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A case of recurrent malignant triton tumor successfully treated with radiotherapy

Makoto Yasuda *, Yoko Muto, Toshihiro Kuremoto, Kentaro Murakami, Toshinori Onisihi, Atsuhide Koida, Takaaki Inui, Yasuo Hisa

Department of Otolaryngology-Head and Neck Surgery, Kyoto Prefectural University of Medicine, Kyoto 602-8566, Japan

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ABSTRACT

A 15-year-old female was previously admitted to another hospital because of painless swelling of the lateral right nasal ala for 2 months. Magnetic resonance imaging revealed an expansive enhancing lesion in close proximity to the anterior surface of the right maxillary sinus and lateral wall of the right nasal cavity. Tumor extirpation was performed via the supragingival transantral approach under general anesthesia. Histopathological study revealed a malignant triton tumor (MTT) arising from the nasal vestibule. The patient was referred to our department for consultation regarding additional treatment. Because the surgical margin was positive and MTT has high malignant potential, we recommended expansion re-surgical treatment following immediate free-flap reconstruction and postoperative radiotherapy, but family consent was not obtained. Tumor regrowth was noted 1 month after her first visit to our department. The patient and her family accepted radiotherapy instead of surgical treatment. Complete remission was achieved by radiotherapy alone. No local recurrence or distant metastasis was observed for 30 months after radiotherapy. The conventional mode of treatment for MTT is radical excision followed by high-dose radiotherapy. However, this case is remarkable because our patient experienced complete remission by simple radiotherapy.

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1. Introduction

Malignant peripheral nerve sheath tumors are rare and account for 5–10% of soft tissue sarcomas [1]. These tumors are thought to arise from Schwann cells or primitive mesenchymal cells from the neural crest [2]. Among these tumors, malignant triton tumors (MTTs) characterized by heterologous skeletal muscle (rhabdomyoblastic) differentiation within the tumor are extremely rare [3]. MTTs account for 5% of malignant peripheral nerve sheath tumors [3] and have worse prognosis than malignant peripheral nerve sheath tumors, with crude 2-year and 5-year survival rates of 15% and 11%, respectively [4]. The standard therapeutic approach is aggressive wide surgical removal with adequate surgical margin, and additional postoperative radiotherapy is preferable to achieve better prognosis. We experienced a recurrent case of MTT arising from the right-sided nasal vestibule. The female patient did not accept wide surgical treatment; therefore, we performed simple radiotherapy. Fortunately, complete remission was achieved. Given that surgical removal is the preferred treatment for this tumor, this case was notable because treatment by radiotherapy alone was successful.

2. Case report

A 15-year-old female with no significant medical history and a noncontributory family history, including neurofibromatosis type 1 (NF-1), presented with painless swelling of the lateral

* Corresponding author at: Department of Otolaryngology-Head and Neck Surgery, Kyoto Prefectural University of Medicine, Kyoto 602-8566, Japan. Tel.: +81 75 251 5603; fax: +81 75 251 5604.
E-mail address: myasu@koto.kpu-m.ac.jp (M. Yasuda).

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side of the right nasal ala that had been present for 2 months. Her otolaryngological examination was normal, except for swelling of the right cheek and submucosal swelling of the right-sided nasal vestibule. Her neurological and ophthalmological examination findings were normal.

The patient showed no clinical evidence of Von Recklinghausen disease. Parenteral antibiotic therapy resulted in no reduction of the mass, and her facial mass had grown gradually. Several fine-needle aspiration cytological samples from the supragingival mucosa revealed a few glandular epithelial cells with no atypia, which suggested no malignancy. Magnetic resonance imaging (MRI) revealed an expansive enhancing lesion in close proximity to the right maxillary sinus anterior surface and the right nasal cavity lateral wall (Fig. 1).

Based on the clinical and radiographic findings, the presumptive diagnosis was a benign or malignant tumor arising from the nasal vestibule. Tumor extirpation was performed via the supragingival transantral approach under general anesthesia. The tumor was close to the skin surface and had caused skin perforation, and almost the entire tumor could be resected by naked eye visualization (Fig. 2). A part of the right major alar cartilage had to be resected together with the tumor because of adhesion. Hematoxylin and eosin staining revealed the proliferation of spindle cells with a wavy nuclei foamed bundle arrangement against a background of eosinophilic plasma, which suggested malignant peripheral nerve differentiation (Fig. 3a). Immunohistochemical staining of the tumor for desmin and myogenin was focally and occasionally positive, respectively, suggesting heterologous rhabdomyoblastic differentiation (Fig. 3b, c). Considering these findings, the resected tumor was diagnosed as a malignant peripheral nerve sheath tumor with a rhabdomyoblastic component, namely MTT. The surgical margin was pathologically positive. The patient was referred to our department for consultation regarding future treatment.

