Neurilemmoma is a benign neoplasm that originates from the Schwann cells that cover the peripheral nerves. This tumor presents as a painless, slow-growing mass and it may develop at any age. Neurilemmomas are most frequently located in the soft tissues of the head and neck, but they are also encountered on the flexor surfaces of the upper and lower extremities. Intraosseous neurilemmoma is extremely rare and fewer than 200 examples of this entity have been described in the literature. It accounts for less than 0.2% of all primary bone tumors.

We present here a rare case of intraosseous neurilemmoma of the mandible in a patient who was substantially older than the average neurilemmoma patient and the tumor occurred more anterior than its usual location in the mandible.

**CASE REPORT**

A 77-year-old woman presented with a painful swelling in the right chin, which had evolved over approximately 3 years and recently become aggravated. A clinical examination revealed an asymmetrical facial appearance due to swelling of the right chin. No skin color change or sensation of heat was evident. The patient did not complain of facial numbness, but she did complain of paresthesia of the right lower lip and chin area. She wore complete dentures on both sides of the jaw. Moreover, although she complained of tenderness on intraoral palpation of the right lower vestibule, there was no pus discharge. Panoramic radiography showed a \(3 \times 2\) cm-sized well-delimited radiolucent lesion in the anterior region of the right mandibular body (Fig. 1A). A block CT scan revealed an expansile, well-defined, unilocular lesion with thinning of the cortex. The lesion was surgically removed and it was found to be a \(2 \times 1.7\) cm-sized, bright yellowish, hard mass with hemorrhage and cyst formation. Histologically, the mass was a moderately cellular neoplasm and it showed distinct nuclear palisading, numerous Verocay bodies and tumor cells that were positively immunohisto stained for S-100 protein. Two months after the operation, the patient has remained in a good condition with no signs or symptoms of tumor recurrence.

**Key Words:** Mandibular neoplasms; Neurilemmoma; Bone neoplasms
tical bone was excised. The lesion was found to be a $2 \times 1.7$ cm sized, well-defined, hard mass with a bright yellowish cut surface. There was no evidence of bone invasion. Microscopically, the tumor was a uninodular mass that was limited by the cortical bone (Fig. 2A), and the tumor tissues showed alternating Antoni A and B regions. Vascular congestion, hemorrhage and cyst formation were also present in the mass’s periphery (Fig. 2B). The tumor was characterized by compact spindle cells that

Fig. 1. (A) Panoramic radiograph shows a well-defined, radiolucent lesion in the anterior region of the right mandibular body. (B) Block CT scan reveals an expansile, well-defined, unilocular lesion with thinning of the buccal and lingual cortical plates of the right mandibular body.

Fig. 2. (A) In low power view, the tumor is limited by a cortical bone and is composed of alternating Antoni A and Antoni B areas. (B) Vascular congestion, hemorrhage, and cyst formation are observed in the mass periphery. (C) Tumor cells are arranged in short bundles or interlaced fascicles and show typical nuclear palisading to form Verocay bodies. (D) Tumor cells showed immunopositivity for S-100 protein.
were arranged in short bundles or as interlaced fascicles. In addition, the tumor cells had twisted nuclei and indistinct cytoplasmic borders, and some of these nuclei formed typical palisades around an acellular eosinophilic area (Verocay bodies) (Fig. 2C). Mitotic figures and necrosis were not observed. The tumor cells were immunopositive for S-100 protein (Fig. 2D) but immunonegative for smooth muscle actin and CD34. The histopathologic diagnosis was intraosseous neurilemmoma. Two months after the operation, the patient has remained in good condition with no signs or symptoms of tumor recurrence.

**DISCUSSION**

Neurilemmoma is a benign neoplasm that originates from Schwann cells of the neural sheath. Intraosseous neurilemmoma is a rare tumor and it accounts for less than 0.2% of all primary bone tumors. However, the mandible is the most common site when encountering an intraosseous neurilemmoma. In the mandible, the posterior segments of the body and ramus are the most frequent sites of occurrence because of the protracted intraosseous path of the inferior alveolar nerve. The present case is somewhat less common because of the tumor’s anterior location. Neurilemmoma may involve bone via three mechanisms: 1) it may arise intramedullary within bone, 2) it may arise within the nutrient canal and cause canal enlargement, or 3) it may initially arise as a soft tissue or periosteal tumor and later penetrate bone. The last two occurrences are most frequent. In this described case, we were able to suggest the second mechanism because the proximal part of the right mental nerve had adhered to the mass and we exclude the third mechanism because no cortical bone destruction had occurred.

Clinically, neurilemmoma is a slow-growing tumor and it may be present for years before becoming symptomatic. Chi et al. has reported that swelling is the most common symptom of intraosseous neurilemmoma involving the mandible, but pain or paresthesia is present in about 37% and 11% of the cases respectively. It has shown a slight female predilection, with a 1.5:1 female-to-male ratio and 82% of the patients are younger than 50 years of age with the peak prevalence in the second and third decades of life. Furthermore, it occurs more frequently in the posterior body/ascending ramus than in the anterior body, with a ratio of 2.5:1. Radiographically, neurilemmomas may be either uniloculated or multiloculated, and they invariably produce well-defined radiolucencies in the posterior mandible, which may resemble many benign processes such as odontogenic keratocysts, periodontal cysts or ameloblastoma. In the present case, the possibility of neurilemmoma was not considered during the first radiologic study because of its less common intraosseous location in the anterior region of the mandibular body. Histopathologically, neurilemmoma provides a characteristic alternation of two types of tissue arrangement, i.e., Antoni A and Antoni B. The Antoni A areas are relatively cellular, and when they are more differentiated, they may exhibit nuclear palisading, whorling and Verocay bodies. In contrast, the Antoni B areas are less cellular and less organized, and they often contain prominent thickened blood vessels. In addition to classic neurilemmoma, there are several histopathologic variants that include the cellular, plexiform, epithelioid, ancient, and melanotic types. Microscopically, intraosseous neurilemmoma should be differentiated from desmoplastic fibroma, well-differentiated fibrosarcoma, fibrous dysplasia and nonossifying fibroma. Diffuse immunoreactivity for S-100 protein is routinely observed in neurilemmomas, but this is not seen in other lesions. When attempting to differentiate neurilemmoma from malignant spindle cell neoplasm, it is important to remember that atypia may be visualized as a degenerative change, but atypical mitoses are never present in benign neurilemmoma. Fortunately, malignant transformation of neurilemmoma is exceedingly rare, and no such transformation has been reported for intraosseous neurilemmoma.

Because it is a well-encapsulated lesion, the treatment of choice for neurilemmomas is conservative surgical enucleation with periodic follow-up. Recurrence is uncommon and in the present case, the patient has been followed up for two months with no clinical or radiographic signs of recurrence.

We present here a rare case of intraosseous neurilemmoma of the mandible. The majority of such tumors reported in the mandible have involved a posterior location, so our described case is somewhat unusual because of its anterior location and the patient had an older age at onset.

**REFERENCES**