MELANOMA AND MELANOCYTIC NAEVI

MELANOMA IN A CHILD WITH LI-FRAUMENI SYNDROME

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Li fraumeni syndrome is an autosomal dominant cancer syndrome with a mutation in the TP53 gene. Many studies have shown a connection with a wide range of tumors, including melanomas, but there are some controversies because the absolute number of melanoma cases is few.
A 16-year-old boy visited our clinic with 1.2cm x 0.9cm sized asymmetric black to brown colored plaque on his right lateral thigh that he noticed since 6~7 years ago. He had no remarkable medical history and had no surgical history. Histopathological finding showed atypical melanocytic proliferation in epidermis with no dermal invasion. Melanoma in situ was diagnosed and wide excision with 1.0 cm safety margin was performed.
Since his diagnosis was unusual for his young age, extensive questions regarding family history was done. His grandfather died of sarcoma and his father had a history of thyroid cancer. Interestingly, his sister was currently admitted in our hospital for adrenocortical carcinoma with liver and lung metastasis. Considering his unique family history and the onset of cancer at a young age, genetic tests were performed to exclude Li-fraumeni syndrome in his living family members. Mutation of the P53 gene was found in his sister and the patient. We are currently undergoing further evaluation for possible cancer of other parts of the body, and patient's parents are also conducting a genetic test for the p53 gene.