Intraoral Rhabdomyosarcoma in a Young Boy

Aravinda Konidena, Manasi Kode

INTRODUCTION

Soft tissue sarcomas are malignant extraskeletal tumors of mesenchymal origin and uncommon in children. Rhabdomyosarcoma is most common and constitutes about 10 to 15% of the malignant neoplasms affecting children. The tumor is located with a high incidence in the head and neck region followed by the extremities and perineal region. We recently encountered one such interesting case of intraoral rhabdomyosarcoma and reporting the same with relevant review of literature.

CASE REPORT

A 5-year-old male patient presented with painful swelling in the right side of the lower jaw since two months. The swelling was growing slowly, painful and there was spontaneous exfoliation of the right mandibular deciduous 1st and 2nd molars. There was an associated discomfort in mastication and deglutition. Rest of the medical history was not contributory.

Facial examination revealed asymmetry due to a large, ill-defined swelling of 4 × 4 cm approximately extending partly below the inferior border of the mandible causing obliteration of the right nasolabial fold (Fig. 1). The swelling was firm, tender and diffuse. There were no extraoral draining sinuses, pigmentation or ulcerations. Right submandibular lymph nodes were firm, mobile and nontender. The whole complement of primary teeth except for the mandibular right molar teeth was present. All the permanent 1st molars were erupting. The remaining deciduous molars were carious so those were the maxillary central incisors. Intraoral examination revealed ulcerated fleshy, friable mass covering the mandibular ridge, measuring approximately 5 × 4 cm, extends from distal of deciduous canine to the ramus posteriorly (Fig. 2). The surface of the lesion showed indentations of the maxillary teeth. There was no pus discharge or sinus present. The lesion was firm and tender on palpation.

The right lateral oblique view of the mandible revealed irregular bony destruction along with resorption of the anterior border of ramus of the mandible. The orthopantamograph showed an irregular destruction of bone extending from distal of mandibular right canine to the ramus (Fig. 3). The area was devoid of sclerotic border, but the inferior border of the mandible was intact. Displacement of the right mandibular 1st and 2nd molar tooth buds was seen. Anatomy of the mandibular canal was indistinct in that region.

ABSTRACT

Rhabdomyosarcoma constitutes about 10 to 15% of the malignant neoplasms affecting children. Common sites of involvement include the head and neck, extremities and retroperitoneum. Intraoral rhabdomyosarcomas are rare and can affect tongue, palate, oral mucosa or gingiva. A case of rhabdomyosarcoma is being reported along with a review of literature.

Keywords: Rhabdomyosarcoma, Children, Head and neck.
Incisional biopsy was taken and H&E stained sections showed round, undifferentiated cells with hyperchromatic nuclei and clear vacuoles (Fig. 4). The nuclei were moderately large and ovoid. The chromatin was coarsely granular and the nucleoli were inconspicuous. Mitotic figures were rarely seen and the stroma was myxoid and rich in thick-walled vessels. Small number of cells had an eosinophilic cytoplasm with eccentric nuclei. The overall picture was suggestive of rhabdomyosarcoma. Surgical excision of the lesion was planned along with the extraction of permanent 1st and 2nd molars. However, the patient was lost to follow-up.

The presenting symptoms for rhabdomyosarcomas of the orofacial region include painful infiltrative growth of short duration, paresthesia, loss of teeth and trismus characterized by fast growth. Pain, proptosis, diplopia, strabismus, decreased hearing, nasal obstruction, dysphagia, cervical lymphadenopathy are other signs and symptoms. Pande S et al reported two cases of rhabdomyosarcoma mimicking gingival epulis associated with spontaneous exfoliation of teeth.

Staging of rhabdomyosarcoma according to intergroup rhabdomyosarcoma study: 1. Localized disease, tumor resected completely, regional lymph nodes not involved. 2. Localized disease with microscopic residual disease or regional disease with or without microscopic residual disease. 3. Incomplete resection with gross residual disease. 4. Metastatic disease.

Histologically, the major subtypes are embryonal, alveolar and pleomorphic. Embryonal rhabdomyosarcoma has hypercellular, spindled areas intermixed with myxoid, hypocellular areas. Botryoid type is included in embryonal type occurring in a hollow viscus. Embryonal rhabdomyosarcoma has a favorable prognosis. Alveolar variant has aggregates of round to oval neoplastic cells, separated by irregularly shaped fibrous trabeculae forming ill-defined alveolar spaces. Pleomorphic or classical variant is the adult type seen in adults over 40 years of age.