Because MTT has high malignant potential and no distant metastases were identified on positron emission-computed tomography (PET-CT), we recommended expansion re-surgical treatment, including resection of cheek skin and parts of the external nose and upper lip following immediate free-flap reconstruction and postoperative radiotherapy. We explained our plan to the patient and her family, but consent was not obtained because of cosmetic concerns. One month after her first consultation day at our hospital, she noticed re-swelling on the right side of her face. Fine-needle aspiration of the supragingival mucosa revealed a class 5 malignant tumor. CT revealed a regrowth mass outside her right nasal ala (Fig. 4). We recommended radiotherapy instead of surgical treatment as soon as possible to treat the recurrence. They finally accepted our second plan, and radiotherapy alone (60 Gy over 6 weeks) was initiated for her right cheek and maxilla. Complete remission was achieved and her post-therapeutic course was uneventful. She had no local recurrence or distant metastasis during the 30 months after radiotherapy.

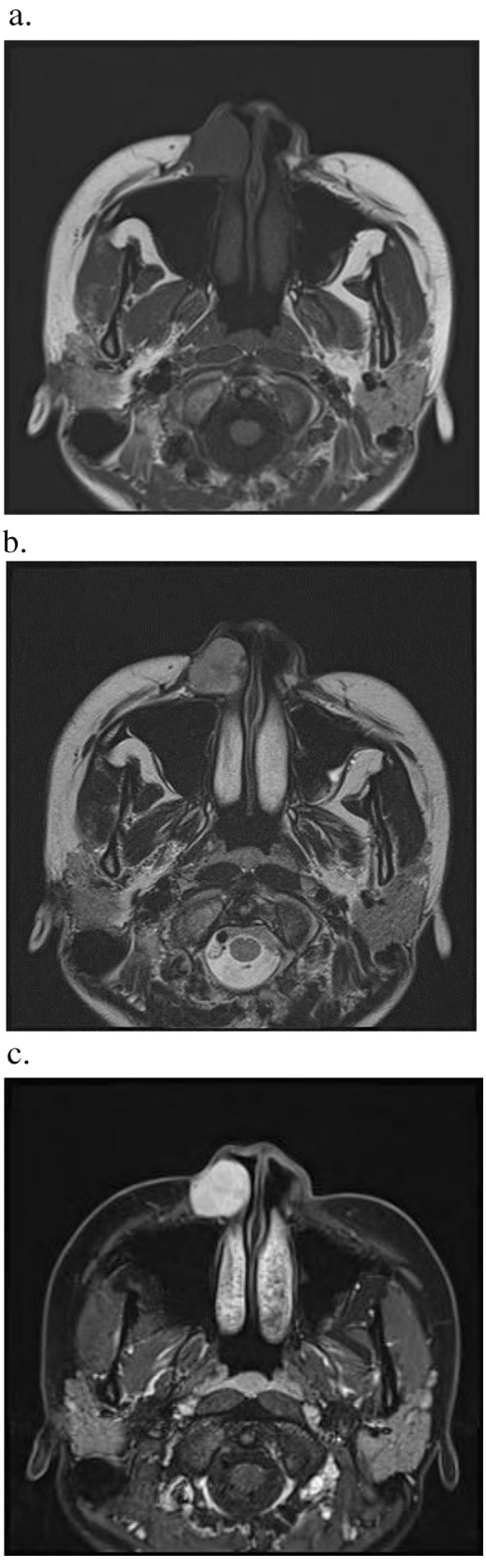

Fig. 1. Preoperative axial MRI showing an expansive and well-enhanced mass within the right nasal vestibule extending to the subcutaneous area of the maxilla. (a) T1-weighted image, (b) T2-weighted image, (c) fat-suppressed T1-weighted image with gadolinium injection.

3. Discussion

MTT is a malignant peripheral nerve sheath tumor with rhabdomyoblastic differentiation [5] and was first described in

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