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Letter to Editor

Synovial sarcoma of the hard palate: The third case in the medical literature

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Abstract
Synovial sarcoma is a high-grade soft tissue sarcoma that rarely arises in the head and neck region. It affects the parapharyngeal space and the hypopharynx most commonly and it has different presentations based on the affected site. In extremely rare occasions, it involves the hard palate such as in our case where a 24-year-old female patient presented with a mass lesion involving the left hard palate, which was identified clinically and by imaging studies. The histopathological assessment confirmed that it was a monophasic synovial sarcoma which was also confirmed with further molecular studies. The patient underwent surgical excision and postoperative radiotherapy. Her close follow up over a 6-year period that followed her curative treatment has demonstrated no evidence of disease recurrence or distant metastasis. Surgical excision is the mainstay of treatment for synovial sarcoma and adjuvant radiotherapy is advised. Long-term follow up is recommended because of the remote possibility of late recurrence of the tumor.

Introduction

We present the third case of synovial sarcoma of the hard palate with a long follow-up period without recurrence or metastasis. Synovial sarcoma, a high-grade histological variety of sarcomas, is the fourth most common entity after malignant fibrous histiocytomas, liposarcomas, and rhabdomyosarcomas [1]. It accounts for 7–10% of all soft tissue sarcomas. The extremities in young adults are the most commonly affected site [2].

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Three percent of synovial sarcomas arise in the head and neck, [3] predominantly in the hypopharynx and parapharyngeal spaces, with the majority being in the paravertebral connective tissue and less commonly in the larynx [4]. In 1954, Jernstrom reported the first case of head and neck synovial sarcoma [5]. Hard palate synovial sarcoma has been mentioned in only two medical reports so far.

Monophasic and biphasic synovial sarcomas are the two subtypes of synovial sarcoma described in the literature with the monophasic type containing only spindle cells and the biphasic containing both spindle and epithelioid cells [1].

Case report

A 24-year-old patient presented to our clinic with a swelling of the hard palate that caused discomfort during feeding. She had no other head and neck or upper aerodigestive tract manifestations. She had no history of smoking or alcohol consumption and her past medical history was otherwise unremarkable. The clinical assessment confirmed the presence of a firm 5 cm × 3 cm mass lesion involving the left side of the hard palate that was extending to the alveolar process. No other head and neck abnormalities were detected and there was no clinical evidence of cervical lymphadenopathy. An incisional biopsy revealed a diagnosis of synovial sarcoma positive for SS18 gene rearrangement at 18q11.2 by fluorescence *in situ* hybridization (Fig. 1). Immunohistochemical staining was done as well (Figs. 2 and 3).

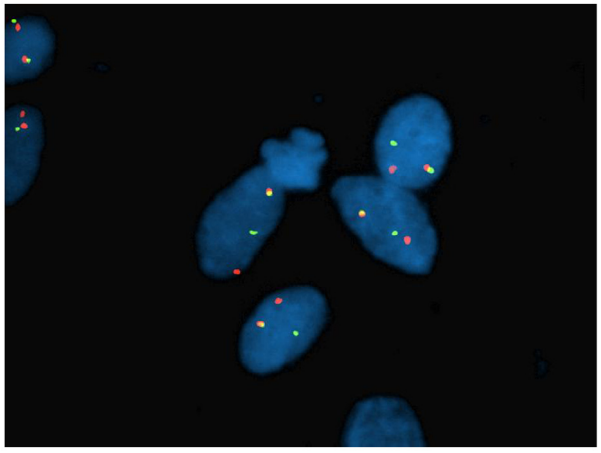

Fig. 1 Fluorescence *in situ* hybridization.

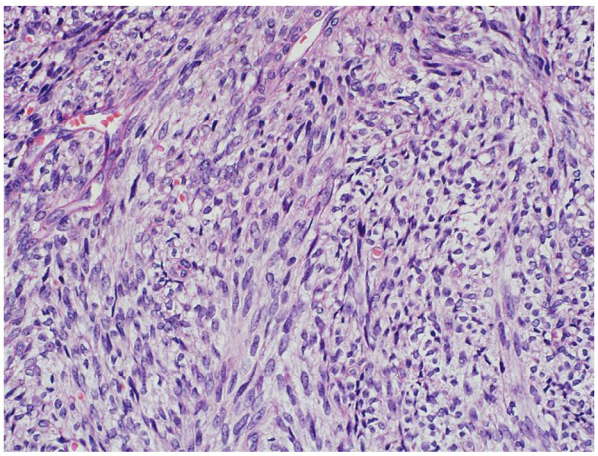

Fig. 2 Monophasic synovial sarcoma showing fascicles of spindle shaped cells.

