J Korean Med Sci 1998; 13: 696-700
ISSN 1011-8934

Mesenchymal Chondrosarcoma of the Hyoid Bone
: A Case Report

Mesenchymal chondrosarcoma is a rare tumor that is distinctly different from classic chondrosarcoma. The prognosis of this tumor is poor, with a high incidence of locoregional and distant metastases. It shows a predilection for the head and neck, however mesenchymal chondrosarcoma of hyoid bone has rarely been reported. We experienced a case of mesenchymal chondrosarcoma of the hyoid bone in a 39-year-old woman. She underwent excision of the tumor by right hemihyoidectomy. Histologically, a combination of cellular zones composed of undifferentiated small cells and chondroid zones typically presented a bimorphic appearance. CD99 (DN16) immunohistochemical stain demonstrated that all undifferentiated small cells had strong reactivity with a distinct membranous pattern. There was microscopic tumor extension to the resection margin of the hyoid bone, however, no evidence of reccurence is noted at follow-up of 4 months with neck CT.

Key Words: *Chondrosarcoma, mesenchymal; Hyoid bone; Antigens, CD99*

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Received: May 4, 1998
Accepted: August 3, 1998

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INTRODUCTION

Since Lichtenstein's and Bernstein's description in 1959 (1), mesenchymal chondrosarcoma has been recognized as a rare tumor arising both in the bone and the soft tissues. It occurs predominantly in adolescents and young adults. This unusual malignancy is histologically characterized by areas of densely cellular and undifferentiated mesenchymal cells admixed with islands of immature hyaline cartilage. Approximately 10% of chondrosarcomas arise in the head and neck site, such as the skull base, maxilla, mandible, and larynx (2). Involvement of the hyoid bone is exceptional. In the literature, only 13 cases of primary chondrosarcomas of the hyoid bone have been reported (3). However, all of the reported cases (2-6) were classic or clear cell chondrosarcoma, and there have been few cases of mesenchymal chondrosarcoma arising in the hyoid bone.

CASE REPORT

A 39-year-old woman was seen at otorhinolaryngology clinic with a painless swelling in the right submandibular area for 4 months. On physical examination a 2×3 cm-sized, soft mobile mass was palpable in the right submandibular region. A neck computed tomograph (CT) revealed a mixed density mass with finely stippled calcification and irregular destruction of the right greater cornu of

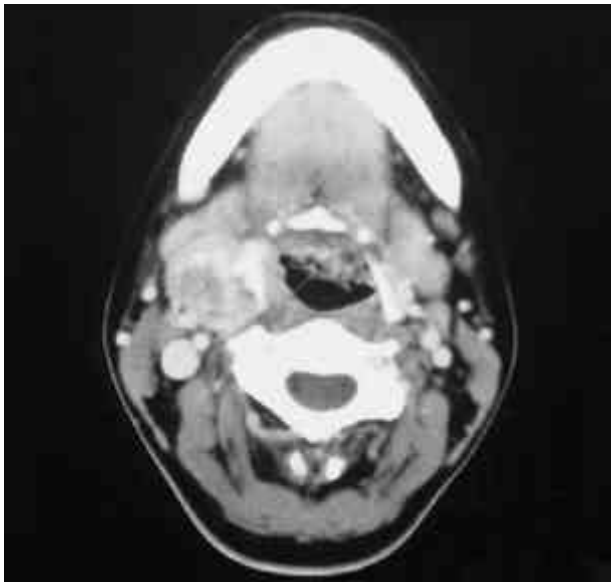

Fig. 1. A neck CT reveals a well-defined osteolytic bone tumor which appears to arise in the right greater cornu of the hyoid bone. It shows mixed density with fine stippled calcification with destruction of the hyoid bone.

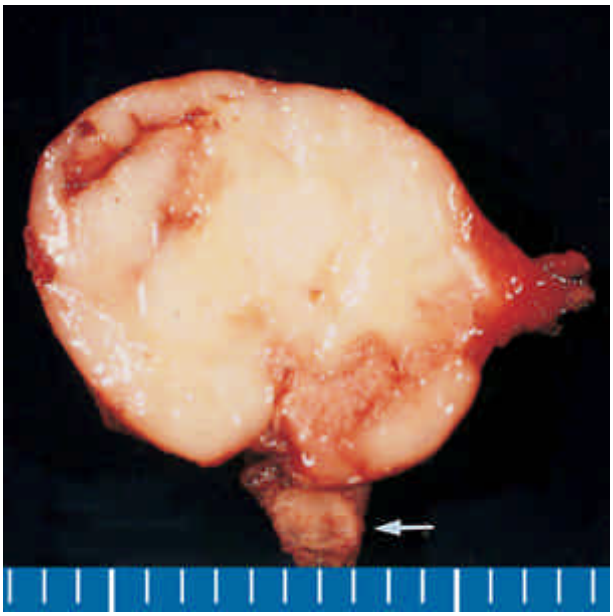

Fig. 2. Grossly, a well-circumscribed round soft mass, 2.2×2.0 ×2.0 cm, shows a thin fibrous capsule and pale brown homogenous cut surface. Multifocal hemorrhagic and calcific areas are also seen. A portion of partially resected hyoid bone is attached to the tumor (arrow).

