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Malignant mucosal melanoma in the olfactory cleft of a 10-year-old child

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ABSTRACT

Mucosal malignant melanoma is a rare but aggressive neoplasm with high rates of recurrence and death. It is known that two-thirds of mucosal melanoma cases arise in the nasal cavity and paranasal sinuses in adults. However, there have been few studies until now on children with mucosal malignant melanoma and the related treatment. We report on a 10-year-old girl with mucosal malignant melanoma presented as a nasal polyp, which was removed via endoscopic sinus surgery and adjuvant chemotherapy without recurrence.

1. Introduction

Mucosal malignant melanoma is a neoplastic disorder in the mucous membrane caused by malignant transformation of the melanocytes, which are neuroectodermal origin cells. It is rare, accounting for 0.7–1.3% of all malignant melanomas of all sites in the US; however, it is more aggressive than cutaneous malignant melanoma with high rates of recurrence and death [1,2]. Incidence of melanoma arising in the nasal cavity and paranasal sinuses in adults reported varies from 23.3% to two-thirds of mucosal melanoma accounting for less than 5% of all sinonasal neoplasms [3–6]. As mucosal malignant melanoma is a rare disease, there have been few reports until now on cases involving children. We describe the report of a 10-year-old girl with mucosal malignant melanoma presented as a nasal polyp, which was removed via endoscopic sinus surgery and adjuvant chemotherapy without recurrence. This is the first case report of primary malignant mucosal melanoma in the nasal cavity diagnosed in children and its treatment.

2. Case report

A 10-year-old girl who had recurrent epistaxis for three months without previous other nasal problems visited a local ENT clinic. A mass in the left nasal cavity was found and she was sent to a secondary ENT clinic and received left endoscopic sinus surgery on the assumption that it was a nasal polyp. When the pathologic report confirmed it was a malignant melanoma, she was again referred to the tertiary medical center for further evaluation and treatment.

During the physical examination, the left olfactory cleft area was suspected as the site of the tumor's origin and there was a suspicious remaining lesion. A paranasal sinuses (PNS) CT revealed soft tissue density without definite enhancement in the left nasal cavity, especially from the olfactory cleft to superior turbinate and middle turbinate (Fig. 1). On the PNS MRI T2 imaging, a low signal intensity mass was distinguished from the high signal intensity lesion in the left ethmoid sinus which was believed to be an inflammatory lesion rather than a residual tumor. Gadolinium enhanced lesion was suggestive of a suspicious residual malignant lesion in the PNS MRI T1 imaging (Fig. 2). The PET CT showed an abnormal FDG uptake. Thus, on the assessment of a nasal cavity mucosal melanoma, revision endoscopic exploration and remnant mass excision in the nasal cavity were performed. The suspicious melanotic spot area was removed with a safety margin including remnant superior turbinate, posterosuperior part of middle

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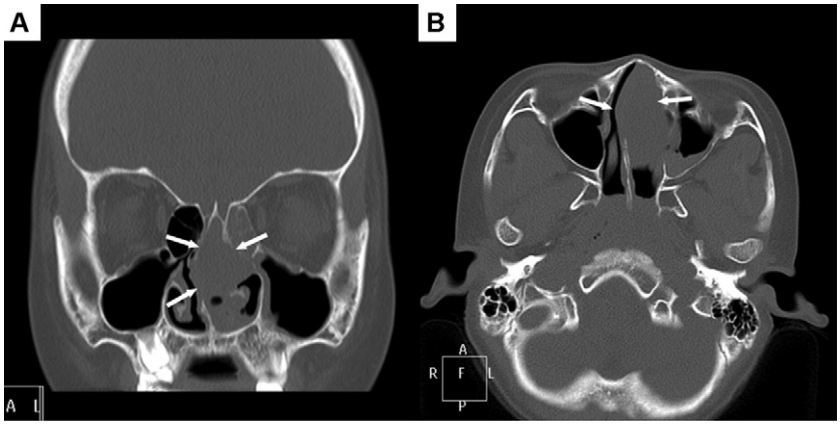

Fig. 1. Initial olfactory cleft to superior turbinate origin mass bulging to septum (A) coronal and (B) axial section (white arrows indicate the contour of the mass).

