Alveolar soft part sarcoma of the mandible: A rare case

Abstract

Alveolar soft part sarcoma (ASPS) is an enigma both for clinicians and pathologists. It is a rare soft tissue neoplasm affecting young adults. A 72-year-old female presented with a history of painful ulcer on the right side of the oral cavity associated with bleeding and difficulty in chewing. She underwent wide local excision of the lesion with segmental mandibulectomy and supraomohyoid neck dissection. Histopathological examination was suggestive of ASPS with involvement of resection margins. The patient developed rapid relapse after surgery and did not respond to chemotherapy and radiotherapy. She ultimately died of disease 10 months postsurgery.

Key words: Alveolar soft part sarcoma, chemotherapy, mandible, radiotherapy, surgery

INTRODUCTION

Alveolar soft part sarcoma (ASPS) is an extremely rare soft tissue sarcoma. It accounts for approximately 0.5–1% of all soft tissue sarcomas and occurs mainly in adolescents and young adults. ASPS occurs most commonly in soft tissues of lower extremities (44%) followed by head and neck region (27%), with the majority in orbital soft tissues and tongue. ASPS is a slow growing tumor with relatively indolent course and poor prognosis. It has a propensity for distant metastasis to lungs, brain, and bone. ASPS has been reported in various unusual locations in head and neck region, including, mandible, larynx, and paranasal sinuses. We report an unusual case of ASPS originating from the alveolar ridge of mandible in 72-year-old woman.

CASE REPORT

A 72-year-old woman presented with 3 months history of painful ulcer on the right side of the oral cavity associated with bleeding from ulcer and difficulty in chewing. There was no history of any addiction. On neck examination, a solitary right upper cervical lymph node measuring 1 cm × 1 cm was palpable. On examination, there was an ulcerative lesion on right alveolar ridge extending from lateral incisor till first molar and involving lower gingivobuccal sulcus. Laboratory examination comprising of hematology, liver, and renal functions were normal. Chest X-ray and ultrasound of abdomen and pelvis were normal. Orthopantomogram showed a lytic lesion involving the right side of the mandible. Preoperative biopsy was suggestive of sarcoma. The patient underwent wide local excision, segmental mandibulectomy, and supraomohyoid neck dissection. The mandibular reconstruction was done with iliac crest graft. Gross examination of the specimen revealed grey white tumor on the alveolar ridge measuring 3.5 cm × 2.3 cm × 1.2 cm. The tumor was soft to firm in consistency and eroded adjacent bone. Histopathological examination showed a relatively circumscribed tumor arranged in nests separated by thin fibrovascular septae [Figure 1a]. The tumor cells were round to polygonal with distinct cell borders, vesicular nuclei, abundant granular eosinophilic cytoplasm and low mitotic activity. Periodic acid-schiff (PAS) positive diastase resistant cytoplasmic inclusions were present focally. The tumor cells showed diffuse cytoplasmic positivity for vimentin and focal positivity for S-100 [Figure 1b] and were negative for chromogranin, smooth muscle actin, desmin and HMB-45. The posterior resection limit was involved by the tumor. Three out of 11 lymph nodes showed metastatic tumor deposits. The patient was advised postoperative radiotherapy in view of positive margins and lymph node metastasis but owing to poor compliance, she did not undergo adjuvant radiotherapy and came 4 months postsurgery with recurrence of disease at the primary
eosinophilic cytoplasm (b) tumor cells showing focal positivity for S-100

diseases, positive surgical margins, complex anatomic sites and
radiotherapy or chemotherapy has not yet been defined.

either survivable or metastasis free survival was detected. Large
tumor size (>5 cm), presence of metastasis at diagnosis and higher age
at diagnosis were identified as poor prognostic factors.[6,10] Long-term
follow-up is of paramount importance as 20% of the patients may
develop distant metastasis more than 10 years after initial surgery.[9]

CONCLUSION

Alveolar soft part sarcoma of the alveolar ridge of mandible is a
rare neoplasm. Complete surgical excision with negative pathologic
margins is the treatment of choice. The role of radiotherapy and
chemotherapy is not clearly defined.

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