ABSTRACT

Background: Sclerosing rhabdomyosarcoma is the rarest variant of rhabdomyosarcoma, accounting for 5-10% of all rhabdomyosarcoma cases. Pathologically, these tumors can provide a variety of features and can resemble other tumors, thus often difficult to diagnose. We report a case of sclerosing rhabdomyosarcoma of the gingiva with histomorphology mimicking a malignant peripheral nerve sheath tumor.

Case Presentation: The patient was a 39-years old Balinese male who noticed a lump on the left gum since 9 months ago. Head CT scan revealed a solid mass with soft tissue density at the left lower gingiva that extend superiorly and laterally, involving the left parapharyngeal and masticator spaces, infiltrated into the left mylohyoid muscle and left medial and lateral pterygoid muscles, and caused destruction of the left mandibular ramus and alveolar process of the left maxilla. The initial biopsy showed the proliferation of atypical spindle cells with a pattern resembling peripheral nerve sheath tumor and concluded as a low-grade malignant peripheral nerve sheath tumor. Wide excision specimen consists of proliferation of low-grade atypical spindle cells arranged in a fascicular pattern with foci of strap cells embedded in glassy eosinophilic sclerosis matrix. Some parts of the tumor showed palisading nuclei. The tumor cells were diffusely positive for desmin and negative for S100. This case was finally diagnosed as sclerosing rhabdomyosarcoma.

Conclusion: Sclerosing rhabdomyosarcoma should be considered when atypical spindle lesions occur between the hyalinized stromal matrix of the head and neck in adults. Because of wide differential diagnosis, an adequate sampling, careful microscopic examination, and immunohistochemistry staining can avoid misdiagnosis.

Keywords: Sclerosing Rhabdomyosarcoma, Sarcoma, Gingiva, Peripheral Nerve Sheath Tumor.


INTRODUCTION

Rhabdomyosarcoma is a malignant tumor originating from skeletal muscle cells. The latest World Health Organization (WHO) classification system classifies rhabdomyosarcoma into four groups: embryonal, alveolar, pleomorphic, and spindle cell/sclerosing rhabdomyosarcoma. Among all these types, the type most commonly found is embryonal rhabdomyosarcoma, with an incidence of 4.5 cases per 1 million in children aged < 15 years in the USA. Spindle cell/sclerosing rhabdomyosarcoma is the rarest variant of rhabdomyosarcoma, accounting for 5-10% of all rhabdomyosarcoma cases. It can affect children and adults and is more common in men.2,3

Sclerosing rhabdomyosarcoma has a variable morphology with characteristic spindle cells with a prominent hyalinizing matrix. Pathologically, these tumors can provide a variety of features and can resemble other tumors such as salivary gland tumors, fibrosarcoma, osteosarcoma, chondrosarcoma, angiosarcoma, spindle cell carcinoma, spindle cell melanoma, malignant peripheral nerve sheath tumor, and malignant triton tumor, so that it is often challenging to diagnose histopathologically, coupled with rare cases that cause the pathologist to be unfamiliar.2,4

Based on those mentioned above, this case study aims to evaluate the sclerosing rhabdomyosarcoma of the gingiva with histomorphology resembling a peripheral nerve sheath tumor.

CASE PRESENTATION

The patient is a male, 39 years old, Balinese, who noticed a lump on the left gum since 9 months ago. The lump grows to the palate and pushes to the left cheek, accompanied by pain when chewing without painful swallowing. The patient has had difficulty opening his mouth for the past 7 months. The patient had no previous medical or surgical history. On physical examination, vital signs were within normal limits.
There was a solid mass on the left superior gingiva, greyish-white, with a cross-section of 3 cm that extended to the left buccal (Figure 1). Other laboratory tests were within normal limits.

