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MALIGNANT MELANOMA IN CHILDHOOD*

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Metastatic malignant melanomas are extremely rare in children. The incidence of malignant melanoma prior to puberty is less than 1% of all melanomas. Inspite of this rarity, the disease is at least as aggresive in children as in adults[1-3].

Trozak et al[4] in 1975, reviewed the literature and reported a total of 68 cases of malignant melanoma in childhood. Very few additional cases have been reported since that time[5-7]. The purpose of this paper is to present a case report and discuss the diagnosis and management of this rare tumor in childhood.

Case Report

A four-year-old girl was admitted to the Department of Pediatrics in April 1985 because of a mass in the occipital region which was first noted by the mother as a small pigmented mass on birth. Prior to admission the lesion had been growing rapidly.

Physical examination revealed a lobulated, irregular, bluish-black mass in the occipital region measuring 10 x 15 x 10 cm. Ulcerations and bleeding points were present at the surface of the mass (Figure 1). Right exophthalmos and right cervical lymphadenopathy were also noted. A solid, irregular, mobile 8 x 8 cm mass was palpated at the right lower quadrant of the abdomen. Ascites was also present.

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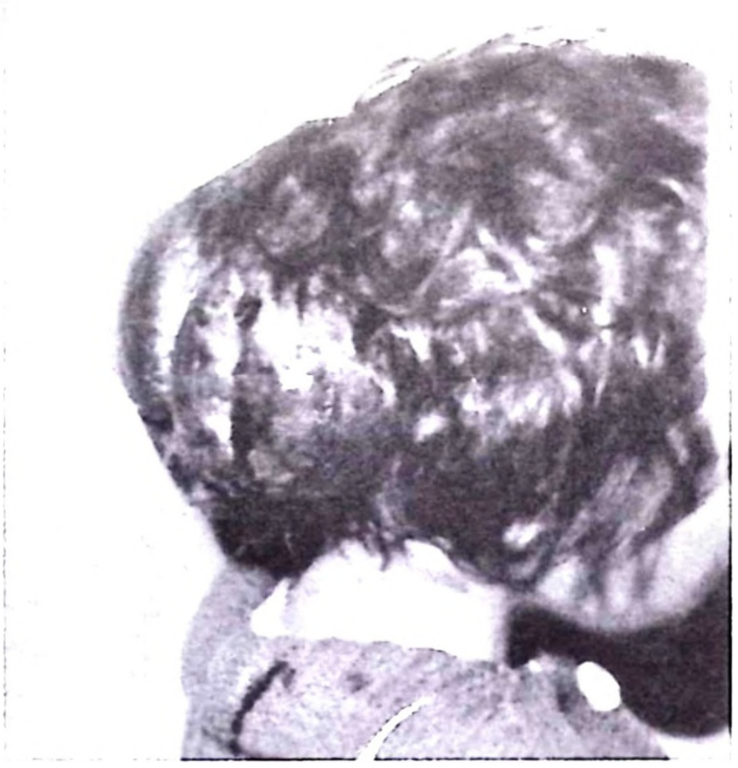

Figure 1

Bilobulated occipital mass.

A chest roentgenogram showed diffuse pulmonary metastatic infiltration (Figure 2). Radiologic studies also showed a defect in the occipital bone. A computed tomography of the brain demonstrated that the mass was extradural (Figure 3). The mass in the abdomen was also detected by ultrasonography.

Incisional biopsy of the occipital mass and excisional biopsy of the cervical lymph node was performed for definitive diagnosis. The incisional biopsy of the primary lesion revealed malignant melanoma (Figure 4) and histologically the lymph node showed metastatic malignant melanoma.

Since the clinical and the pathological diagnosis revealed a stage III malignant melanoma, surgical excision of the occipital mass was no planned. No surgical procedure was performed for the abdominal mass either.

Radiation therapy was not given. It was planned to use dimethyl triazeno imidazole carboxamide (DTIC) 600 mg/m^2 body surface and cyclophosphamide (Cytoxan®) 600 mg/m^2 body surface three times at three week intervals. Unfortunately, after the second month of the therapy the parents refused to continue the therapy and the routine follow-up examinations. During the last two weeks, however, it had been noted that the general condition of the child had worsened, the mass had become larger and ascites had increased. We received no news of the patient and assume that she died a short time after leaving the hospital.