Immunohistochemically, specific markers for rhabdomyosarcoma are desmin and myogenin. There are contradictory reports about the value of DNA ploidy in the prediction of clinical response, histopathology, prognosis and the outcome of treatment. Kilpatrick et al by image analysis and flow cytometry conclude that DNA content is not predictive of prognosis. Important prognostic indicators of survival include extent of disease, distant metastases and age of the patient.

The best possible clinical outcome is frequently achieved via a multimodal approach. Sophisticated surgical techniques with reconstruction are used as a primary modality. Rhabdomyosarcoma is a radiotherapable lesion but results in arrest of craniofacial skeletal growth and the appearance of radiation induced secondary tumors. In addition, multiagent neoadjuvant chemotherapy, including vincristine, adriamycin, cyclophosphamide, and actinomycin D reduces the micrometastases and the malignant cell population and thus favors long time survival with fewer complications. Rhabdomyosarcomas are associated with high rates of recurrence and distant metastases through lymphatic and hematogenous spread to lungs, lymph nodes and bone marrow followed by heart, brain, meninges, pancreas, liver and kidney.

DISCUSSION

Rhabdomyosarcoma was first described in 1946 and they originate from the striated muscle and the common sites include extremities, head and neck, genitourinary tract and retroperitoneum. Rhabdomyosarcoma (RMS) arises from the immature mesenchymal precursor cells committed to skeletal muscle lineage or embryonal muscular tissue origin displaced during early development. WHO defined rhabdomyosarcoma as a highly malignant tumor of rhabdomyoblasts in varying stages of differentiation with or without cross-striation.

Rhabdomyosarcoma commonly affects children below 7 years of age. Some authors found bimodal age distribution, the first peak in children aged 2 to 6 years and the second peak in adolescents. The predominant sites include pharynx, orbit, oral cavity (soft palate, posterior mandibular region, cheek, lips, tongue) followed by paranasal sinuses, neck and ear. Rare cases were reported on gingiva, floor of the mouth, parotid, longstanding ameloblastoma. Rhabdomyosarcoma of head and neck is classified anatomically as parameningeal (including RMS of the nose, nasopharynx, paranasal sinuses, middle ear, mastoid, infratemporal fossa, and pterygopalatine fossa) and nonparameningeal (including RMS of the scalp, orbit, parotid gland, oral cavity, oropharynx, and larynx).

The presenting symptoms for rhabdomyosarcomas of the orofacial region include painful infiltrative growth of short duration, paresthesia, loss of teeth and trismus characterized by fast growth. Pain, proptosis, diplopia, strabismus, decreased hearing, nasal obstruction, dysphagia, cervical lymphadenopathy are other signs and symptoms. Pande S et al reported two cases of rhabdomyosarcoma mimicking gingival epulis associated with spontaneous exfoliation of teeth.

Staging of rhabdomyosarcoma according to intergroup rhabdomyosarcoma study: 1. Localized disease, tumor resected completely, regional lymph nodes not involved. 2. Localized disease with microscopic residual disease or regional disease with or without microscopic residual disease. 3. Incomplete resection with gross residual disease. 4. Metastatic disease.

Histologically, the major subtypes are embryonal, alveolar and pleomorphic. Embryonal rhabdomyosarcoma has hypercellular, spindled areas intermixed with myxoid, hypocellular areas. Botryoid type is included in embryonal type occurring in a hollow viscus. Embryonal rhabdomyosarcoma has a favorable prognosis. Alveolar variant has aggregates of round to oval neoplastic cells, separated by irregularly shaped fibrous trabeculae forming ill-defined alveolar spaces. Pleomorphic or classical variant is the adult type seen in adults over 40 years of age.

Immunohistochemically, specific markers for rhabdomyosarcoma are desmin and myogenin. There are contradictory reports about the value of DNA ploidy in the prediction of clinical response, histopathology, prognosis and the outcome of treatment. Kilpatrick et al by image analysis and flow cytometry conclude that DNA content is not predictive of prognosis. Important prognostic indicators of survival include extent of disease, distant metastases and age of the patient.

The best possible clinical outcome is frequently achieved via a multimodal approach. Sophisticated surgical techniques with reconstruction are used as a primary modality. Rhabdomyosarcoma is a radiotherapable lesion but results in arrest of craniofacial skeletal growth and the appearance of radiation induced secondary tumors. In addition, multiagent neoadjuvant chemotherapy, including vincristine, adriamycin, cyclophosphamide, and actinomycin D reduces the micrometastases and the malignant cell population and thus favors long time survival with fewer complications. Rhabdomyosarcomas are associated with high rates of recurrence and distant metastases through lymphatic and hematogenous spread to lungs, lymph nodes and bone marrow followed by heart, brain, meninges, pancreas, liver and kidney.
REFERENCES