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Case Report
Clinical Pathology

Recurrent adamantinoma of the mandible

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Abstract. Adamantinoma is an extremely rare tumour originating from bone that can be divided into classical and osteofibrous dysplasia (OFD)-like adamantinoma. This low-grade malignancy almost exclusively occurs in long bones, and only few cases of mandibular adamantinoma have been reported. Here, we report the case of a 30-year-old female with a 2-year history of right mandible tenderness. Radiological examinations showed a lytic lesion involving the right mandible without a well-defined margin. Biopsy confirmed the diagnosis of adamantinoma. She underwent a segmental mandibulectomy and reconstruction with a fibula flap. The definitive diagnosis was OFD-like adamantinoma. However, the tumour recurred after 5 years. The residual mandible and fibula flap were widely involved. A total mandibulectomy was performed. Five years later, there is no evidence of recurrence or metastasis. We recommend that adamantinoma be treated by radical resection with clear margins, and long-term surveillance is necessary due to the likely high local recurrence rate and the potential for late tumour metastasis.

Key words: adamantinoma; mandible; low-grade malignant neoplasm; recurrent; long-term follow-up.

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Adamantinoma is a rare low-grade malignant bone neoplasm that occurs nearly exclusively in the long bones and in patients in the second to fifth decades of life[1]. Adamantinoma tumour cells are characterized by a biphasic differentiated nature. The tumour is composed of epithelial cells in a fibrous stroma, which demonstrates various patterns. It is categorized into two types: classical adamantinoma and osteofibrous dysplasia (OFD)-like adamantinoma. The tumour mostly involves the long bones[2], and very few cases of adamantinoma occurring in the head and neck region have been reported. The purpose of this report is to present a rare case of mandibular OFD-like adamantinoma.

Case report

A 30-year-old female presented in June 2010 with a 2-year history of right mandible tenderness. The pain had worsened over the last 1 month. She reported no prior trauma. Physical examination revealed mild expansion on the lingual side of the right mandible with tenderness. Radiological examinations showed a $4.3 \times 2.7 \times 1.4$ cm lytic lesion involving the inferior side of the right mandible without a well-defined margin (Fig. 1a, b). The dental roots, soft tissue, and cervical lymph nodes were not involved. A biopsy sample showed that the gross tumour appearance was greyish white and solid. The pathological diagnosis was adamantinoma.

On the basis of the clinical and radiological findings, a segmental mandibulectomy was performed with approximately 1.5-cm surgical margins. The bone margins were clear histopathologically. The mandible was reconstructed with a fibula free flap on the right side (Fig. 1c). Histologically, the tumour showed fibrous proliferation with epithelial cells in strands and irregular bone trabeculae. The tumour cells were immunohistochemically reactive for cytokeratin (CK) and vimentin, but were non-reactive for epithelial membrane antigen (Fig. 2). The pathological diagnosis was OFD-like adamantinoma.

The patient maintained routine follow-up. In order to improve the appearance of

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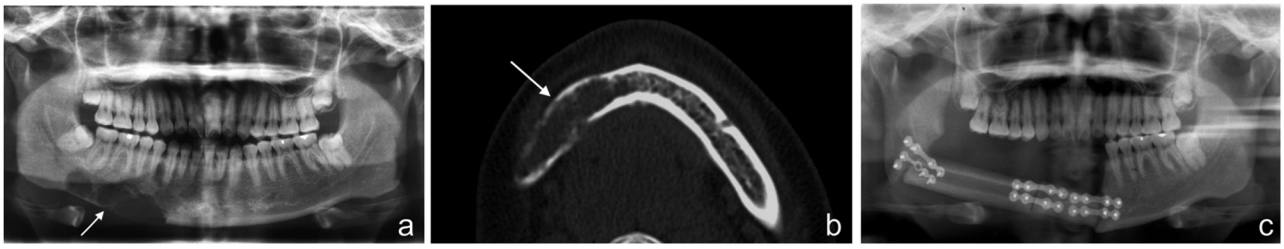

Fig. 1. Radiological manifestations of the primary osteofibrous dysplasia-like adamantinoma in the mandible and the outcomes of treatment. (a) Panoramic radiograph showing a lytic lesion in the right mandible. (b) Computed tomography showing the expansive and osteolytic appearance of the lesion, with cortical destruction. (c) Panoramic radiograph showing the extent of the segmental mandibulectomy and the fibula flap reconstruction.

