Extrafollicular Adenomatoid Odontogenic Tumor in the Maxillary Incisor Region Disguised as Gingival Swelling

Abstract

Adenomatoid Odontogenic Tumor (AOT) is a benign, non-invasive tumor with slow but progressive growth, mainly affecting younger patients, predominantly females. It is more often located in maxilla, involving an unerupted or erupted tooth, mostly canine. There are three variants, namely follicular, extra-follicular and peripheral. Permanent cuspids account for 60% of all follicular and 89% of all extra-follicular AOT. This article discusses a unique case of extra-follicular AOT in 9 year old male patient associated with partially erupted maxillary central incisor.

Keywords: *Adenomatoid odontogenic tumor, cone-beam computed tomography, extrafollicular, radiographic variant*

Natasha Saini, Bindu Kadian, Tanu Rajain, Saurabh Narang, Ritu Namdev

Department of Pedodontics and Preventive Dentistry, Post Graduate Institute of Dental Sciences, Rohtak, Haryana, India

Introduction

Adenomatoid odontogenic tumor (AOT) is a slow-growing, asymptomatic epithelial odontogenic lesion, which was first reported by Nakayama in 1903, with indisputable proof based on both clinical and micropathologic features.[1] However, many names were used in the past, until when Philipsen and Birn in 1969 proposed the more appropriate currently used term AOT to describe the lesion.[2] This term was adopted by the WHO in 1971 under "Histological Typing of Odontogenic Tumours, Jaw Cysts, and Allied Lesions."[3] In the second edition of the WHO "Histological Typing of Odontogenic Tumours," the AOT has been defined as "a tumor of the odontogenic epithelium with duct-like structures and with varying degrees of inductive change in the connective tissue. The tumor may be partly cystic, and in some cases, the solid lesion may be present only as masses in the wall of a large cyst. It is generally believed that the lesion is not a neoplasm."[4] It constitutes 2.2%–7.1% of all odontogenic tumors.[5] Philipsen *et al*.,[6] in their collaborative retrospective study in 2007, confirmed that majority of the tumor is found in the second and third decades of life, with the highest number of cases in 10–19 years of age. It has twice the higher tendency to occur in females than males, globally. It is categorized into three groups, namely, follicular, extrafollicular, and peripheral. As the name suggests, follicular and extrafollicular are the intrabony type, while peripheral is extraosseusly located. Permanent cuspids account for 60% of all follicular and 89% of all extrafollicular AOT. As far as peripheral AOT is concerned, gingiva in the anterior maxillary region is the most frequent location.[7] This case report describes a rare case of AOT associated with partially erupted maxillary incisors.

Case Report

A 9-year-old male patient was referred by a private dental practitioner to the Department of Pedodontics and Preventive Dentistry, Postgraduate Institute of Dental Sciences, Rohtak, India, with the chief complaint of swelling in the upper front teeth region and noneruption of the same for the past 1 year. Detailed history by his father revealed that a slowly progressive, nontender swelling of the gum over the front teeth is present for 7–8 months. His medical history was noncontributory. The patient was in good general health and presented no conspicuous extraoral findings. Intraoral examination revealed a solitary smooth well-defined swelling of 2 cm × 1.5 cm in size in the left maxillary region, involving marginal gingiva of the

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Address for correspondence:
Dr. Natasha Saini, Department of Pedodontics and Preventive Dentistry, Post Graduate Institute of Dental Sciences, Rohtak, Haryana, India.
E-mail: nitusaini23@gmail.com

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left maxillary incisors extending superiorly up to labial vestibule, anteroposteriorly from the midline to distal margin of the left maxillary lateral incisor. Overlying mucosa was of the normal color [Figure 1]. On inspection, the surface of the swelling was smooth and nonulcerated giving an appearance of gingival enlargement. On palpation, the swelling was nontender, nonpulsatile, nonfluctuant, and firm in consistency. Pocket depth with respect to labial surface of the central incisor was 11 mm. Both 21 and 22 showed a positive response to the pulp vitality test. Radiographically, a diffuse radiolucency was seen with respect to 21 in orthopantomograph [Figure 2]. However, to understand the radiolucency better, cone-beam computed tomography (CBCT) was done which revealed a well-circumscribed unilocular radiolucency with a well-defined radiopaque border of 15.4 mm × 18.8 mm × 14.2 mm, extending from the mesial surface of 21 to lateral surface of 22 and vertically from an incisal third of the incisor to junction of the middle third and apical third of the root [Figures 3-5]. The radiolucency overlapped the buccal surface of the root and most of the crown portion of the left maxillary central incisor. No tooth resorption was observed. Based on the clinical and radiographic findings, a provisional diagnosis of a benign bony tumor of odontogenic or nonodontogenic origin was made.

