Metastatic Malignant Melanoma in A Child

Phaibul Sutthiwan, MD, FACS, FRCS (C)
Nivat Chantarakul, MD

Malignant melanoma in pediatric age group is a rare lesion. Some confusion exist in diagnosing this disease prior to the criteria suggested by Allen & Spitz but became established thereafter. One of the criteria is the tumor's capability to metastasize. The case herein reported is a 4 year old girl presented with enlarged submandibular cervical glands. Excisional biopsy of one of the nodes and subsequent wide excision of all enlarged node with free margin were accomplished in treating this lesion. Histologic examination revealed metastatic malignant melanoma. The primary lesion was not located despite thorough examination. A lesion at her face excised at one and a half year of age was considered possible its primary lesion but histologic documentation was not available.

Malignant melanoma is a rare tumor in the prepubertal period with less than 80 well documented cases reported.[1-4] In the past, prior to the findings of Allen and Spitz in differentiating it from juvenile melanoma, it was usually diagnosed in childhood.[5] Reexamination revealed the number of cases that fit into the pathological criteria for diagnosis of true malignant melanoma are markedly reduced in cases that have been diagnosed and registered.[5,6] One of the accepted criteria in documenting them to be malignant in nature is their capability to metastasize. We recently treated a case of 4 year old girl with metastatic malignant melanoma. Due to its rarity, it seems appropriate to record the case and to review some of the literatures.

From the Departments of Surgery and Pathology, Faculty of Medicine, Siriraj Hospital, Mahidol University, Bangkok, Thailand.

CASE REPORT

A 4 year old girl was referred from a provincial hospital due to the presence of masses under the left mandible for two years. Her past history was quite interesting in that, at the age of 1½ year, she had a lesion removed from the left side of the face. The lesion was located at the area of the left angle of the mouth at the same level as the nose. Neither the histological diagnosis nor the specimen were obtainable. The mother stated that she recovered uneventfully from the operation. About a year after the excision of the face lesion, the patient began to develop a mass at the lower border of the left mandible. It was growing slowly with no other accompanying symptoms. The mother brought her back for further medical advice and was referred to us.

At the time of hospitalization, the physical examination revealed that T 37.5° C., P. = 100/min., R = 20/min., Body weight = 12 kg. There was an old scar approximately 1.5 cm. in length at the location where a lesion was removed previously. In the left submandibular region, there was an enlarged mass which was clinically compatible with enlarged lymph nodes. A total of four glands were found, two of which were about 2 cm. and the other two about 0.5 cm. in diametre. They were firm in consistency, non tender, movable with distinct borders and smooth surfaces. Apart from her rather small stature, other physical findings were within normal limits. CBC showed

Hct = 34%, WBC = 10,000/cu. mm^3; N 50%, M 3%, E 21% with hypochromic microcytic rbc 1 +, and platelets slightly increased. Urinalysis was negative. Chest x-ray was also within normal limits and showed no hilar lymphadenopathy. Preliminary diagnosis was lymphoma. She then underwent excisional biopsy of one of the nodes for histologic diagnosis. On gross examination, the specimen consisted of a well encapsulated mass measuring 2.0 x 1.5 x 1.0 cm., smooth surface and firm in consistency. Cross-section showed a non-homogenous brownish-grey surface. Microscopically, the mass was identified as a lymph node which was mostly replaced by closely packed tumor cells. They were arranged in alveolar pattern surrounded by scanty fibrous stroma. The cells were mostly large polygonal shape and contained pale acidophilic cytoplasm with large bizarre nuclei and prominent nucleoli, some of which had a varying amount of melanin pigment granules in the cytoplasm that was confirmed by Masson-Fontana stain. Many melanophages were scattered among the tumor cell mass (Figure 1, 2). The diagnosis of metastatic malignant melanoma was reached. The patient again underwent wide excision of the remaining nodes and surrounding tissues including the left submandibular salivary gland ten days later. Further histologic examination confirmed the diagnosis and revealed the tissues excised to possess a tumor free margin. The patient made an uneventful recovery and remained well and free from recurrence and metastasis upon followup three months later.

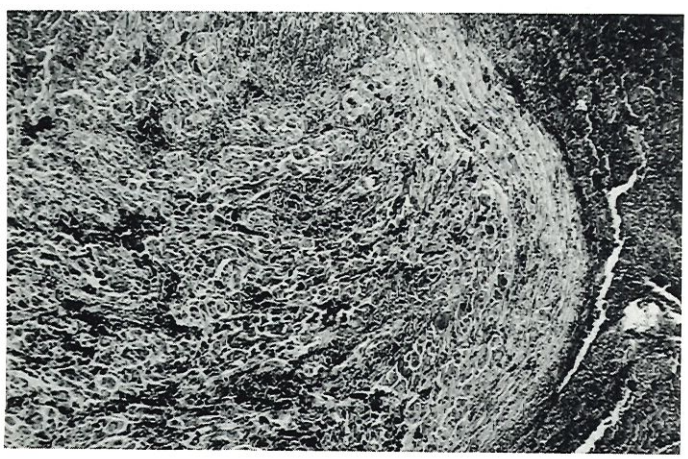

Fig. 1 Showing the lymph node was replaced by a closely packed area of tumor cells containing melanin pigments (dark cells) mixed with tumor cells without melanin pigment (light cells) (H & E x 35).

