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Giant Malignant Melanoma Of Scalp In A 15 Year Old Boy With Widespread Metastases At Presentation

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Abstract : *Malignant melanoma is the most common potentially lethal neoplasm of skin. About 50-70% of all melanomas develop spontaneously, while the rest develop from a pre-existing nevus. Malignant melanoma is largely a disease of adults. Incidence of childhood melanoma is very low. Here, we describe a case of malignant melanoma in a fifteen year old boy who presented with a scalp swelling (12x10 cm). History given by parent revealed a small pigmented lesion on the scalp for last twelve years, which underwent a rapid enlargement over the last six months. Fine needle aspiration cytology of scalp swelling showed cytomorphological features of malignant melanoma. Histopathology of an incisional biopsy from the swelling confirmed the cytological diagnosis of malignant melanoma. The patient underwent imaging of various organs which revealed widespread metastasis- in mediastinal lymph nodes, liver and spleen. We report this case of giant melanoma in a teenaged boy because of its extreme rarity, challenging presentation and interesting findings.*

Keywords – *Giant melanoma, nevus, scalp, spleen metastasis*

I. Introduction

Malignant melanoma is the malignant neoplasm arising from melanocytes and/or its precursors. Apart from skin, it arises in the mucous membrane of different organs, including nose, anal canal. Malignant melanoma is primarily a disease of adults. It accounts for only 1.3% for all cancers less than 20 years of age; though contributes for up to 7% of all cancers between 15 to 19 years of age.[1] Melanoma in children is very rare and in majority of the cases it is associated with giant congenital nevus.[2] The superficial spreading form of malignant melanoma, which has a much better prognosis, is uncommon in childhood; most melanomas in children are nodular in type which has a grave prognosis.[3] In this case report we present a case of malignant melanoma in the scalp, probably, arising from a pre-existing nevus, in a fifteen year old boy.

II. Case Report

A fifteen years old boy presented to Neurosurgery outpatient department with a nodular swelling over the scalp measuring about 12cm x 10 cm [Figure 1A]. According to the mother, he had an apparent small flat pigmented macule (~0.5cm x 0.5 cm) on the scalp, which was first noticed by her when the child was three years of age. This lesion underwent a sudden rapid enlargement over the last six months and attained huge (12cm x10cm) size. He also developed complaints of headache, chest discomfort, intermittent fever, jaundice along with gradual loss of weight and appetite over a course of four to five months. The patient was advised for fine needle aspiration cytology from the scalp swelling. A brownish black particulate material was aspirated. Cytological smears from the aspirate were highly cellular, comprising of round to plasmacytoid cells having moderate cytoplasm, vesicular nuclei with coarse chromatin and large prominent nucleoli [Figure 1B]. Intracytoplasmic as well as extracellular brownish black pigment was noted [Figure 1B]. Overall cytomorphology was consistent with malignant melanoma.

A small incisional biopsy was performed from the scalp lesion for a final diagnosis before undertaking any radical therapy. The sent specimen measured 1.5cm x 1cm. Sections examined showed nests of atypical melanocyte/tumor cell diffusely infiltrating in to the dermis [Figure 1C]. Individual tumor cells were round to plasmacytoid with scanty to moderate eosinophilic cytoplasm, vesicular nuclei and large prominent nucleoli; some containing intracytoplasmic brownish black pigment [Figure 1D]. The tumor cells displayed frequent mitoses, including atypical forms. Extracellular accumulation of brownish pigments was also seen. Special stain for melanin, Masson Fontana, was positive in those intracytoplasmic and extracellular pigments [Figure 1E].

Immunohistochemistry with HMB45 revealed positive immunoreactivity in the tumor cells [Figure 1F]. The histomorphology confirmed the case as malignant melanoma. Clark's level and Breslow thickness could not be determined because only superficial incisional biopsy was carried out.

