Abstract: The aneurysmal bone cyst (ABC) rarely occurs in the jaws. It represents approximately 1.5% of all non-odontogenic and non-epithelial cysts of the jaws. The literature contains conflicting reports on the clinical and radiological features of ABC of the jaws. The radiographic appearance of ABC varies from a unicystic radiolucent or moth-eaten radioluency to an extensive multilocular lesion. In this article, we describe the transition of an ABC in the maxillofacial region from a unilocular radiolucent lesion to a radiopaque lesion in a 40-year-old female over a 10-month period, which indicates diversity in the clinical and biologic behavior of ABCs. (J Oral Sci 53, 529-532, 2011)

Keywords: aneurysmal bone cyst; mandible; radiolucent; radiopaque.

Introduction
Aneurysmal bone cyst (ABC) was first recognized as a distinct entity in 1942 when Jaffe and Lichenstein (1) described two cases of a “peculiar blood containing cyst of large size”. Later, Bernier and Bhaskar (2) first reported a case occurring in the jaws in 1958. The World Health Organization defines aneurysmal bone cyst as “a benign tumor-like lesion with an expanding osteolytic lesion, consisting of blood-filled spaces of varying size separated by connective tissue septae containing trabeculae or osteoid tissue and osteoclast giant cells” (3). The term “aneurysmal” is used to define the blown-out distension of part of the contour of the affected bone and “bone cyst” to indicate that when the lesion enters through a thin shell of bone, it appears largely as a blood-filled cavity.

ABCs occur most frequently in the long bones and vertebral column and only 2% of the lesions appear in jaws. The radiographic features of ABCs affecting the jaws are not pathognomic. They vary from a unilocular radioluency to a “ballooned out” multilocular radioluency with a honeycomb or soap-bubble appearance. This article reports an unusual radiographic presentation of ABC of the mandible with transition from a radiolucent to radiopaque lesion in the same individual.

Case Report
A 40-year-old female reported to the Department of Oral Medicine and Radiology, with the chief complaint of swelling on the right angle of the mandible and mild and intermittent pain for 1 month. Extra-oral examination revealed a well-defined, tender and bony hard swelling at the right angle of mandible with normal overlying skin (Fig. 1). The patient’s medical and family history was
unremarkable and there was no history of trauma. No evidence of swelling was present intra-orally. However, tooth 48 showed Grade I mobility.

A panoramic radiograph revealed a single unilocular lesion with well-defined borders anteriorly and inferiorly, extending from the periapical area of distal root of 47 to the angle of mandible posteriorly, superiorly extending from the cervical one third of distal root of 48 until the inferior border of mandible, which was not visible (Fig. 2). On CT examination, a well-defined expansile lytic soft tissue lesion was seen at the right angle and ramus of mandible with thinning of cortex and loss of continuity at many places and extension into the surrounding soft tissues (Fig. 3). A diagnosis of ameloblastoma of the right angle and ramus of mandible was made.

The patient was advised to undergo an incisional biopsy but she failed to report for the scheduled procedure. Ten months after her initial visit, the patient reported again with almost similar clinical presentation. The mobile tooth 48 had exfoliated. A panoramic radiograph (Fig. 4) showed Grade I mobility. Figure 2 shows a panoramic radiograph revealing a single unilocular lesion at the right angle of the mandible, whereas Figure 4 shows a well-defined expansile lytic soft tissue lesion at the right angle and ramus of mandible with thinning of the bony cortex. Figure 3 and Figure 5 depict axial CT scans showing a well-defined expansile lytic soft tissue lesion and a well-defined non-enhancing expansile lesion at the right angle and ramus of mandible, respectively, with thinning of the bony cortex.
4) taken this time showed a single predominantly radiopaque round lesion with a regular and distinct border blending with the normal bone anteriorly and superiorly. Inferiorly, the cortex of mandible was lost. Internal structure was predominantly radiopaque with dense bone formation antero-superiorly and expansion of jaw bone in an inferior direction.

CT mandible revealed a well-defined non-enhancing expansile lesion at the right angle and ramus of mandible with thinning of bony cortex and irregular breaks at many places and extension into the surrounding soft tissue (Fig. 5). The provisional diagnosis was cemento-ossifying fibroma. Fibrous dysplasia and aneurysmal bone cyst were included as differential diagnosis. The patient was referred to the Department of Oral Surgery where an enbloc resection distal to 45 until the base of right coronoid process and condyle of mandible was performed. The histopathological examination showed presence of many dilated vessels, containing blood separated by thick fibrous septae with osteoclastic giant cells, at places showing osteoid consistent with the diagnosis of aneurysmal bone cyst (Figs. 6 and 7).