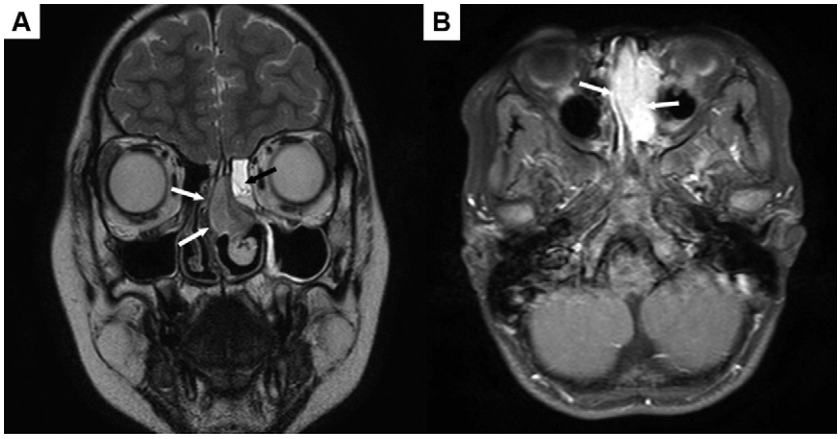

Fig. 2. Paranasal sinuses MRI imaging of the mass in the left nasal cavity which originates from the olfactory cleft (A) coronal T2 imaging, which can distinguish a mass with low signal intensity (white arrows) from the lateral ethmoid fluid collection with high signal intensity (black arrow), (B) axial T1 gadolinium enhanced lesion, suggestive of malignant lesion.

turbinate, mucosa of septum just anterior to the sphenoid foramen, and mucosa of the skull base. The final pathologic report confirmed that there was no tumor involvement with pigmented laden macrophages in middle turbinate, mucosa of septum anterior to sphenoid foramen; however, there was a residual malignant melanoma in the remnant superior turbinate and anterior skull base (Fig. 3). Immunohistochemistry for residual malignant melanoma lesion was positive for S-100 and vimentin, focal positive for HMB45, positive in 1% with Ki-67, but negative in p53 (Fig. 4). The final diagnosis was a nasal cavity malignant mucosal melanoma and the pathologic stage was T3N0M0, stage III, according to the American Joint Committee on Cancer 2010 cancer staging system. As the complete resection was thought to be made, high dose Interferone (IFN) therapy was recommended. Cisplatin, Vinblastine, Dacarbazine (CVD) chemotherapy for melanoma was applied as adjunctive therapy; Cisplatin 90 mg/m^2/day intravenously, day 1 only, Vinblastine 1.5 mg/m^2 intravenously daily, days 1–4, Dacarbazine 800 mg/m^2 intravenously, day 1 only. A total of 6 cycles of CVD chemotherapy every 4 weeks was applied for the patient with some mild side effects consisting of nasal swelling and vomiting for several days. During the chemotherapy sessions, the patient was followed up without any melanotic spot on the physical examination and imaging correlated with no recurred lesion in PNS CT (Fig. 5), and PET CT imaging. After a high dose Interferone (IFN)-α2b induction 22M unit was applied intravenously for one month, the patient showed systemic petechia as a side effect, which was resolved in a week. Following induction, 12 months of maintenance phase, IFN-α2b 10 million IU/m^2 subcutaneously, 3 times per week and IFN-α 11 million IU/m^2 unit subcutaneously 3 times per week were applied.

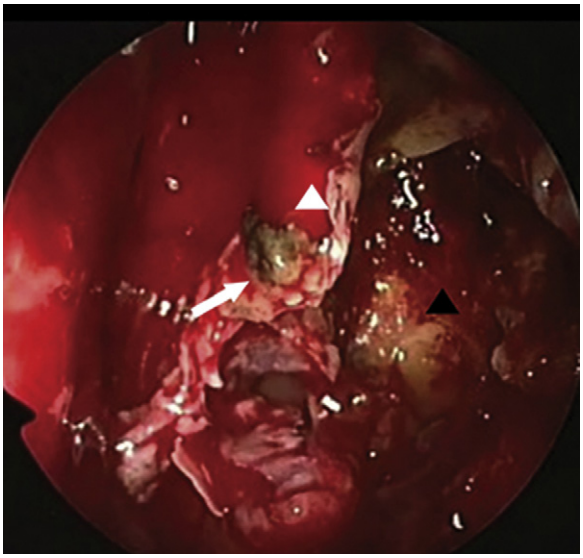

Fig. 3. Intraoperative endoscopic view of superior remnant lesion with black pigmentation (white arrow), superior turbinate (white arrowhead), posterior ethmoid cavity (black arrowhead).

The patient has been followed regularly at the outpatient clinic and has remained disease-free in the one-and-a-half-year follow-up.