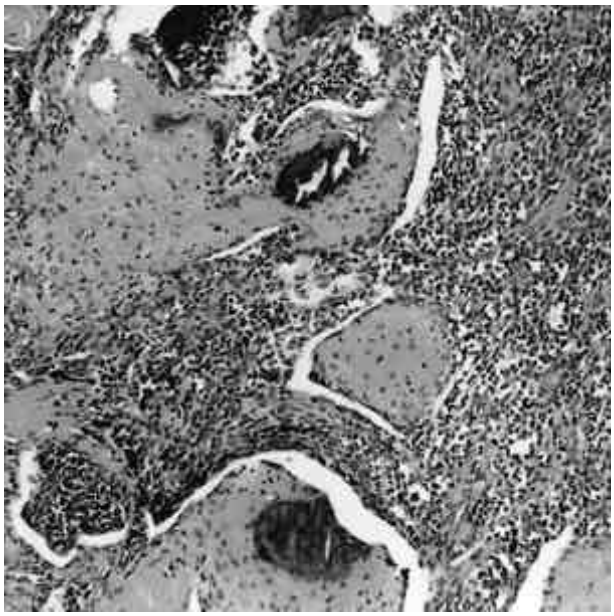

Fig. 3. Typical bimorphic pattern. Chondroid zone is surrounded by proliferation of undifferentiated cells with abrupt transition and ossification or calcification of the cartilaginous islands was occasionally encountered (H&E, ×100).

the hyoid bone (Fig. 1). A CT-guided fine needle aspiration biopsy obtained a 2.2×0.1 cm-sized, grayish-white tissue and microscopically represented nests of small undifferentiated tumor cells with staghorn-like vasculature, suggestive of hemangiopericytoma. No chondroid area was observed.

She underwent surgical excision of the tumor via a transcervical approach. An encapsulated tumor originated from the right greater cornu of the hyoid bone. The tumor including the right greater cornu and part of the hyoid bone, was resected. Grossly, the tumor was fairly well circumscribed and covered with a thin fibrous pseudocapsule. Cut surface of the tumor was pale gray and firm with multifocal calcified areas (Fig. 2).

Microscopically, the lesion was characterized by highly cellular proliferation of undifferentiated small cells, alternating with zones of differentiated cartilaginous area. The border between the two areas was clearly demarcated (Fig. 3). In the cellular areas, undifferentiated cells frequently were arranged in a vague alveolar pattern (Fig. 4A), or a herringbone pattern (Fig. 4B). Hemangiopericytoma-like pattern (Fig. 4C) was also found. Cytologically, the undifferentiated cells had round, elongated or spindle-shaped nuclei with dispersed chromatin, scanty cytoplasm and ill-defined border. The mitotic count was less than 4/10 high power fields (HPF) in the majority of the tumor. The degree of cellular pleomorphism was mild to moderate. The islands of hyaline cartilage were hypercellular and had features of low-grade to intermediate-grade chondrosarcoma. While ossification or calcification of the cartilaginous islands was occasionally encountered, lace-like deposits of osteoid with irregular appearance typically seen in osteosarcoma were not found. The tumor cells infiltrated to the adjacent skeletal muscle and the marrow space in the resection margin of the hyoid bone. Reticulin stain showed a well-developed and coarse reticulin network surrounding individual, or small clusters of undifferentiated cells. Immunohistochemistry showed diffuse weak vimentin expression and focal neuron specific enolase (NSE) expression. S-100 protein expression was limited to chondrocytes and chondroblasts within lacunae. Smooth muscle actins were also focally expressed in spindle-shaped tumor cells. Immunohistochemical staining revealed diffuse expression of CD99 (DN16) in small undifferentiated cells with variable intensity (Fig. 5) and the spindle cell areas tended to be more intensely expressed than the alveolar area. Ultrastructurally, tightly arranged tumor cells displayed a high nuclear/cytoplasmic ratio and a few primitive cell junction. The nuclei showed an euchromatin pattern with peripheral condensation and some medium-sized nucleoli. The cytoplasm was scanty, and revealed a moderate number of rough endoplasmic reticulum, a few mitochondria, and a large amount of glycogen particles (Fig. 6).