Decision was made to surgically remove the lesion under local anesthesia. After reflecting the flap on the facial aspect of the maxilla, a thin but intact buccal cortical bone was found attached to the lesion [Figure 6]. After removing it, a reddish cystic tissue became evident which was completely enucleated [Figure 7]. Separating the lesion from the adjoining bone caused no problem. There was no evidence of oronasal communication, and the palatal mucosa was intact. The wound was closed by primary intention. Excised tissue specimen was fixed in 10% formalin and sent for the histopathological examination. The histopathological report revealed hyperchromatic, cuboidal to columnar, spindle-shaped tumor cells arranged in nests where they show duct formation, rosette pattern, and solid islands. In these ducts, the hyaline rim is seen. Around these tumor mass, there is condensation of thick collagen bundles with extravasation of RBCs being seen. At few foci, tiny calcifications are evident. It was suggestive of AOT [Figure 8].

The postoperative course was uneventful. The patient was followed up regularly, and no recurrence of the swelling was noted. CBCT was taken at 2-year follow-up,

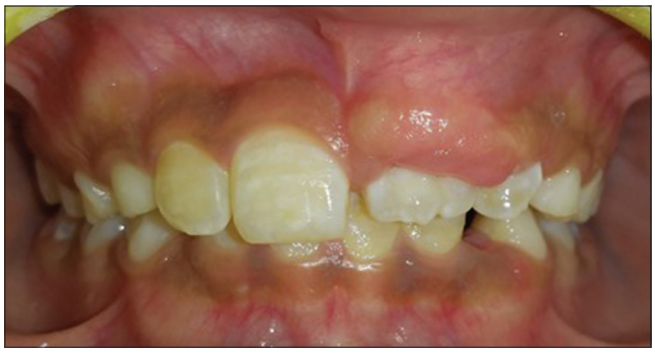

Figure 1: Clinical examination revealing swelling in the anterior maxillary region involving 21 and 22

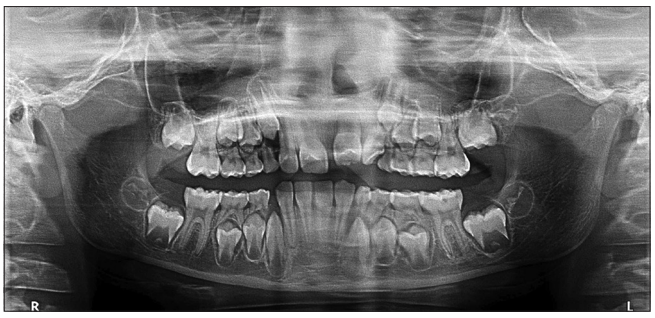

Figure 2: Orthopantomograph

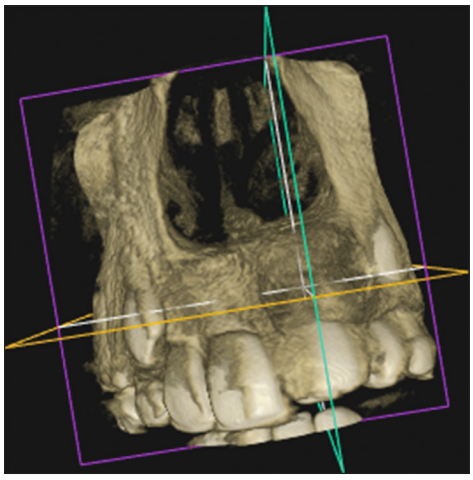

Figure 3: Cone-beam computed tomography showing a unilocular radiolucency involving buccal cortex in the 21 and 22 region