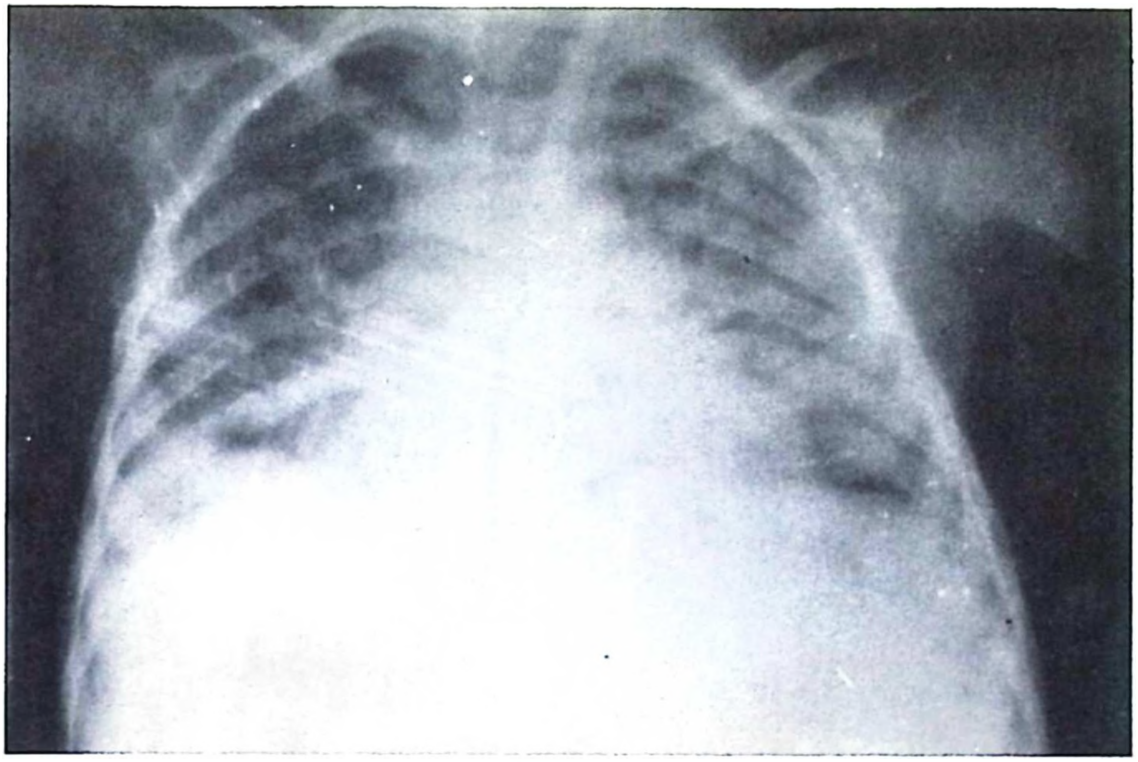

Figure 2

Chest roentgenogram demonstrating diffuse pulmonary metastatic infiltration.

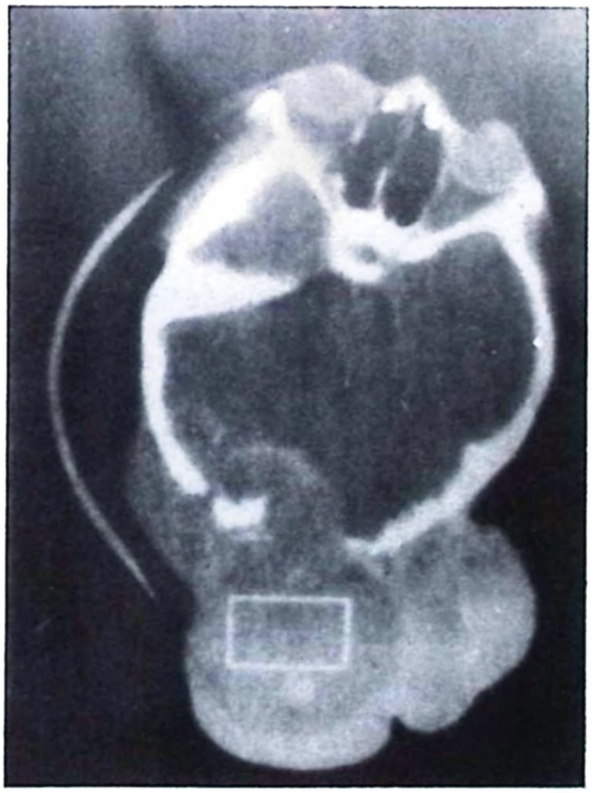

Figure 3

Computed tomography of the brain demonstrating extradural occipital mass.