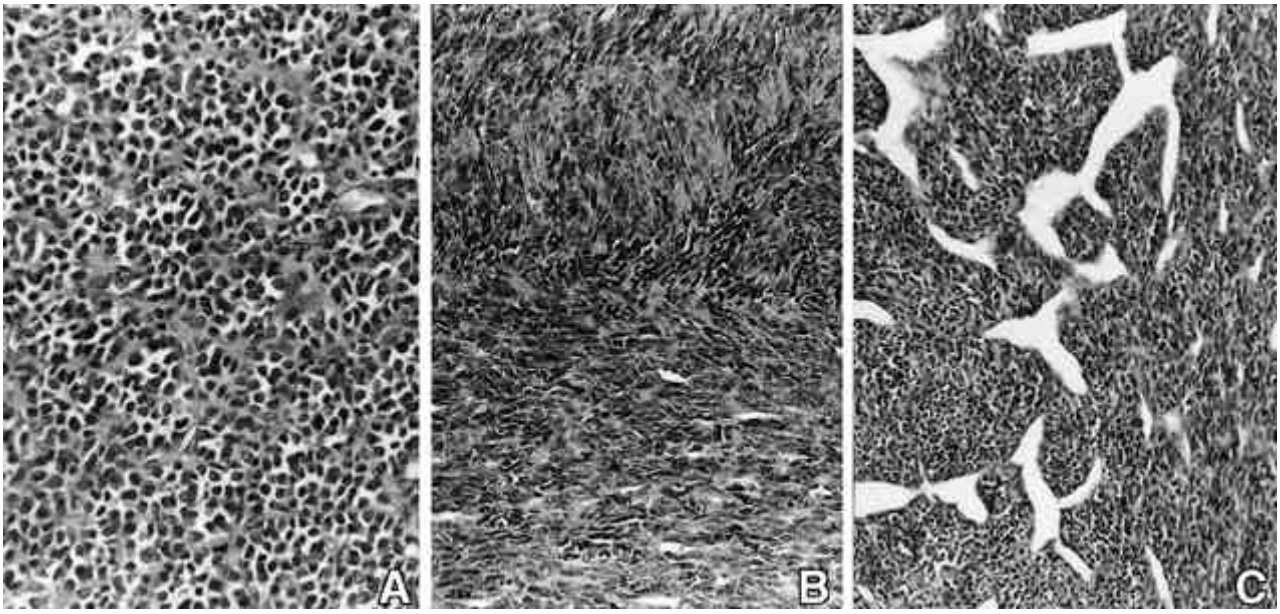

Fig. 4. Proliferation of small undifferentiated cells in a alveolar pattern (A) or a herringbone pattern (B) is prominent. And perivascular clustering of tumor cells and vascular proliferation with hemangiopericytomatous pattern is also seen (C).

Fig. 5. Immunohistochemical staining with CD99 is diffusely expressed in small undifferentiated cell. Spindle cell areas are more intensely expressed (×400).

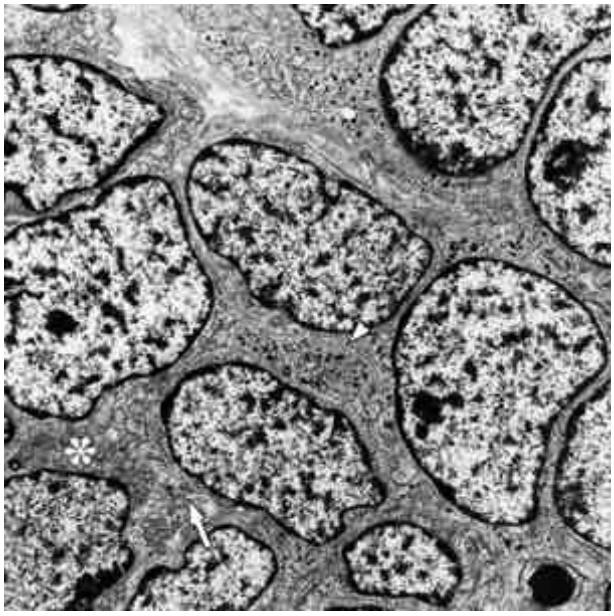

Fig. 6. Ultrastructurally, tightly arranged tumor cells show primitive, undifferentiated appearance with scanty cytoplasmic organelle including some mitochondria (asterisk), RER, glycogen particles (arrow head) and a few primitive intercellular junction (arrow) (×6,900).

DISCUSSION

Mesenchymal chondrosarcoma is a rare tumor that constitutes approximately 13% of chondrosarcoma of bone (7). The tumor has been described predominantly in the axial skeleton or cranial bones (8-10). The rib, vertebrae, pelvic bones, femur and craniofacial bones, including skull, were the most commonly involved sites (10). The hyoid bone is a rare location for chondrosarcoma to develop with only 13 previously reported cases and all

cases were classic or clear-cell chondrosarcoma (2-6).