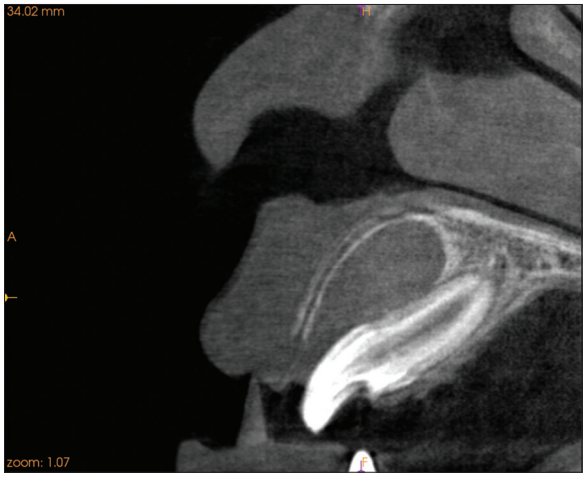

Figure 4: Cone-beam computed tomography showing unilocular radiolucency extending from the junction of the middle third and apical third of 21 up to the middle third of the crown

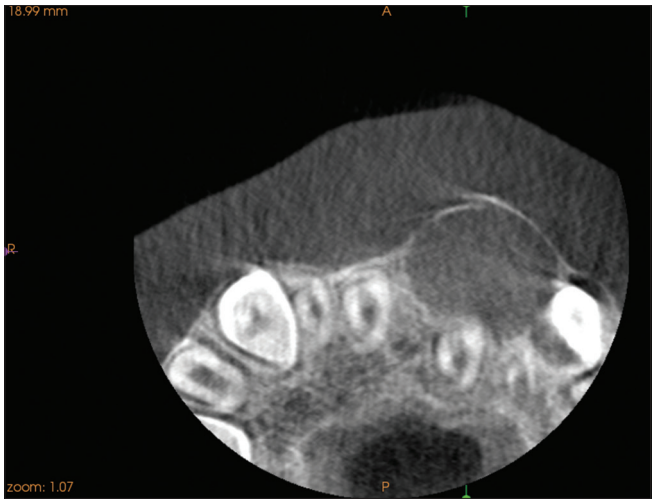

Figure 5: Cone-beam computed tomography showing unilocular radiolucency extending from the junction of the middle third and apical third of 21 up to the middle third of the crown

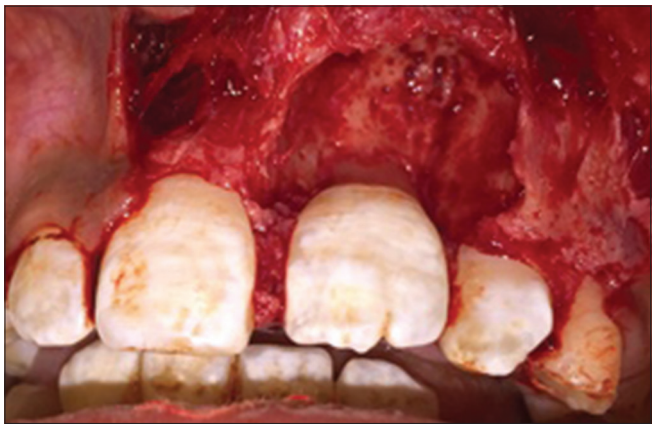

Figure 7: Complete enucleation of the lesion

there was no evidence of recurrence of pathology, and CBCT revealed complete bone formation/normal bone healing [Figures 9-11].

Discussion

Earlier in the past, AOT was first described under the term epithelioma adamantinum, by Steensland in 1905.[8] However, Philipsen *et al*.[9] in their recent updated review of AOT recognized cases documented by Nakayama in 1903 to be the first complete description of AOT in Japanese journal. Since 1903, many authors presented AOT under various terms, including adamantinoma by Harbitz, epithelial odontome by James and Forbes, adenoameloblastoma by Bernier and Tiecke, cystic complex composite odontoma by Miles, and adenomatoid ameloblastoma by Ishikawa and Mori, until Philipsen and Birn in 1969 presented a review based on 76 cases of AOT, which clearly distinguished it from ameloblastoma and, in addition, provided sufficient evidence in support of lesion to be renamed as AOT.[10] In 1971, the WHO in their international histological classification of tumors No.