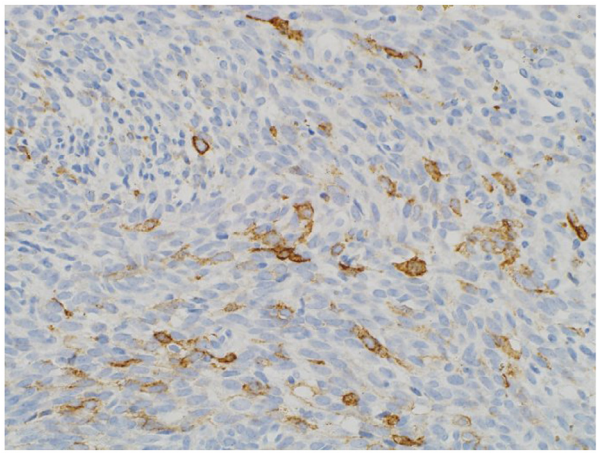

Fig. 3 Immunohistochemical study for epithelial membrane antigen (EMA) showing positive cells.

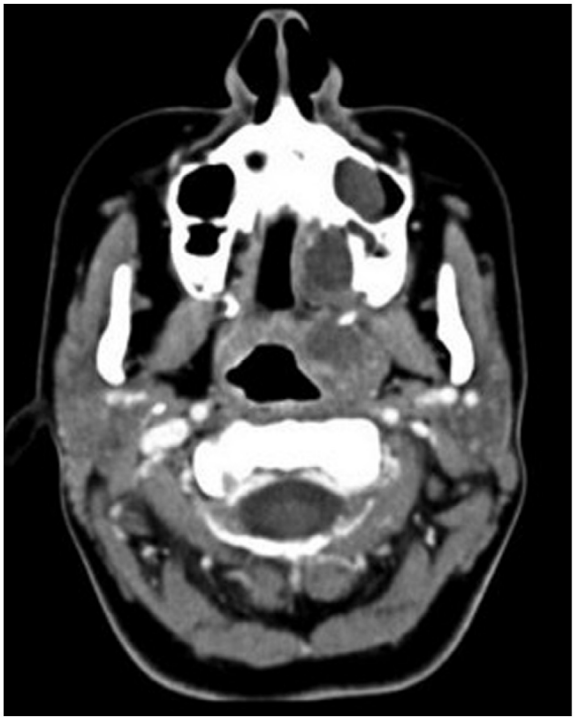

Fig. 4 Axial computed tomography showing a mass occupying the undersurface of the hard palate on the left side extending posteriorly into the lateral wall of the nasopharynx. Anteroposterior extension is around 5 cm. Side-to-side extension around 1.5 cm and thickness about 2.5 cm in the posterior region.

Computed tomography (CT) of the head and neck delineated the extent of the lesion (Fig. 4) and the positron emission tomography—CT showed normal distribution in the chest, abdomen, pelvis, and musculoskeletal system, with no definitive activity noted in the maxilla.

The patient underwent left partial maxillectomy as well as excision of the left side of the hard palate and soft palate. Part of the ipsilateral lateral pharyngeal wall was also resected. All resection margins were negative. The defect was reconstructed with a dental prosthesis.

The final pathology report confirmed that the tumor was synovial sarcoma with a cumulative size of 5 cm in maximum dimension. There was no evidence of vascular invasion and, again, all surgical margins were negative for malignancy.

The patient received postoperative radiation, which consisted of 25 sessions of 50 Gy intensity-modulated radiotherapy.

The post-treatment positron emission tomography—CT scan and other imaging modalities confirmed that there was no evidence of residual disease, recurrence, or distant metastasis.

Over the past 6 years, our Head and Neck Multidisciplinary Team has closely followed up the patient and, so far, she has had no evidence of local disease recurrence or distant metastasis.