In general, radiologic identification of mesenchymal chondrosarcoma is readily recognized as malignant, and usually the diagnosis of an ordinary chondrosarcoma is rendered (11). Clinical signs and symptoms are not characteristic, and pain as well as swelling or mass are the main symptoms in most cases (11).

In typical lesions, mesenchymal chondrosarcoma shows a paradoxical histologic combination of highly cellular zones composed of small undifferentiated cells and islands of low grade or even apparently benign chondroid substance, which may be calcified and even ossified. However, relative amounts of chondroid matrix and small cells differ from tumor to tumor, and small undifferentiated cell component may be identical with that of Ewing's sarcoma, primitive neuroectodermal tumor (PNET) or small cell osteosarcoma by light microscopy (12-15). Similarly, ultrastructural studies (14, 16) of the small cell component of mesenchymal chondrosarcoma cannot reliably distinguish Ewing's sarcoma and PNET or small cell osteosarcoma from mesenchymal chondrosarcoma. When overtly chondroid areas are inconspicuous, or are not represented in biopsy specimens, the histologic diagnosis of mesenchymal chondrosarcoma is difficult. Abundant periodic acid-Schiff (PAS)-positive cytoplasmic glycogen granules may make histologic differentiation between mesenchymal chondrosarcoma and Ewing's sarcoma (10). Prominent reticulin deposits encircling individual or small clusters of tumor cells are a more favorable feature of mesenchymal chondrosarcoma than small cell osteosarcoma (12). However, a search for other areas containing chondroid or osteoid matrix is important to solve the problem. In our case, multiple foci of cartilaginous islands were easily found and the chondroid areas were cytologically appeared to be low-grade to intermediate-grade chondrosarcoma. Even if the cartilaginous component is also present with the case of small cell osteosarcoma, it shows usually high grade. The previously described characteristic histologic patterns were an alveolar pattern, a hemangiopericytoma-like pattern and a herringbone pattern (8, 10, 12, 13). All three histologic patterns were intermingled in our case. In the spindle cell area, herringbone patterns and increased mitotic figures were evident.

The previously reported antigenic profiles of mesenchymal chondrosarcomas were vimentin positive, NSE focal positive, S-100 protein positive, and smooth muscle actin negative (12-14). S-100 protein expression in the chondroid area has been described to be most distinctive in this tumor (12, 13). Immunoreactivity to smooth muscle actin has been reported in a few studies (12, 13, 17).

Granter et al. (15) investigated the immunoreactivity of mesenchymal chondrosarcoma to CD99, which has been reported to be a sensitive and specific marker for Ewing's sarcoma and PNET, and found reactivity in 11 out of 11 cases (100%). We performed CD99 immunohistochemical staining. All undifferentiated small cells showed strong reactivity with a distinct membranous pattern; however, the cartilaginous areas showed no expression. The results may correspond to a theory that mesenchymal chondrosarcoma may be related to Ewing's sarcoma and PNET (17).

It is very difficult to differentiate this tumor from other small round cell tumor ultrastructurally except for specialized chondroid or osteoid area. However, if abundant amount of cytoplasmic organelle, mainly RER and Golgi apparatus and flocculent dense material within the dilated cistern or abundant glycogen pool and neuronal differentiation such as microtubules or neurosecretory granules are present, these findings favor small cell osteosarcoma or Ewing's sarcoma and PNET (16). In our case, although a fairly large amount of glycogen particles were encountered in some tumor cells, the majority of the tumor cells showed primitive, undifferentiated appearance with poorly developed, scanty cytoplasmic organelle. Definite neuronal differentiation or osteoid-like material was not seen.

Surgical excision is the treatment of choice for mesenchymal chondrosarcoma and complete resectability is one of the important prognostic factors (2, 5, 9-11). Irradiation or chemotherapy, traditionally used for Ewing's sarcoma, may be indicated for lesions which are not amendable to ablative surgical treatment (9, 11). In our case, there was no gross evidence of residual tumor at the time of surgery. However, unfortunately, there was a focal microscopic tumor that extended to the marrow space at the resection margin. However, she refused adjuvant chemotherapy or radiotherapy. At postoperative follow-up of 4 months, no evidence of reccurence is noted in follow-up neck CT.

The prognosis of mesenchymal chondrosarcoma is fairly unpredictable. Local recurrence or metastasis can occur after more than 20 years. Mayo Clinic reported a 5-year survival rate of 54.6% and a 10-year survival rate of 27.3% (10).

In this report we present a very rare case of mesenchymal chondrosarcoma of the hyoid bone in 39-year-old woman.

Acknowledgement

The monoclonal antibody for CD99 (DN16) was kindly donated by the immunologic laboratory of Dr. Seong Hoe Park (Professor, Department of Pathology, Seoul National University College of Medicine). We are grateful to him for the assistance.

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