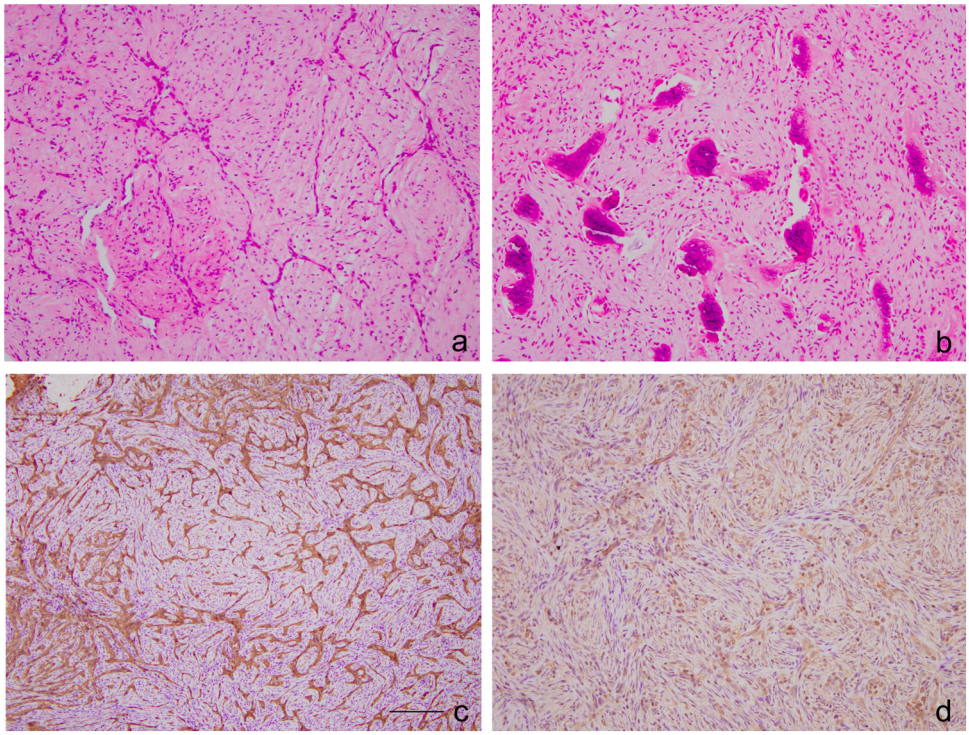

Fig. 2. Pathological features of the mandibular osteofibrous dysplasia-like adamantinoma from the resected specimen. (a) Cords and nests of epithelial cells juxtaposed with fibrous stroma (haematoxylin–eosin stain). (b) Areas that resemble osteofibrous dysplasia containing irregular bone spicules (haematoxylin–eosin stain). (c) The tumour cells showed positivity for cytokeratin. (d) The tumour cells showed positivity for vimentin.

the facial lower third, the mandible was augmented with a cortical non-vascularized iliac bone graft in May 2013. Further surgery was performed to improve the symmetry of the lower border of the mandible in April 2014. However, in December 2015, she complained of numbness of the left lower lip of 1-month duration. Physical examination showed mild expansion of the left mandible without limited mouth opening. Panoramic radiography and computed tomography (CT) scans demonstrated multifocal lytic lesions, which involved the body and ramus of the left mandible, the ramus of the right mandible, and even the fibula flap (Fig. 3a–c). The surrounding soft tissue and cervical nodes were not involved. A chest X-ray revealed no lung metastasis. A CT scan of the lower limbs showed no abnormalities. Biopsy confirmed recurrent adamantinoma.

A total mandibulectomy was performed to encompass the entire recurrent tumour, and the surgical defect was restored with a new microvascular fibula flap on the left side (Fig. 3d). Microscopically, the histological characteristics of the recurrent tumour were the same as those of the primary tumour, and the Ki67 level was 7–8%. The pathological diagnosis was recurrent OFD-like adamantinoma. The patient was followed regularly, and at 5 years after surgery, no recurrence or metastasis was evident.

Discussion

Adamantinoma is an extremely rare, low-grade malignant primary bone tumour that accounts for less than 1% of all primary bone neoplasms. The tumour was named ‘adamantinoma’ by Fischer in 1913, because it resembled ameloblastoma histologically[3]. Adamantinoma is microscopically characterized by a background of spindle-cell stroma containing islands of epithelial cells that show varying types and proportions. It commonly occurs in the long tubular bones, particularly in the tibia. Other sites include the humerus, ulna, femur, fibula, radius, and rib[2]. Reports of mandibular adamantinoma in the literature appear to be extremely rare. A single report by Dini et al. was identified, relating the case of a 55-year-old man with a cervicothoracic spinal adamantinoma and a mandibular adamantinoma[4]. In that case report, however, it was reported that the mandibular

