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Monophasic synovial sarcoma of the nasopharynx

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ABSTRACT

Synovial sarcomas are rare, aggressive malignant neoplasms, and less than 10% of cases involve the head and neck. Cases that involve the nasopharynx are even more exceptional and little information is available concerning their diagnosis and management. We report the third case of synovial sarcoma of the nasopharynx, which was diagnosed as a monophasic type and was successfully treated with a complete surgical excision followed by irradiation. The present case indicates that appropriate immunohistochemical and cytogenetic analysis are essential for accurate diagnosis of monophasic synovial sarcoma in unusual locations. A review of the literature indicates that synovial sarcoma of the nasopharynx exhibits an improved prognosis following tumor resection and postoperative adjuvant radiation unless it invades adjacent bones, even though the tumor is larger than 4 cm.

1. Introduction

Synovial sarcoma (SS) is a rare, aggressive, malignant neoplasm that typically affects young patients and commonly arises in the soft tissues of the extremities [1,2]. Less than 10% of cases involve the head and neck [1–4]. In the region of the head and neck, the nasopharynx is an unusual site for this tumor. To our knowledge, two previous cases of primary SS involving the nasopharynx have been reported [5,6]. Owing to the paucity of cases of SSs in the nasopharynx, information regarding appropriate therapy for this tumor is limited. We report the third case of a SS arising from the nasopharynx that occurred in a 37-year-old man, along with a review of the available literature on nasopharyngeal SSs.

2. Case report

A 37-year-old man presented with right-sided symptoms of nasal obstruction and epistaxis that had progressively worsened over the past 4 weeks. Flexible fiberscopic examination revealed a large, lobulated soft tissue mass with an extensive hemorrhagic and necrotic appearance occupying the nasopharyngeal cavity and extending to the oropharyngeal space (Fig. 1). A computed tomography (CT) scan with intravenous contrast revealed that the tumor originated from the post-inferior edge of the nasal septum without any bony involvement (Fig. 2A). Magnetic resonance imaging (MRI) analysis demonstrated a lobulated soft tissue mass that filled the nasopharyngeal space, reaching the oropharynx (Fig. 2B). [18]-Fluorodeoxyglucose positron emission tomography analysis indicated a positive accumulation in the nasopharynx with a maximum standard uptake value of 4.8 and no evidence of lymphatic or distant metastases. The biopsy of the tissue mass suggested a spindle cell neoplasm including fibrosarcoma, malignant vascular tumor, and monophasic SS. He was treated surgically with curative intent. Under general anesthesia, local excision via the oral cavity was performed with sufficient surgical margin of about at least 10 mm of macroscopically healthy tissue. Because the tumor arose from the superior aspect of the soft palate and was pedunculated without bony involvement, the full-thickness local excision of the soft palate was done. As nearly left half of the soft palate was left, immediate reconstruction using the right palatal island flap to close the postoperative defect of the soft palate enabled restoration of the functionality of the patient's phonation and deglutition. The resected tumor, which measured 80 mm × 45 mm × 20 mm, was rubbery and lobulated. The histopathological examination and immunohistochemical analysis exhibited a spindle cell appearance with focal epithelial membrane antigen (EMA) immunoreactivity, which was consistent with a monophasic SS (Fig. 3A–C). A diagnosis of SS was confirmed using conventional cytogenetic analysis that revealed the following clonal karyotypic abnormalities: 46, Y, t(X;18)(p11.2;q11.2), t(5;20)(q13; p11.2) (Fig. 3D). Although the surgical margins were microscopically tumor-free, postoperative local radiation at a dose of 60 Gy was administered. At the 48 month follow-up no evidence of recurrence and no new relevant symptoms were observed.

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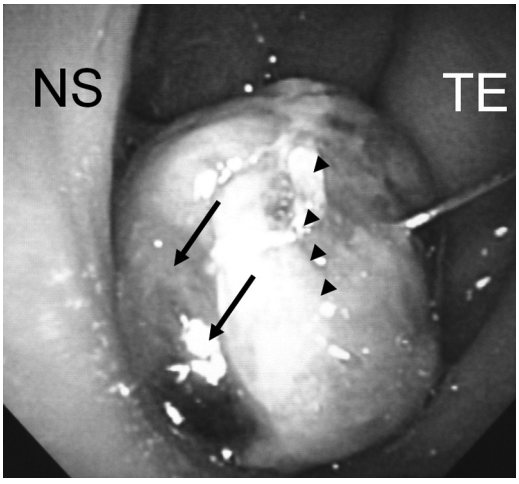

Fig. 1. Endoscopic view of the left-sided nasopharynx showing a large, lobulated soft tissue mass with an extensive hemorrhagic (arrows) and necrotic appearance (arrowheads). NS: nasal septum; TE: tubal eminence.