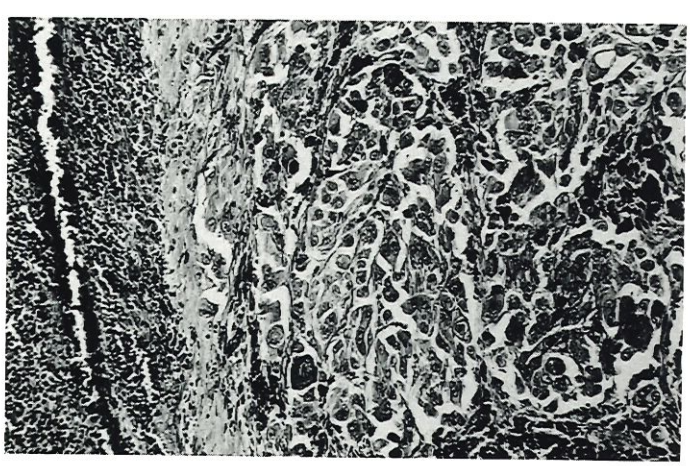

Fig. 2 Showing the lymphoid tissue was replaced by closely packed area of large polygonal cells containing pale acidophilic cytoplasm, large bizarre nuclei with prominent nucleoli which arranged in alveolar pattern. Some tumor cells have dark color which are melanin pigment granules (H & E x 100).

DISCUSSION

In general, malignant melanoma is relatively uncommon tumor with incidence of less than 5 in 100,000 population. It is accounted for only 3 per cent of cutaneous malignant neoplasm but causes 67 per cent of the deaths attributable to skin cancer, therefore emphasizing its importance.[7] Recent epidemiologic evidence has documented an increase in the incidence in adults, but not in children, and in the mortality from this disease.[8,9] The principal factors influencing the incidence of melanoma are racial susceptibility, skin pigmentation and latitude of domicile.[10] The Irish, Norwegians and Swedish all have higher incidences of melanoma than people of similar skin color living in the same latitude. Skin pigment gives protection but the pigmented races have a higher incidence of melanoma in the less pigmented regions of the body such as the sole of the foot and the various squamous mucosae. There is a direct relationship with latitude of residence and its duration, incidence of melanoma being higher with proximity to the equator. It strongly suggests that sunlight plays a major role in the development of melanoma.[11] Apart from these, there seem also to be endogenous factors responsible for familial melanoma and for the development of melanoma in young persons.

In childhood under 14 year of age or in the prepubertal period, malignant melanoma of the skin is an extremely rare tumor with the incidence reported vary from 0.3 -1.5 per cent of cutaneous malignancy.[9] There also exists the problem of overdiagnosis especially in differentiation of the true malignant melanoma from melanocytoma or juvenile melanoma. Prior to the recongnition of juvenile melanoma as a benign tumor by Allen and Spitz, malignant melanoma in children were considered as less aggressive and having relatively good prognosis. McGovern stated that the criteria for both clinical and histological diagnosis are the same for children as for adults, however the mortality is greater in children.[12]

It is found that about 40 per cent of malignant

melanomas in children arise in giant congenital hypertrophic nevi. Malignant change in such a congenital nevi was reported to range from 1.8 - 30 per cent.[3] Even though the data are not sufficient, it does appear that the giant congenital nevus is a premalignant lesion and should be excised whenever possible.[13]

Regarding the treatment, if at all possible, the biopsy of the lesion should be an excisional biopsy specimen. Even though incisional biopsy does not adversely affect the prognosis, maximum histologic information is obtained from an excisional biopsy specimen. The treatment of primary melanoma is almost always surgical except on rare occasions. The usefulness of an elective regional lymph node dissection (RLND) seems to correlate with tumor thickness.[14] When the lesion is less than 0.76 mm. a RLND is unnecessary with clinically negative nodes, however when the depth is greater than 1.5 mm. a RLND is prognostically beneficial. Lesion in between these levels must be studied further. But currently the surgeons must decide upon the appropriateness of a RLND individually.

In the case reported, undoubtedly is a case of malignant melanoma demonstrated by its capability to metastasize to the regional lymph nodes. Its primary lesion has yet to be determined but its relationship to the lesion removed at the age of 1½ year is highly likely. It is very unfortunate that the specimen of the face lesion was not obtainable for histologic review. For this metastatic lesion, the patient received only wide excision with a tumor free margin of the surrounding tissues. Neither chemotherapeutic agent nor immunotherapy were initiated. Further follow up is mandatory in determining the course of treatment and her prognosis.

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