Non-enhanced and enhanced head CT scan revealed a solid mass with soft tissue density at the left lower gingiva that extend superiorly and laterally, involving the left parapharyngeal and masticator spaces, infiltrated into the left mylohyoid muscle and left medial and lateral pterygoid muscles, and caused destruction of the left mandibular ramus and alveolar process of the left maxilla. The mass revealed contrast enhancement after contrast administration, suggesting a malignant left gingival mass-submandibular lymph nodes enlargement, suspicious of lymph node metastases. Fluid density is noted in the right frontal, left sphenoid, and bilateral ethmoid and maxillary sinuses-left deviation of the nasal septum. Intracranial structures are unremarkable (Figure 2). Examination of the chest x-ray and ultrasound of the liver did not show any metastases.

Then the patient underwent an incisional biopsy. Microscopically, the biopsy specimen showed a proliferation of spindle cells with mild to moderate atypia, forming irregular fascicles with dense and loose areas. Some of them showed a nuclear palisading pattern, resembling peripheral nerves sheath tumors. Then it was concluded as an atypical neurofibroma with a differential diagnosis of low-grade malignant peripheral nerve sheath tumor (MPNST) (Figure 3).

Three months later, the patient underwent wide surgical excision of the tumor, left maxillectomy, left hemimandibulectomy, and removal of the sublingual and submandibular lymph nodes. Macroscopically, the tumor was 10.5 cm long, white to gray solid (Figure 4). Microscopically, the tumor mass consists of a proliferation of low-grade appearance spindle cells arranged in a fascicular pattern with oval to spindle hyperchromatic nuclei. Some strap cells can be identified (Figure 5A and 5B). Some of the tumor cells appeared rounded, arranged in cell nests between glassy eosinophilic sclerosis matrix (Figure 5C). Few scattered mitoses were also seen. It was also noticed that

**Figure 1.** (A) A bulging mass was seen in the left buccal region; (B) On intraoral examination, a reddish-white solid mass with necrotic, solid consistency appears on the left gingiva (arrowhead).

**Figure 2.** (A and B) Head CT scan showed a solid mass of the left lower gingiva that infiltrates left mylohyoid muscle; (C) The mass extends laterally and superiority into the left parapharyngeal and masticator space, infiltrates the left medial and lateral pterygoid muscles; and (D) The mass causes destruction of the left mandibular ramus (arrow) and alveolaris process of the left maxilla (arrowhead).
CASE REPORT

with a ratio of 6:1. Clinically, this tumor usually appears as a painless mass but can give local compression symptoms. In children, sclerosing rhabdomyosarcoma is more common in the paratesticular region. At the same time, in adults, it mainly occurs in the deep soft tissue in the head and neck area and can destroy surrounding organs, and can even invade intracranial tissue. Other locations are the urinary bladder, abdomen, retroperitoneum, trunk, and extremities. In our case, the tumor appeared at the left lower gingiva that extends superiorly and laterally, involving the left parapharyngeal and masticator spaces, infiltrated into the left mylohyoid muscle and left medial and lateral pterygoid muscles, and destroyed the left mandibular ramus and alveolar process of the left maxilla.

Figure 3. Microscopic appearance of the biopsy specimen. (A) Mild atypical spindle cells showed nuclear palisading (arrow) (HE, 100x); (B) On high power view, the tumor cells were spindle with elongated, wavy nuclei mimicking peripheral nerve tumor (head arrow) (HE, 400x).

Figure 4. Macroscopic view of the resected tumor. (A) The tumor encased mandible; (B) The tumor mass was grayish-white and had a solid inconsistency on the cut section.

the part of the tumor that formed a nuclear palisade pattern resembled the arrangement/pattern of growth of tumor cells originating from peripheral nerves, such as malignant peripheral nerve sheath tumor (MPNST) (Figure 5D). No evidence of tumor metastasis to the sublingual and submandibular lymph nodes microscopically.

Immunohistochemical stains applied with appropriate control and tumor cells showed strongly positive for desmin and negative for S-100 (Figure 6). Based on the morphological and immunohistochemical findings, this case was concluded as sclerosing rhabdomyosarcoma. After wide excision, the treatment is followed by radiotherapy.

