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Congenital Malignant Melanoma
— A Case Report —

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Herein reported is a case of congenital malignant melanoma in a premature male baby from a 25-year-old healthy mother who was found to have hydramnios at the 29th week of gestation. The pregnancy was interrupted because of a large posterior neck mass detected by ultrasonography. The large neck mass of the baby was a malignant melanoma involving deep dermis and subcutaneous tissue. The skin over the mass showed a large area of pigmentation with hairs and the pigmentation involved the occipital scalp and posterior neck. Microscopically, the tumor cells were monotonous and showed polygonal and epithelioid appearance with prominent nucleoli indicative of malignant melanoma of a minimal deviation variety. Neither junctional components nor benign dermal nevus cells were noted. There were no distant metastasis or underlying leptomeningeal melanosis. This tumor is presumed to have developed from either preexisted congenital giant pigmented nevus with loss of benign components or de novo origin.

Key Words: *Melanoma, Congenital tumor, Giant pigmented nevus, Minimal deviation melanoma*

INTRODUCTION

Pigmented moles are found in approximately 1% of newborn infants and they can rarely become malignant in infancy and childhood (Illig et al., 1985; Rhodes et al., 1982). Only 0.3 to 0.5% of all melanoma is diagnosed before the age of 13 years (Rhodes, 1983). Moreover, malignant melanoma in newborns and congenital malignant melanoma are extremely rare.

Skov-Jensen et al. (1986) reviewed 45 cases of malignant melanoma in children and found only four cases of congenital malignant melanoma in the literature. Two of these 4 cases were suspected as transplacental transmission from the malignant melanoma of their mother with subsequent development of malignant melanoma with metastasis during infancy. Remaining two cases were malignant melanoma of the skin at the time of birth from healthy mothers, and both cases died of widespread metastasis within a short period of time after birth. Until 1980, there are less than 10 cases of congenital melanoma reported in the literature (Dargeon, 1950; Derric & Thompson, 1958; Oldhoff & Koudstaal, 1968; Sweet & Connerty, 1941).

The extreme rarity of reported cases of congenital malignant melanoma prompted this report along with the interest on its pathogenesis.

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CASE REPORT

The patient was a premature male baby who died two hours after birth. His mother was a healthy 25 year old female with 0-1-0-1 in parity. Because of hydramnios the mother underwent ultrasonography which revealed a large mass in the occipital area of the fetus. Meningoencephalocele was suspected and termination of pregnancy was recommended and done at the

29th week of gestation. Past medical and family histories of the mother were not contributory.

Postmortem examination

The body weight was 1,900 gm. Measurements were as follows; crown to rump 31 cm; head circumference 30 cm; chest circumference 26 cm. and abdomen circumference 26cm. There was a large round and firm soft tissue mass involving the posterior neck and occipital area (Fig. 1), The overlying skin of the scalp showed drak black pigmentation with hairs and mass involved both dermis and subcutaneous tissue (Fig. 2).

On cut sections small areas of necrosis was also seen. The tumor extended and attached to the periosteum of the skull bone in the occipital region. The tumor measured 10×9×5.5 cm in maximum extents and weighed 300 gm. The underlying brain weighed 2300 gm and showed no pigmented lesion both in parenchyma and meninges. Remaining viscera including placenta were free of any pigmented lesion. No lymph node enlargement was found.

Microscopically, the tumor was a solid pigmented lesion, composed of diffuse monotonous intradermal proliferation of epithelioid or polygonal melanocytes larger than lymphocyte without tendency to maturation. The major portion of the tumor masses was heavily pigmented with melanin. The nuclei were round to oval and hyperchromatic with prominent nucleoli. Mitoses were rare. There were areas of necrosis and features of pseudoglandular arrangement. Junctional component of melanocytes was not demonstrated. (Fig. 4, 5).

The tumor cells were strongly positive in both S-100 protein (Fig. 6) by peroxidase-antiperoxidase method and Fontana Masson stain. The iron was negative. No reticulin fibers were seen among tumor cells. Some immature blood elements were noted in the vascular lumen, probably representing prematurity of the fetus. Electron microscopic study of the tumor cells from paraffin embedded tissue showed many melanosomes and melanin pigments dispersed or in clump in the the cytoplasm of the tumor cells (Fig. 7).