**Discussion**

Aneurysmal bone cyst is usually considered to be a reactive lesion of the bone rather than a cyst or true neoplasm. Most believe that ABC is the result of a vascular malformation within the bone. The cause of the malformation is however a topic of controversy. It is not clear whether the lesion is primary or occurs in a pre-existing bone lesion (1).

Hillerup and Hjorting-Hansen proposed the theory that ABC, central giant cell granuloma and simple bone cyst arise from an intramedullary haematoma that may be the causative factor for the development of ABC (4). Hemodynamic disorders and arteriovenous malformations are thought to increase intraosseous venous pressure with expansion of the vascular tissue bed, leading to bone resorption and resulting in the “cystic” appearance on the radiograph (5). Chromosomal alterations of segments 17p and 16q have also been described. Panoutsakopoulos et al. described three cases of ABC with chromosomal abnormalities; band 16q22 being involved in all three patients (6).

Eighty percent of the ABCs occur in patients below 20 years of age with no gender predilection. However, studies have claimed a slight female preponderance. ABC is most common in those regions of the skeleton where there is a relatively high venous pressure and high marrow content (7). This explains its rare occurrence in the skull bones in which there is low venous pressure. However, when present, the mandible is most commonly affected (mandible-maxilla ratio 3:1), with a higher predilection for molar and ramus region.

ABC is extremely variable in its clinical presentation, ranging from a small indolent asymptomatic lesion to a rapidly growing, expansile, destructive lesion causing pain, swelling, deformity, neurological symptoms, pathologic fracture and perforation of the cortex. Radiographically, aneurysmal bone cyst may appear as a unilocular or multilocular radiolucency with expansion and thinning of the surrounding cortical bone. It has also been described as having a honeycomb or soap bubble appearance since it may be traversed by thin bony septa. However, there is no pathognomonic radiographic appearance for ABC. The periphery is usually well-defined and the shape is circular or “hydraulic”. After an ABC becomes large, there is propensity for extreme expansion of the cortical plates and it can displace or
A characteristic radiographic feature of ABC is the “ballooning” distension of periosteum with a thin outline of reactive, subperiosteal bone. The radiographic picture at the second visit of this case also favoured the ballooned or distended or blown-out appearance of ABC. Most cases in the jaws reported in the literature were radiolucent except for one case which was radiopaque.

The aneurysmal bone cyst has been reported to evolve through four radiologic stages: initial, active growth, stabilization, and healing (10). In the initial phase, the lesion is characterized by a well-defined area of osteolysis with discrete elevation of the periosteum. This is followed by a growth phase, in which the lesion grows rapidly with progressive destruction of bone and development of the characteristic “blown-out” radiologic appearance. The growth phase is succeeded by a period of stabilization, in which the characteristic “soap bubble appearance” develops, as a result of maturation of the bony shell. Final healing results in progressive calcification and ossification, with the lesion transforming into a dense bony mass. Our case is unique as it represents the rapid evolution of a unilocular radiolucent lesion to a radiopaque lesion in the same individual over a period of 10 months.

Microscopically, numerous cavernous, sinusoidal spaces filled with blood are surrounded by loose, fibrous connective tissue. The connective tissue septa contain small capillaries, multinucleated giant cells, inflammatory cells, extravasated erythrocytes, and hemosiderin. The multinucleated, osteoclast-like giant cells often aggregate adjacent to the sinusoidal spaces. Trabeculae of reactive, woven bone are often evident within the connective tissue. Since a normal epithelial lining is lacking, the lesion is also referred to as pseudocyst. Our histological findings were consistent with the abovementioned characteristics.

Depending on the size, site and extent of the lesion, the treatment options range from curettage, enucleation, percutaneous sclerotherapy, diagnostic and therapeutic embolization, block resection and reconstruction, and systemic calcitonin therapy. The recurrence rate is fairly high, ranging from 19% to 50% after curettage and approximately 11% after resection. This indicates the need for regular follow-up.

In conclusion, aneurysmal bone cyst of the jaws represents an enigmatic pseudocyst with variable clinicopathological, histological and radiographic presentations, therefore, often posing a diagnostic dilemma. To the best of our knowledge, this is the first case reported of the unusual transition from the initial radiolucent stage to a mature radiopaque stage with large amount of bone formation in the same individual.

References