DISCUSSION

Based on the latest WHO classification, sclerosing rhabdomyosarcoma is a particular entity other than embryonal rhabdomyosarcoma, alveolar rhabdomyosarcoma, and pleomorphic rhabdomyosarcoma. Sclerosing rhabdomyosarcoma is a rare type of rhabdomyosarcoma, accounting for only 5-10% of all rhabdomyosarcoma cases. These tumors can affect children and adults. In contrast to embryonal rhabdomyosarcoma and alveolar rhabdomyosarcoma, which are more common in children, pleomorphic rhabdomyosarcoma is more common in old age. Sclerosing rhabdomyosarcoma was more common in men than women.
rhabdomyosarcoma includes fibrosarcoma, osteosarcoma, chondrosarcoma, angiosarcoma, spindle cell carcinoma, spindle cell melanoma, malignant peripheral nerve sheath tumor, and malignant triton tumor.\(^4\) Immunohistochemistry is most helpful in differentiating sclerosing rhabdomyosarcoma from other malignancies because the other tumors do not give diffusely positive results for myogenin, Myo-D1, or desmin.\(^4,11,12\)

Although they can produce a positive stain, malignant triton tumour will be stained with focal positives in areas with rhabdomyoblastic differentiation only, not diffusely stained on all tumor cells.\(^4\)

In our case, we also found a proliferation of spindle cells that form a nuclear palisade arrangement, giving a malignant peripheral nerve sheath tumor appearance, resulting in a misdiagnosis during the initial biopsy. However, in the resected tissue, the histomorphology of sclerosing rhabdomyosarcoma became more apparent with extensive hyalinized stroma and strap cells foci. This is because, in the resection tissue, an adequate sample can be taken from the entire tumor area. In addition, the diagnosis was also confirmed by desmin immunohistochemistry examination, which showed a diffuse positive result in all tumor cells.

Rhabdomyosarcoma is generally treated with surgery, chemotherapy, and radiotherapy. Chemotherapeutic agents typically consist of actinomycin D, doxorubicin, ifosfamide, cyclophosphamide, etoposide, or vincristine.\(^4\) Despite various treatment techniques, the prognosis for sclerosing rhabdomyosarcoma remains poor, with recurrence and metastasis rates of around 40-50%.\(^1,13,14\)

If the patient survives, there are usually sequelae due to therapy such as facial asymmetry, trismus, dental abnormalities, and hyposalivation.\(^13\)

The limitation of this case report is that a complete immunohistochemical panel has not been carried out because of low resources. However, desmin and S-100 immunostains have been able to confirm the diagnosis.

**CONCLUSION**

Sclerosing rhabdomyosarcoma is a rare type of rhabdomyosarcoma. These tumors should be considered when atypical spindle lesions occur between the hyalinized stromal matrix of the head and neck in adults. Because of wide differential diagnosis, an adequate sampling, careful microscopic examination, and immunohistochemistry staining can avoid misdiagnosis.

**CONFLICT OF INTEREST**

There is no conflict of interest regarding the manuscript.

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**Figure 5.** Microscopic appearance of the resected specimen. (A) The resected specimen showed proliferation of low-grade appearance spindle cells arranged in fascicular pattern; (B) Strap cells with dense eosinophilic cytoplasm (arrows) are also seen (HE, 400x); (C) Rounded cells are embedded in a glassy eosinophilic matrix (HE, 400x); and (D) The part of the tumor cell that forms the nuclear palisading pattern—the pattern Mimicking Peripheral Nerve Origin (MPNST) (HE, 40x).

**Figure 6.** Desmin immunostaining. (A) The tumor cells were diffusely stained positive for Desmin (IHC, Desmin, 100x); (B) The tumor cells were negative for S-100. (IHC, S-100, 100x).
ETHICAL CONSIDERATION

This case report compliance with ethical standards was obtained with number 2297/UN14.2.2.VII.14/LT/2021 prior to the study being conducted.

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AUTHOR’S CONTRIBUTION

I Wayan Juli Sumadi contributed to the concept, design, definition of intellectual content, data analysis, preparation, editing, manuscript review, and guarantor. Dandy Citra helped in literature search, data acquisition, manuscript preparation, manuscript editing, and manuscript review. Elysanti Dwi Martadiani and I Nengah Wiadnyana Steven Christian helped define intellectual content, collecting clinical data, data analysis, manuscript editing, and manuscript review.

REFERENCES