. Successful excision of the SLN in the right anterior submandibular nodal basin was documented with intraoperative gamma camera imaging and probe survey, both demonstrating only background activity in the surgical field (Fig. 3). We then undertook primary re-excision with 1-cm margins, the institutional tumor board’s recommendation considering patient size and lesion location. Frozen section reported negative margins. Coverage was achieved with a full thickness skin graft.

Pathology demonstrated negative margins with micrometastases present in the subcapsular sinus of the SLN. The treatment options were discussed with the family and expectant observation was chosen. Monthly follow-up was undertaken for the first six months, with biannual visits planned thereafter for five years, after which the

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pediatrician in the patient’s home community will take over surveillance. There is no evidence of disease at 12 months.

2. Discussion

Currently, metastases are the only reliable indicators for melanoma in comparison to Spitz nevi [2]. Spitzoid-type malignant melanoma may be suggested by a preponderance of histopathological features typically associated with melanoma: spindle and/or epitheloid melanocytic cells with asymmetry, ill-defined lateral borders, intra-epidermal pagetoid spread, deep extension, absence of maturation, atypia, and diffuse sheets of cohesive dermal cells [3].

Compared to adult disease, pediatric melanoma shows markedly different behavior [4,5]. The five-year melanoma-specific survival rate for children under 10 years of age is 88.9%, lower than in older patients [6]. Lesion thickness is cited as a most important factor with regard to metastases and survival in adults, but the implications of deeper lesions in infancy and childhood are not clear [6,7]. Investigators have yet to discover whether morphological features such as heavy spindle cell presence have any effect on outcomes [4].
There is no consensus regarding standard treatment for pediatric melanomas [1,4,6,8]. Wide local excision, recommended in adults, has been extrapolated to children. The literature reflects that patients with ulcerated melanomas and a thickness greater than 1-mm or a Clark's level IV–V thinner than 1 mm should undergo SLN dissection [4,5]. Although SLN metastases have been shown to have a higher incidence (up to 40% positive nodes) among pediatric patients when compared to adults, the incidence of recurrence in children is lower [8]. Furthermore, SLN biopsy has not led to improved survival in children [6,9]. The significance of microscopic nodal involvement in children, present in this case, has not been evaluated adequately [4]. The family decided against neck dissection after discussion, due to the morbidity of the procedure (particularly at this age) and the unclear significance of micrometastases without gross disease in the SLN.

3. Conclusion

Melanomas are unusual in the pediatric population and even rarer in infants, but careful monitoring of apparently innocent lesions is important. This case is significant for the benign-appearing, unpigmented lesion developing under a year of age. The thickness and location of the melanoma, as well as the positive SLN biopsy, add to the challenges of the case. A literature review shows that many important aspects of pediatric melanoma, including the most effective diagnostic, staging, and treatment modalities, have yet to be fully illuminated and differentiated from adult disease. Research is required to determine the significance of micrometastases on SLN biopsy. A specialized registry targeting pediatric melanoma may be helpful in defining these characteristics, prognosis and long-term survival data.

References