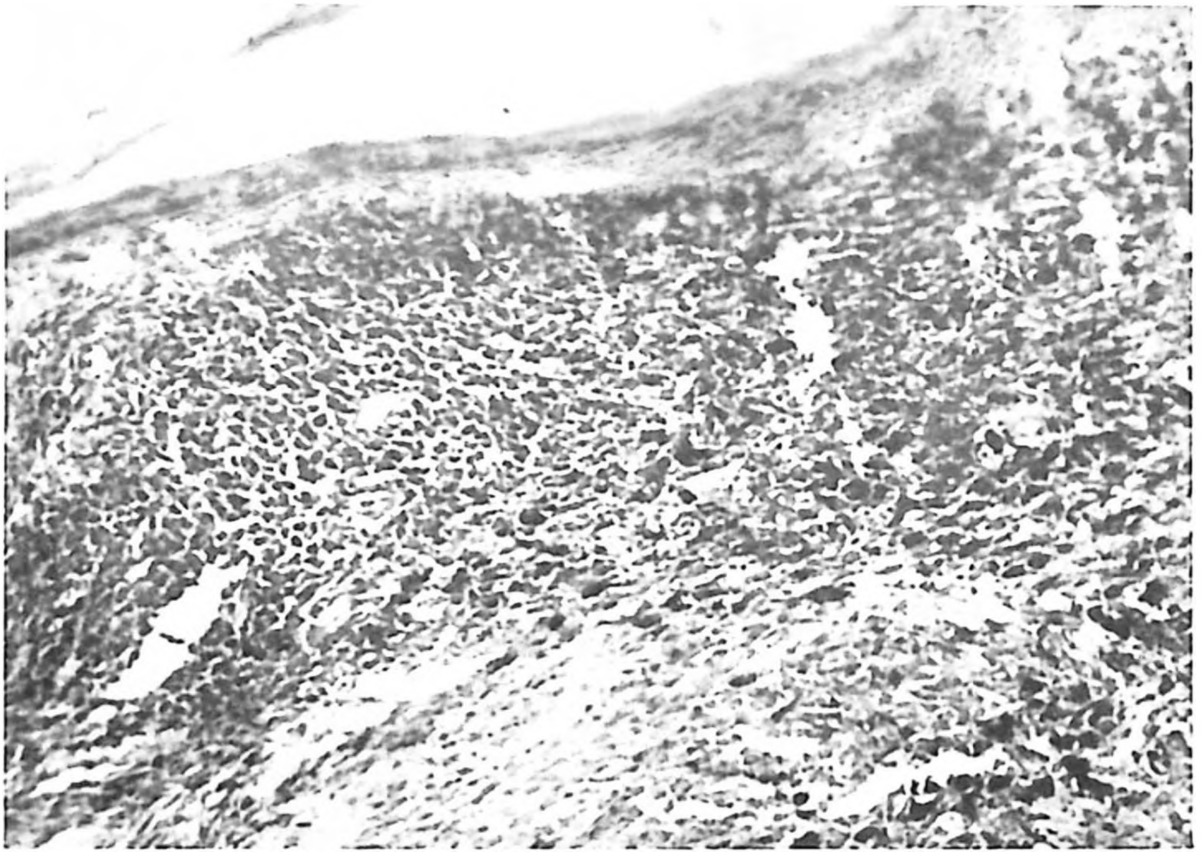

Figure 4

Primary lesion. Malignant melanoma. Superficial keratotic epithelium and diffuse tumoral invasion consisting of pigmented pleomorphic melanocytes (HE x 125).

Discussion

The diagnosis of malignant melanoma is usually based on a history of recent change in clinical signs rather than on the direct clinical appearance[8] (Table I). Definitive diagnosis of the disease can only be made after the specimen has been examined histologically.

Although 3% of all infants are born with pigmented nevi, the incidence of malignant melanoma before puberty is very low[4,9]. Skov-Jensen[3] and Trozak et al[4] divided malign melanoma in childhood into three categories (Table II). Congenital malignant melanoma (Type I) occurs as a result of metastasis of maternal malignancy to the placenta or fetus. This type of malignant melanoma is seen during the early days of life and is usually fatal within a short time with widespread metastases[3,4]. Metastatic malignant melanoma in children before puberty (Type II) is clinically analogous to melanoma in adults[4].