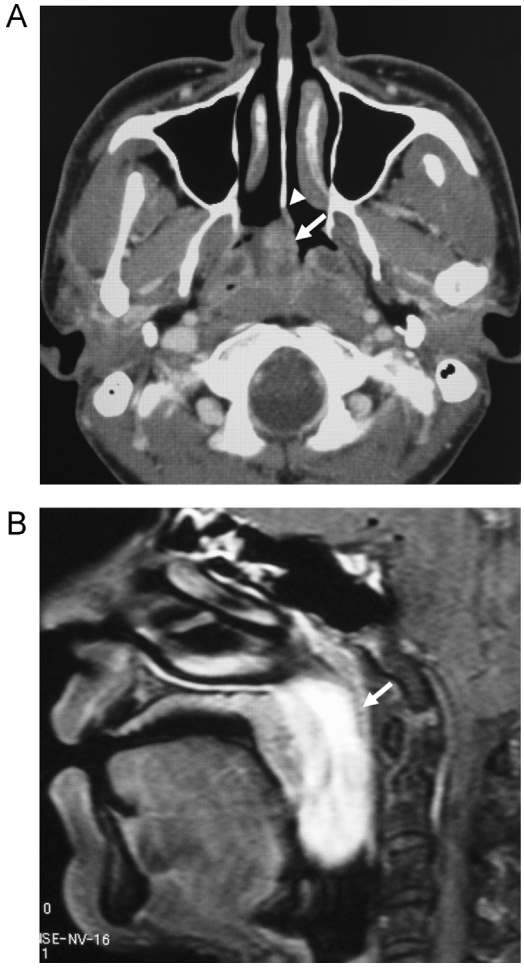

Fig. 2. Preoperative cross-sectional images of the nasopharynx. (A) A CT scan with intravenous contrast shows a slightly enhanced mass (arrows) in the nasopharynx. Note that the tumor exhibits a thin mucosal pedicle (arrowhead), seemingly arising from the posterior edge of the nasal septum. (B) A gadopentetate dimeglumine-enhanced T1-weighted sagittal image (TR, 770 ms; TE, 18 ms) shows a nasopharyngeal hyperintense soft tissue mass suspended to the oropharynx (arrow).

3. Discussion

We report an unusual case of SS of the nasopharynx, an extremely rare site for the occurrence of SS. Despite the name, SSs do not originate from synovial tissue, but rather from pluripotential mesenchymal cells near or even remote from articular surfaces [7]. SSs most commonly arise in the lower extremities adjacent to the knee [1,2], and less than 10% of cases involve the head and neck [1–3]. In the head and neck, the most common location of SSs is the neck, where they are proximal to the prevertebral areas from the skull base to the hypopharynx [3,4]. Other locations of SS in the head and neck reported in the literature include the orofacial region [8], the orbita [9], the sinonasal region [10], and the pharynx [11,12]. Only two previous cases arising in the nasopharynx have been described in the literature to our best knowledge [5,6]. Of the previous reports [5,6], one case arose from the posterior nasopharyngeal wall, and the other arose from the Eustachian tube. However, the tumor in the present case arose from the superior aspect of the soft palate, which constituted the inferior wall of the nasopharynx. These manifestations of this tumor indicated there was no subsite of predilection in the nasopharynx. The characteristics of the three cases of nasopharyngeal SS are listed in Table 1.

Head and neck SS presents both diagnostic and treatment challenges because of the limitations of knowledge and experience for this neoplasm, particularly for cases arising in the nasopharynx. Microscopically, synovial sarcoma is classified into two types: biphasic and monophasic [13]. The biphasic type is the classic type that contains epithelial and spindle-cell components in various proportions, whereas the monophasic fibrous type contains only spindle cells and is a relatively common tumor in soft tissue sites, but uncommon in the head and neck [13]. The diagnosis of biphasic SS is generally straightforward, whereas a monophasic SS diagnosis requires confirmation of epithelial differentiation by either immunohistochemical staining or detection of cytogenetic alterations. Positive immunohistochemical staining for cytokeratin or epithelial membrane antigen helps to confirm the diagnosis of monophasic synovial sarcoma. Other markers, including CD99, CD56, and Bcl-2, may display positivity [13]. In the present case, positive immunohistochemical staining for EMA helped to confirm the diagnosis of monophasic SS. The previously reported cases of SS in the nasopharynx consisted of one monophasic type and one biphasic type, and our patient is the second case of a monophasic type in the nasopharynx. Another potential diagnostic marker is chromosomal rearrangements. The t(X;18) translocation is a specific marker of synovial sarcomas [13]. In the present report, this clonal karyotypic abnormality was confirmed by the conventional cytogenetic analysis. The present case indicates that appropriate immunohistochemical and cytogenetic analysis are essential to diagnose monophasic SS in unusual locations.

SS is an aggressive tumor and overall prognosis for head and neck synovial sarcoma is not good because it exhibits a tendency toward local recurrence as well as toward metastases; the overall 5-year survival rate has been reported to be 47–72% [3,13]. At present, adequate surgical excision is the most appropriate procedure to prevent local recurrence. The deep mid-face region, including the nasopharynx, is one of the most difficult areas in which to gain wide surgical access. For larger tumors in particular it is necessary to consider the reconstruction for the mucosal defect following tumor excision. However, all of the nasopharyngeal synovial sarcomas were extirpated completely via transoral [5] or endonasal [6] approach without reconstruction of the surgical defect except for our patient. Because the lesion in the present report was pedunculated without bony involvement, it was successfully treated by complete resection with reconstruction of the soft palate performed using a local hard palatal mucosal flap.