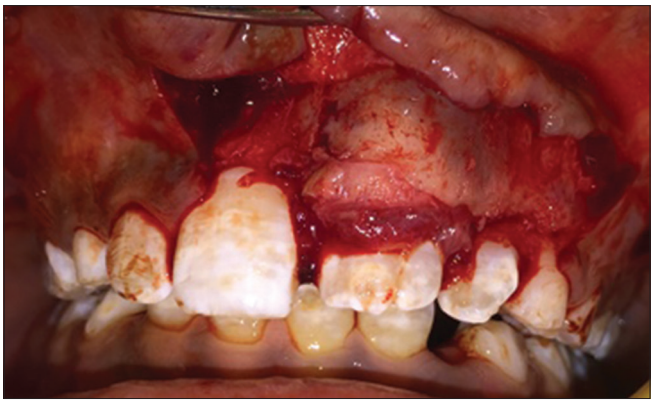

Figure 6: Surgical exposure of the defect revealing expanded and thinned buccal cortical plate

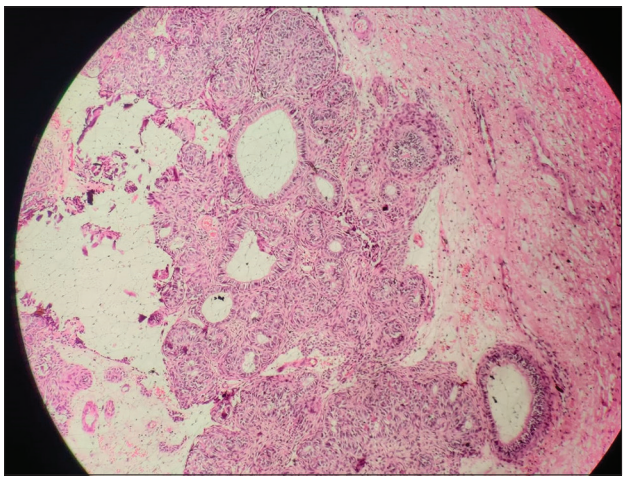

Figure 8: Duct-like structures lined by cuboidal-to-columnar cells within the cellular area. Few foci of calcifications are also evident

5 titled "histological typing of odontogenic tumours, jaw cysts, and allied lesions" adopted the currently used term "AOT."[3] Philipsen *et al*.,[7] based on 500 cases, provided a detailed biologic profile of this unique tumor in 1991 and further updated the clinical and epidemiological profile in 2007 after analyzing a total of 1082 cases from all around the world through a collaborative retrospective study.[6]

In earlier years, the relative frequency of AOT corresponded to 2.2%–7.1%.[5] However, in recent times, data from worldwide publications showed a considerably wider range of occurrence constituting 0.6%–38.5% of all odontogenic tumors.[11,12]

Majority of the AOTs are usually found in 10–19 years of age, with 87.2% cases occurring in the second and third decades of life. AOT tends to occur 1.9 times more in females than males if all age groups and all types are taken in account.[6] However, if geographical aspects are considered, significant differences appear between Asians and non-Asians. Asian cases show a female:male ratio of 2.3:1 compared to non-Asian cases showing a ratio of 1.4:1.[5] The patient in this case report is a 9-year-old Asian

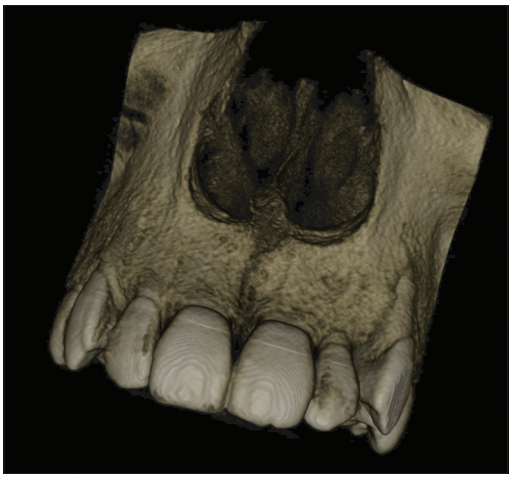

Figure 9: At 2-year follow-up, cone-beam computed tomography showing significant bony healing of the lesion