Following the confirmed diagnosis of malignant melanoma, multiple imaging studies were done depending on the patient's complaints to look for metastases. Computed tomography (CT) scan of brain revealed that the lesion in the scalp had already invaded directly through the skull to the brain [Figure 1G]. CT scan chest showed mediastinal widening and hilar lymphadenopathy [Figure 1H]. CT scan of abdomen revealed multiple hypoechoic lesions in the liver and spleen [Figure 1I]. Based on histopathology, clinical and radiological findings, the patient was diagnosed as a case of primary malignant melanoma of scalp, possibly arising in a pre-existing congenital nevus, with wide-spread dissemination (stage IV disease). He has been planned for debulking of primary lesion, systemic chemotherapy, and adjuvant radiotherapy if required.

III. Figure

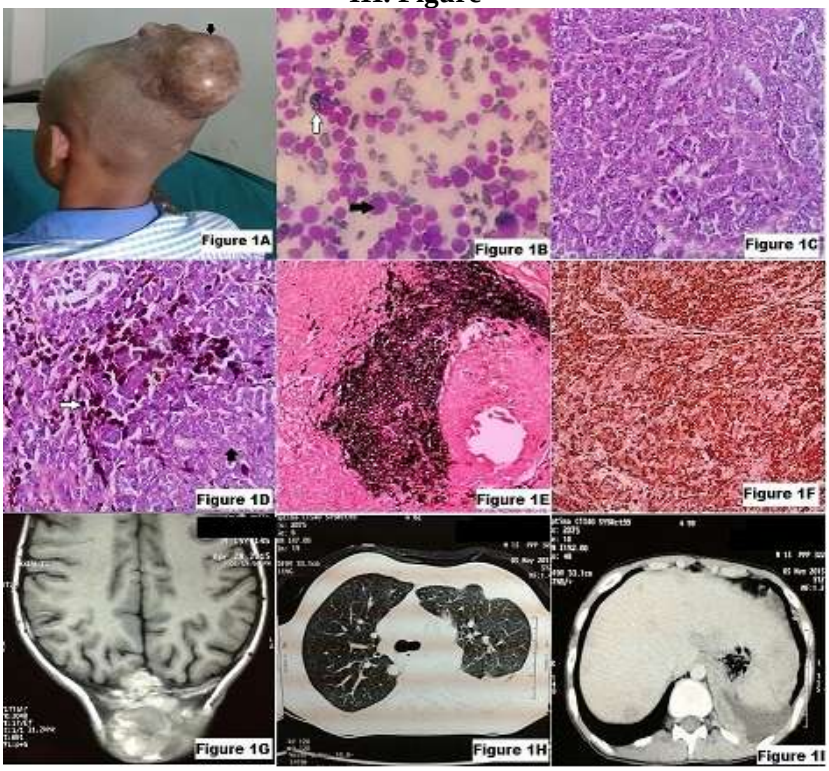

Figure 1A: Photograph: A bosselated swelling (black arrow) measuring 12cm x 10cm over scalp.
Figure 1B: Fine needle aspiration cytology: cellular smear, comprising of round to plasmacytoid cells having moderate cytoplasm, vesicular nuclei with coarse chromatin and large prominent nucleoli (black arrow); intracytoplasmic & extracellular brownish black pigment (white arrow) was noted (Leishman and giemsa, 400x).
Figure 1C - D: Histopathology: Nests of round to plasmacytoid tumor cell with vesicular nuclei and large prominent nucleoli (D, black arrow), eosinophilic cytoplasm, diffusely infiltrating in to the dermis. Some cells are containing intracytoplasmic brownish black pigment, melanin (D, white arrow) (C, Hematoxylin and eosin, 100x; D, Hematoxylin and eosin, 400x)
Figure 1E: Special stain for melanin: Masson Fontana showing black positivity in the intracellular pigments, melanin (100x).
Figure 1F: Immunohistochemistry: tumor cells showing immuno-positivity for HMB45 (DAB chromogen, 100x).
Figure 1G-I: Computed tomography (CT) images: ***G [brain]***: direct invasion of the tumor through the skull into underlying brain parenchyma; ***H [chest]***: mediastinal widening and hilar lymphadenopathy; ***I [abdomen]***: multiple hypoechoic lesions over liver and spleen.