TABLE I

Changes in Clinical Signs which Suggest Malignant Melanoma

Size	Sudden enlargement
Colour	Hyperpigmentation Hypopigmentation
Shape	Development of nodules Elevation Irregular margins
Surface Characteristics	Loss of normal skin lines Hemorrhage after minor trauma Ulceration Serous discharge
Sensation	Itching Tenderness
Surrounding Skin	Inflammation Satellite pigmentation

TABLE II

Classification of Malignant Melanoma Before Puberty

Type I	Congenital malignant melanoma (Transplacental)
Type II	Malignant melanoma developed before puberty
Type III	Malignant melanoma developed before puberty in a nevus pigmentosus giganticus

In Type III malignant melanoma, as in our case, regional lymph node and distant metastasis are frequent. The lesions generally cover the major portions of an anatomical area. They are usually present at birth and they may occur anywhere on the body. Nevus pigmentosus giganticus may be accompanied by such developments as spina bifida, leptomeningeal melanosis and other central nervous system defects like the occipital bone defect observed in our patient[3,4]. This association is most frequent in cases of nevus of the scalp or neck[10]. The malignant change rate is 10% in giant pigmented nevus cases which is higher than in any other type of nevi[10]. Here, malignant changes come about when there is rapid growth of the mass which is an indication for surgical treatment[3,10].

Malignant melanoma may proliferate locally, spread by satellite lesions, extend via the blood stream or lymphatics and finally invade any organ of the body. The prognosis in metastatic cases is extremely poor with a five year survival rate being between 20 and 38%[3,4,9,11]. Clinical staging of malignant melanoma can be accomplished according to the clinical spread of the tumor[8] (Table III). In the case presented, metastases had been detected in the lymph nodes, chest and abdomen, and the melanoma it would seen, was a stage III one.

TABLE III

Clinical Staging of Malignant Melanoma

Stage I	Localized malignant melanoma without metastasis to distant or regional lymph nodes
Stage II	Metastasis limited to regional lymph nodes
Stage III	Disseminated malignant melanoma (Visceral, lymphatic, cutaneous, subcutaneous)

The most important method of treatment of malignant melanoma in childhood, as for adults, is surgical excision of the lesion. Excision of the tumor should include a cuff of skin surrounding the lesion, ideally 4-5 cm in all directions. The excision should also include the underlying deep fascia[2,8,9,11]. A therapeutic lymph node dissection is recommended if the lymph nodes are palpable. The major controversy in the surgery of malignant melanoma is the value of prophylactic lymph node dissection. In certain groups of patients with high Clark and Breslow levels, prophylactic lymph node dissection is a much favored method[8,11,12]. Surgical management of advanced disease is a palliative rather than a curative

procedure. Surgical excision of the mass can give relief to the patient, but should be considered only if the patient is fit for anesthesia[12].

Although chemotherapy can be useful in stages I and II, it's success is limited in advanced cases. However, single-agent or combination chemotherapy is even so widely used in the management of stage III malignant melanomas. DTIC is usually the preferred single-agent chemotherapy at most centers. For combination chemotherapy, bleomycine, lomustine, melphalan, vincristine and Cytoxan® can be used in addition to DTIC[8,12].

Although radiotherapy is being used in the management of malignant melanoma in Europe, it is not the preferred method in other countries where it is regarded as a radioresistant tumor at most centers[9,12].

Since it was suggested that this tumor may be hormone dependent, the use of hormonal therapy in advanced malignant melanoma has been recommended. Although overall response rates are low, antiestrogen tamoxifen is being tried for hormonal therapy[12].

Since melanoma has tumor antigens which produce antibodies against the tumor, immonotherapy has also been used. BCG is the most common agent being used for stimulation of the patient's immune system[8,9,12].

Several approaches to the management of advanced malignant melanoma suggest that once melanoma has spread beyond the primary site, an effective form of therapy is not available. Poor prognosis in these cases is due to late diagnosis. Because of the high risk of malignant change, special attention has to be drawn to giant pigmented nevi and to the cases with recent changes in their clinical appearance.

Summary

A case of metastatic malignant melanoma in a four-year-old girl is presented. Classification, clinical findings and various methods of treatment are discussed.

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