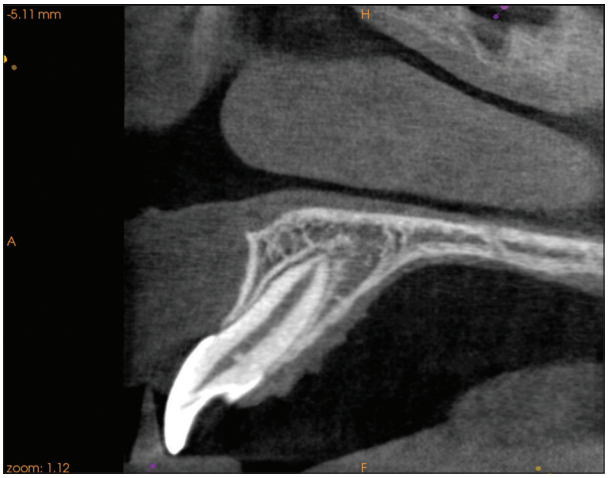

Figure 10: Two-year follow-up cone-beam computed tomography showing bone formation covering buccal root surface of 21

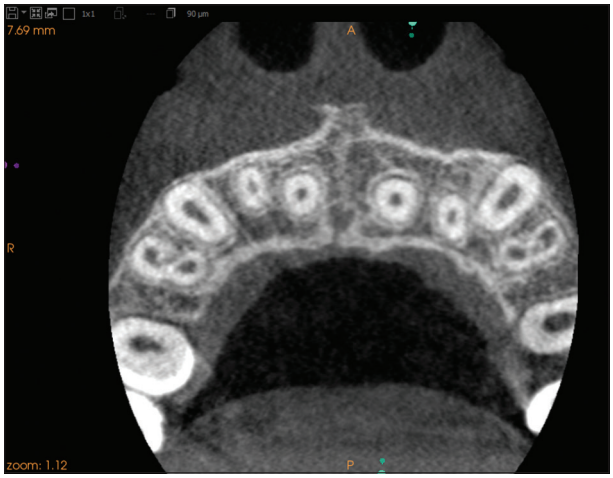

Figure 11: Two-year follow-up cone-beam computed tomography revealing medullary bone formation surrounding the buccal surface of 21 and 22

male. Furthermore, only 14 cases of male patients in the age group 0–9 years were reported by Philipsen *et al*.[6] in 2007. 64.3% of all AOTs are located in the maxilla while 35.7% are found in the mandible, the majority of which are located intraosseously.

AOT is a benign and noninvasive lesion with slow but progressive growth. The patient usually shows no subjective symptoms, except swelling. Cortical expansion and displacement of the adjacent teeth are seen in the intraosseous variant. However, root resorption is rare. Tumor size usually measures 1–3 cm in diameter although larger lesions have been reported in the literature.[13]

It is well established that AOT occurs in central (intraosseous) variant and peripheral (extraosseous) variant. Radiographically, the intraosseous variant is further subdivided into follicular and extrafollicular form, depending on whether the crown of the unerupted tooth is involved or not. In follicular type, tumor surrounds the crown of the embedded tooth, whereas in extrafollicular type, no such association is seen. The peripheral (extraosseous) variant appears as epulis-like fibrous swelling attached to the labial gingiva.[7] The most common type of AOT is follicular one which accounts for 70.8% of total AOTs reported whereas peripheral variant is the least common with only 2.3% cases documented. Extrafollicular AOT constitutes 26.9% of the total AOT cases. Intraosseus variant shows a M:F ratio of 1:2, while for peripheral variant, M:F ratio comes out to be 1:5.3, which is quite remarkable.[6] 62.3% of the follicular and extrafollicular variants are located in the maxilla, whereas 35.3% are found in the mandible. The peripheral or extraosseous variant is exclusively restricted to the anterior maxillary labial gingiva, with only 0.3% of cases being reported in the mandible.[5] Moreover, the most common tooth involved in the follicular AOT is a permanent canine, with maxillary canine alone constituting 40% of the cases and unerupted third molars comprising 2.8%, while in extrafollicular variant, permanent canine accounts for 89% of the cases.[7] Permanent incisors, premolars, molars, and deciduous teeth are rarely involved.[14]