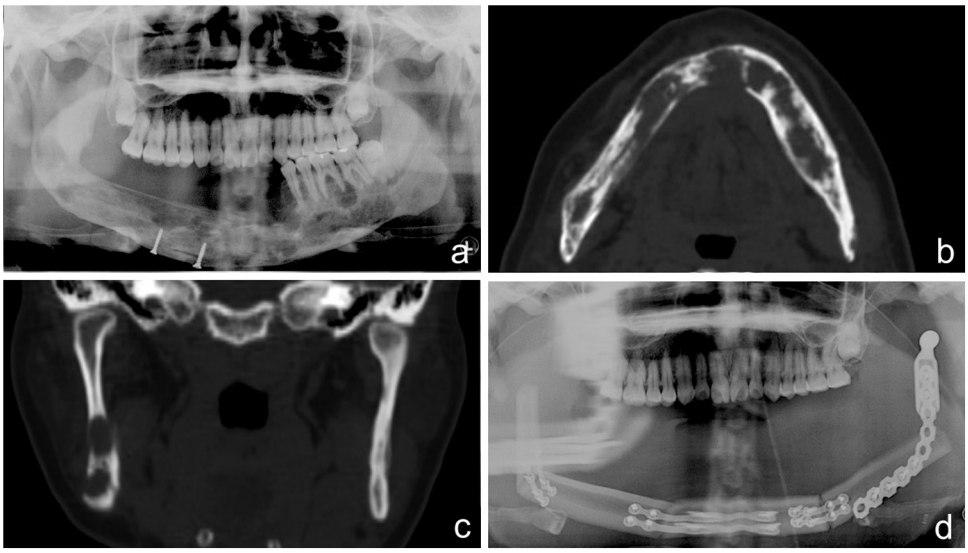

Fig. 3. Radiological manifestations of the recurrent osteofibrous dysplasia-like adamantinoma in the mandible and the outcomes of treatment. (a) Panoramic radiograph demonstrating multifocal lytic lesions without clear margins. (b) Computed tomography showing lytic lesions involving the left mandible and the fibula flap. (c) Computed tomography demonstrating lytic lesions involving the right mandible. (d) Panoramic radiograph showing the total mandibulectomy and fibula flap reconstruction.

adamantinoma was diagnosed by biopsy, and the patient refused further intervention. Thus, no detailed information was provided regarding the mandibular lesion.

There are two types of adamantinoma: classical adamantinoma and differentiated adamantinoma, which is also called osteofibrous dysplasia (OFD)-like adamantinoma. As a subtype, OFD-like adamantinoma was initially identified in 1989 due to its histological differences from the previously recognized classical type. The histopathological feature of differentiated adamantinoma is the predominance of OFD-like stroma with some small nests of epithelial cells. Studies suggest that OFD, OFD-like adamantinoma, and classical adamantinoma are likely related entities and lie along a spectrum of disease, with benign OFD at one end of the spectrum, malignant classical adamantinoma at the other end, and OFD-like adamantinoma in the middle[5].

Patients with adamantinoma of the long bones often have symptoms of pain and slow-growing swelling, and they may have a history of trauma at the affected site[6]. In this case, the patient also presented with numbness of the lower lip, which indicated tumour recurrence. Radiologically, adamantinoma shows an eccentric, expansive, and osteolytic appearance, with moth-eaten margins, skip lesions, and cortical destruction being described frequently[7]. Magnetic resonance imaging is useful for providing information on tumour-free margins and intramedullary extension[8].

As a low-grade malignant tumour, classical adamantinoma of the long bones needs to be treated by radical resection with wide margins, followed by reconstruction[9]. There is little evidence showing that radiotherapy and chemotherapy are effective[6]. For OFD-like adamantinoma, no guidelines can be found due to the scarcity of cases. Some authors recommend observation for small lesions or for young patients, which is similar to the treatment for OFD, but recommend surgical treatment for extensive or symptomatic lesions[2,10]. In the case presented here, a segmental mandibulectomy was performed. The tumour was resected macroscopically with 1.5-cm margins, and the bone margins were clear histopathologically. Afterwards, plate and screw removal, bone augmentation with an iliac bone graft, and other surgeries were performed on the mandible, but we believe that these operations had little to do with tumour recurrence. However, the tumour recurred, and multiple lesions were seen in the ramus and body of the mandible, even in the bone graft, which indicates that the behaviour of OFD-like adamantinoma might be more aggressive than previously thought. This suggests that observation or conservative treatment for OFD-like adamantinoma is not safe or suitable, and radical resection with wide margins might be the best choice to prevent tumour recurrence.

Adamantinoma has a high local recurrence rate and has the potential to metastasize. Houdek et al. performed a retrospective study of 46 patients with adamantinoma of the extremities who were treated surgically, and the local recurrence rate was 15%[9]. Qureshi et al. reviewed 70 patients with adamantinoma of the long bones who received surgery, and the local recurrence rate was 18.6%[6]. Furthermore, Scholfield et al. reported that three of 12 patients with OFD-like adamantinoma experienced local recurrence after surgical treatment[10]. Moreover, a recent long-term follow-up study demonstrated that metastasis occurred in 27% of patients, and the most common site was the lung[9]. In this previous study, the mean time to local recurrence was 8 years and the mean time to metastasis was 7 years, and it was reported that even distant disease in patients could occur more than 20 years after surgical treatment and could lead to death[9]. Houdek et al. also reported that the overall disease-specific survival rate decreased from 92% (10-year) to 53% (30-year)[9].

It is imperative to follow patients with adamantinoma regularly for a lifetime after surgery. For the mandibular case presented herein, the tumour recurred 5 years after the first operation despite clear resection margins. The patient then received a total mandibulectomy and continued follow-up for 5 years. Even though no recurrence or metastasis is evident now, there is no certainty that she is free from disease, and she will need surveillance for life.

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Competing interests

None.

Ethical approval

Not required.

Patient consent

Written informed consent was obtained from the patient.

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