DISCUSSION

Malignant melanoma in infancy and childhood could be divided into three categories: 1) congenital malginant melanoma, 2) Malignant melanoma developed before puberty and 3) malignant melanoma developed prepubertally in a nevus pigmentosus giganticus. Our

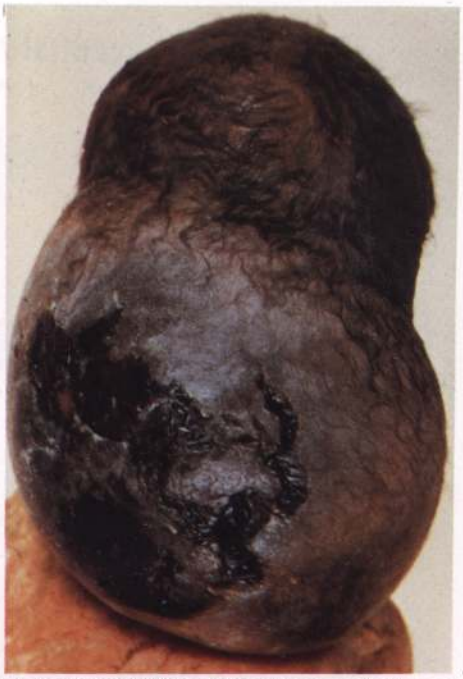

Fig. 1. A large protruding and heavily pigmented tumor mass with surface ulceration is presented in the posterior neck in the premature baby.

Fig. 2. Cut surface of the tumor shows a relatively well circumscribed and heavily pigmented tumor mass involving dermis and subcutaneous tissue.

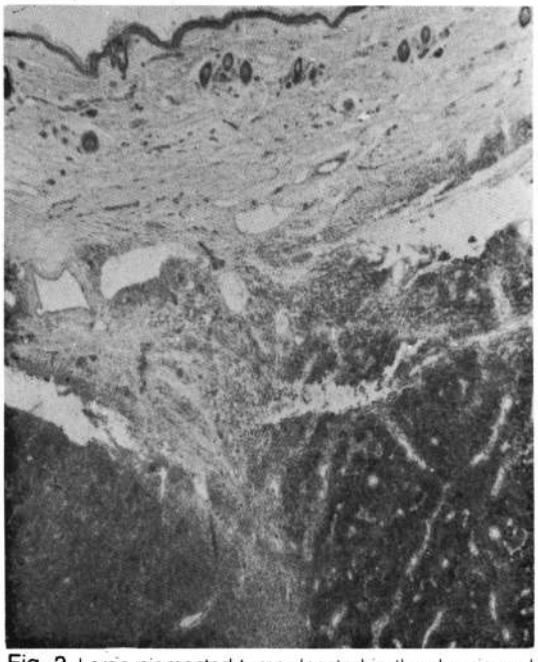

Fig. 3. Large pigmented tumor located in the dermis and sbucutis without definite junctional components in the scalp skin (H & E, ×20).

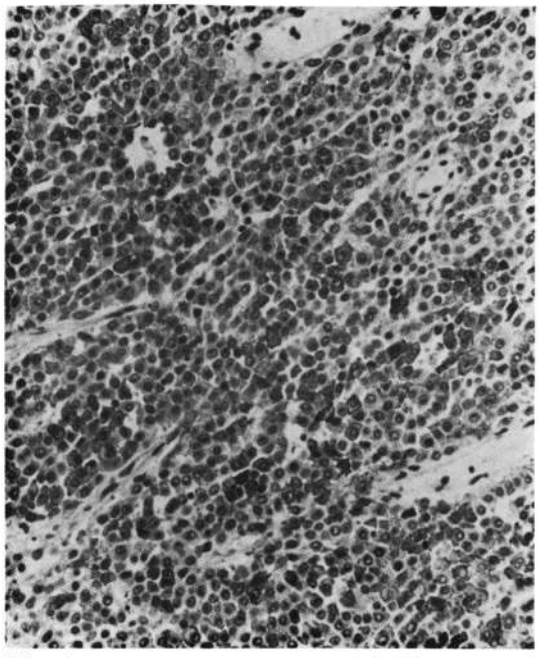

Fig. 4. Tumor masses are composed of diffuse sheets of monotonous and epitheliod melanoma cells with prominent ncucleoli accompanying heavy pigmentation in their cytoplasm. Pseudoglandular pattern is also noted (H&E, ×100).

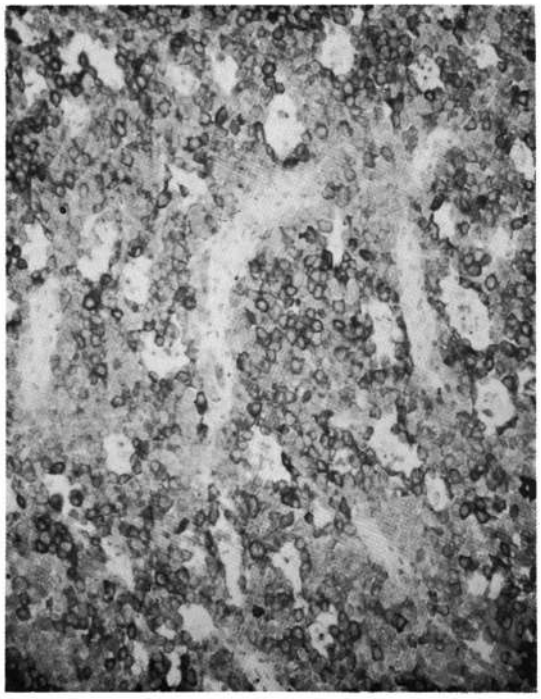

Fig. 5. Tumor cells show strong positive reaction to S-100 protein (Peroxidase-antiperoxidase method.) (×250).