Radiographically, the follicular type shows a well-defined, round or ovoid, unilocular radiolucency associated with the crown and often part of the root of an unerupted tooth, thus mimicking a dentigerous or follicular cyst. In fact, 77% of follicular AOTs are initially diagnosed as dentigerous or follicular cysts.[2] Moreover, many cases have been reported in the literature in recent years, which are believed to arise from a dentigerous cyst or diagnosed as an adenomatoid odontogenic cyst. Most of these cases are of Indian/Asian ethnicity.[9] On the other hand, extrafollicular type manifests as a well-defined, unilocular radiolucency without any pericoronal relationship. Philipsen *et al*.[5] further subdivided extrafollicular type into four types (E_1–E_4) depending on the location of radiolucency. In the first type (E_1), radiolucency is not related to tooth structures neither erupted nor unerupted. In the second type (E_2), radiolucency is interradicular in

position, and tumor expansion leads to the adjacent root to diverge apically. The third type (E_3) of radiolucency is superimposed at root apex, and in the fourth type (E_4), the radiolucency is superimposed at the midroot level. Sometimes, discrete foci of radiopacities in a flocculent pattern are seen in intrabony variant. According to the radiographic classification of AOT, the present case falls in the extrafollicular variant and subtype E_4.

Dare *et al*.[15] proved that the intraoral periapical radiograph is preferable to panoramic radiography, in demonstrating the flocculent pattern of the radiopaque discrete calcified deposits. However, with the advancements in the radiographic techniques, CBCT is preferred over intraoral periapical radiograph and panoramic radiograph to study the detailed internal structure of the lesion, especially the superimposed areas, as it provides a three-dimensional view of the structure and provides high-contrast resolution for calcified foci along with eliminating the superimposition.[16] Recently, Asaumi *et al*.[17] studied the use of magnetic resonance imaging in the radiographic differential diagnosis of AOT.

The differential diagnosis of extrafollicular AOTs includes central giant cell granuloma, benign fibro-osseous lesions (e.g., early cemento-ossifying fibroma), lateral periodontal cyst, lateral radicular cyst, and apical radicular cyst apart from other lesions, such as dentigerous cyst, odontogenic keratocyst, calcifying odontogenic cyst, unicystic ameloblastoma, ameloblastic fibroma, early ameloblastic fibro-odontoma, odontogenic fibroma, and calcifying epithelial odontogenic tumor, which are frequently found in a pericoronal relationship to a tooth but may also appear in a nonpericoronal position to an unerupted or erupted tooth.[10]

Earlier, different hypotheses regarding the origin of AOT have been proposed, but none of them received much support.[10] Later, in 1992, Philipsen *et al*.[18] suggested that the gubernaculum dentis may be implicated in the development of AOT as a vast majority of AOT arises in association with the successional tooth during active dental development. Keeping this in mind, Ide *et al*.[19] presented the review on the early development of AOT and concluded that the embryonic source of more than 96% of AOT was found in the gubernacular cord of a developing permanent incisor, canine, or premolar. For extracoronal variant, the normal eruption of the involved tooth may leave AOT either on a lateral root surface, at the root apex, or superimposed on the root surface.

Due to the similar benign biologic behavior of nearly all typical variants of AOTs and the presence of a smooth, well-developed capsule in most of the lesions, conservative complete surgical enucleation and curettage is considered as the treatment of choice. Conservative treatment is adequate as the tumor is noninvasive, well encapsulated and its lining can be easily separated from the bone. The recurrence is very low following complete removal of the primary lesion.[13] Prognosis is also excellent in most of the cases.[20]

The present case is very unique and unusual as it occurred in a comparatively younger 9-year-old male patient, and also, the location of the tumor, i.e., the maxillary incisor region, is quite uncommon as far as an extrafollicular variant of the AOT is concerned. Due to its smaller size and its mere presence as a gingival swelling, it often leads to misdiagnosis. Very few such cases of extrafollicular AOT associated with maxillary central incisor are reported in the literature.

Conclusion

The AOTs are unique, slow-growing, asymptomatic lesions occurring in a specific age group and a quite restricted anatomic location (the incisor-canine region). However, morphologically, it can be easily confused with other odontogenic lesions occurring in that area. Enucleation was planned in the case described due to small size of the lesion and sclerotic margins. However, histopathological diagnosis is mandatory to arrive at the final diagnosis of the lesion. Furthermore, CBCT is a beneficial modality in describing the radiographic features of the AOT.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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