III. Conclusion

Melanoma is a malignancy of pigment-producing cells, commonly known as melanocytes, located in the epidermis, eyes, gastrointestinal tract, and leptomeninges, oral, nasal and genital mucosa. The incidence of malignant melanoma appears to be increasing significantly worldwide; fair skin, persistent prolonged sun exposure, congenital nevi, genetic mutations involving BRAF and p 16, are common predisposing factors.[4]

Head and neck melanoma usually affects slightly older age group as compared to melanomas of other locations. Malignant melanoma of scalp is quite infrequent, comprises of 3% of head and neck melanomas.[5] The current patient was only fifteen years of age with primary lesion on the scalp.

Melanoma affecting children is a very rare entity and is usually associated with some predisposition, like giant congenital nevus or dysplastic nevi or inherited genetic defects; approximately thirty percent of malignant melanomas in childhood arise from congenital nevi.[3] In the current case, the mother gave a history of a small pigmented lesion over her son's scalp since the child was three years of age. It showed no change until last six months, when it suddenly started increasing in size. At the time of initial presentation, size of the lesion was enormous (12cm x 10cm) with skull bone destruction. Early intervention might have prevented the progression of pre-existing pigmented lesion to malignant melanoma in this patient.

The most common genetic alterations in melanoma is isolated mutation of CDKN2A (p16). Other genetic alterations include mitogen-activated protein kinase (MAPK) pathway (RAS/MEK/ERK) and BRAF, which mediates signalling pathways affecting initiation, progression, and spread of malignant melanoma.[6, 7] Majority of childhood melanoma cases are associated with loss of p16.[8] Screening of these genetic abnormalities can help in early detection and prevention.

Melanoma can metastasize extensively to various organs including brain, but metastases are uncommon until late in the course of the disease. Study done by Fife and his colleagues showed only 7% of patients presented with brain metastases at the time of initial diagnosis.[9] The present case had a huge mass over the scalp, which not only had locally invaded the skull and underlying brain parenchyma, but also had metastasized widely to distant sites including the mediastinum, lymph nodes, liver and spleen at the time of initial presentation. The 5 years survival rate for stage IV melanoma is about 15% to 20%.[5] The course of the disease in current case displayed a very abrupt progression with a potentiality for lethal outcome.

Treatment for stage IV disease is being explored over the years; current treatment options include surgical removal/metastasectomy, chemotherapy with alkylating agents, platinum analogues, and microtubular toxins, given alone or in combinations, and adjuvant radiotherapy. Immunotherapy with IL-2, IFN-α has shown success in treating stage IV disease, especially in high risk groups. Biochemotherapy, combination of cytotoxic agents with immunotherapy, is being investigated in various trials, alone or in comparison to cytotoxic/immunotherapy, and this has shown considerable promise.[10] The current case has been scheduled for primary debulking with subsequent chemotherapy and radiotherapy.

Childhood malignant melanoma on scalp is exceptionally rare. The current case came to us with huge primary scalp mass and widespread metastases involving various organs, developed over a relatively short period of time. The present lesion is thought to arise from a long standing nevus. American Cancer Society has developed five criteria, commonly known as the ABCDEs (Asymmetry, Border irregularity, Color variegation, Diameter >6 mm, Evolving), to easily identify any potential sign of malignant change in a benign nevus. Hence, clinicians and pathologists need to be aware of this disease in children and also the fact that 'ABCDE's in any melanocytic lesion occurring anywhere in the body irrespective of the age should never be neglected, as this usually suggests a malignant change with a probable fatal outcome. General population including adults, older children should also be made aware of 'ABCDE's of this potentially lethal malignancy, as early detection may offer a better survival outcome.

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