Discussion

Synovial sarcoma is a malignant neoplasm that arises from primitive pluripotential mesenchymal cells [6,7]. It is a high-grade histological variety of sarcoma and it accounts for 7—10% of all soft tissue sarcomas. The extremities in young adults are the most commonly affected site [2]. Patients between the ages of 15 years and 40 years are mostly affected [8]. Both sexes are equally affected. In the head and neck region, the parapharyngeal space is the most commonly involved site [9]. In the oral cavity, the posterior tongue is the most common site described in medical reports followed by cheeks, soft palate, retromolar area, and floor of the mouth [10].

Synovial sarcoma of the hard palate is a rare entity, which was first published in the literature in 2004 [11]. To the best of our knowledge, only two cases have been described so far [9,11].

The clinical presentation of head and neck synovial sarcoma is usually determined by tumor site. Since it is a slow growing mass, patients remain asymptomatic until a size sufficient enough to cause pressure symptoms on the adjacent structures is reached [12]. Symptoms may range from a palpable mass causing local pain in lesions arising from cervical or parapharyngeal space to the presence of dyspnea, hoarseness, or dysphonia in the tumors arising from laryngeal sites [13]. Tumors arising from the oral cavity mainly present as a slowly enlarging, painless, nontender, rounded, and deeply seated mass. Some patients may have symptoms of pain and others may complain of sore throat. These tumors are often firm, rubbery, well-circumscribed mass lesions. Some may even be pedunculated, polypoidal, or have areas of ulceration and hemorrhage. Cervical lymphadenopathy may also be found at the time of presentation [10].

CT scans are a useful method of evaluating head and neck synovial sarcomas. The mean size of those lesions range from 2.7 cm to 7 cm, with the majority of them showing a homogeneous well-defined lesion without multiloculations [13]. Calcifications were described however, as well as hemorrhage and necrosis [13,14]. Enlarged lymph nodes maybe detected, however histological analysis is warranted.

Histopathological appearance combined with histochemistry, immunohistochemistry, electron microscopy, and cytogenetic analysis are used for the diagnosis of synovial sarcoma and have been used to confirm the tumor morphology [10]. As mentioned earlier, monophasic and biphasic synovial sarcomas are the two subtypes described in the literature with the monophasic type containing only spindle cells and the biphasic containing both spindle and epithelioid cells [1].

In more than 95% of synovial sarcomas, an SS18-SSX fusion gene can be found and is considered to be a clinically diagnostic marker. In the development of synovial sarcoma, this translocation has been regarded as a chimeric fusion oncogene. In a meta-analysis and systematic review of the prognostic value of SS18-SSX fusion type in synovial sarcoma, there was no significant difference in overall survival or disease-specific survival between patients with SS18-SSX1 and SS18-SSX2, however, progression-free survival and metastasis-free survival were unfavorable in patients with SS18-SSX1 [2].

Surgical excision is the mainstay of treatment of head and neck and hard palate synovial sarcoma and adjuvant radiotherapy improves local control of the disease and overall survival, however, no clear role for adjuvant chemotherapy has been found and more studies need to be conducted to validate its effectiveness [12].

In terms of survival, similar rates have been described in the head and neck when compared to the extremities with the most important prognostic factor being the surgical margin status [9]. Patient age and sex correlate with survival. Five-year event-free survival rate was found in 90% of patients aged 5 cm in diameter have a higher risk of local recurrence, distant metastasis, and mortality when compared to those $\leq$5 cm. The histological subtype has not been found to have a prognostic importance [12].

Metastasis and local recurrence are known to occur late in synovial sarcomas because of the slow-growing nature of these tumors. It is for that reason that long-term follow-up is recommended for those patients. Krieg et al. observed that the 5-year survival in patients with synovial sarcoma was 75.8% and the 10-year survival was 62.9%. However, the 15-year survival was only 46.5%, which reflected that metastasis is a late finding in these patients. The suggested follow up period of >10 years with good patient education and thorough history and physical examination is important in early detection of recurrence [16].

Conclusion

Synovial sarcoma of the head and neck, in general, and of the hard palate, in particular, is a rare disease entity that is best confirmed by radiological, histopathological, and molecular studies. According to most authorities, surgical

excision with negative margins is the mainstay of treatment and adjuvant radiotherapy is advised. The role of chemotherapy, however, is yet to be established. The medical literature reports recommend that patients with synovial sarcoma need to be followed up for >10 years to assess for local recurrence and metastasis. Our case report confirms the soundness of this recommendation.

Conflicts of interest

The authors declare that there are no conflicts of interest.

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Further reading

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