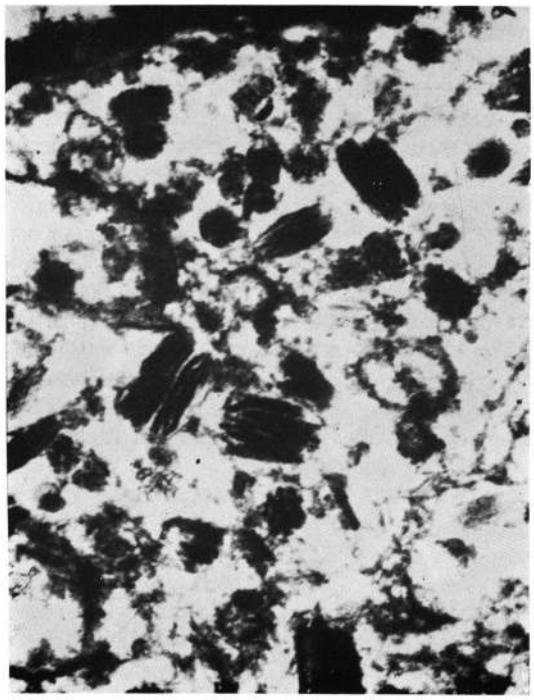

Fig. 6. Many melanosomes are dispersed in the cytoplasm of the tumor cells (EM, ×37,700)

case surely belongs to the first category (Lerman et al., 1970; Penman & Stringer, 1971).

It is indisputable that congenital nevus occasionally provide the setting for the development of malignant melanoma and their malignant transformation may occur even during infancy and childhood (Rhodes & Melsk, 1982). Among congenital nevi the incidence of malignant melanoma arising in a giant melanocytic nevus or in one of the many smaller satellite nevi is about 12 percents. Association with a preexisting pigmented lesion at the site of primary cutaneous melanoma is reported to be 18 to 85% of cases (Rhodes et al 1982; Rhodes, 1983). But actual frequency of malignant melanoma developing from the preexisting congenital nevi in fetal stage is uncertain.

However, when there is no definite area of benign component especially in dermal melanoma, it is very difficult to determine whether the malignant melanoma developed from the reexisting benign nevus or de novo origin. Therefore, the pathogenesis of malignant melanoma in this case is very difficult to be concluded because there is no associated benign or junctional components, although this case had a considerably large pigmented area in the overlying skin of the tumor and the tumor was located in the deep dermis and subcutis without junctional component. Nevus cells in giant congenital nevocellular nevi are known to often penetrate and extend into the deep reticular dermis and subcutaneous with or without junctional components. Three cases out of 7 malignant melanomas arising in giant pigmented nevus revealed no junctional component of nevus cells in the Handrickson and Ross' report (1981). It is therefore suggested that origin of this tumor could be the previous congenital giant pigmented nevus with loss of normal nevus cell components in this tumor, although origin from either cellular blue nevus or de novo can not be excluded. Differential diagnosis includes giant pigmented nevus or cellular blue nevus, but absence of pigmented dendritic melanocytes, no maturation tendency, monotonous polygonal to epithelioid pigmented cells without fascicular arrangement favor the diagnosis of malignant melanoma.

Because cases of congenital malignant melanoma are so rare, standardized classification of this lesion has not been established. However, Handrikson and Ross (1981) classified his 7 cases of malignant melanoma arising from the congenital giant pigmented nevus into 6 histomorphological categories according to neoplastic cell pattern. They are, 1) poorly differentiated small cell cancer 2) malignant cellular blue nevus 3) spindle cell malginant tumor with lamellar cell differentiation 4) so-called minimal deviation melanoma 5) heterologous malignant mesenchymal differentiation including rhabdomyosarcoma and liposarcoma, and 6) undifferentiated spindle cell cancer. Since the tumor cells in this case was composed of monotonous epithelioid and polygonal nevocytic melanoma cells with a quite large amount of cytoplasm with heavy pigmentation showed negligible number of mitoses, and also involved deep dermis and subcutaneous tissue without distant metastasis, this case could be categorized into minimal deviation melanoma. No metastatic lesion was noted in this case although it was a very large tumor mass in primary site. This favors the low grade malignancy of this tumor. It might be worth of noting that head and neck are frequent site involved by giant congenital nevocellular nevus and this pigmented lesion can be associated with leptomeninigeal malanocytosis which causes seizure and hydrocephalus that were not noted in this case (Reed et al 1965: Slaughter et al, 1969).

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