Otosclerosis

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Contents

Abstract

Otosclerosis is a disorder of the bony labyrinth with usual onset in the third decade. The disease is bilateral in a majority of cases and such patients have conductive hearing loss. In case of fenestral otosclerosis hypodense foci are seen on CT/CBCT just anterior to the oval window, at the so-called fissula ante fenestram region. In case of retrofenestral otosclerosis additional foci are seen in the otic capsule around the cochlea.

1 Etiology and Categories

Otosclerosis is a disorder of the bony labyrinth and exclusively affects human beings. In otosclerosis, the ivory-like enchondral bone of the otic capsule is replaced by immature and spongy new bone, and this process of remodeling occurs continuously. The process can become quiescent at any time or may become reactivated, in a way that otosclerotic foci commonly contain both active and inactive regions (Schuknecht 1993a). Unilateral involvement is only seen in about 10–15 % of the cases and there is a 2:1 female predominance. The risk to an affected person of having a child who will eventually develop otosclerosis is 1 in 4 (Donnell and Alfi 1980; Shin et al. 2001a). In comparison with patients with a sporadic form of otosclerosis, the radiologic lesions are more often detectable, bilateral, and severe in the familial forms (Shin et al. 2001a).

Otosclerosis usually has its clinical onset in the third decade, with symptoms of conductive hearing loss due to the impaired movement of the stapes by invasion of the stapediovestibular articulation. Under the age of 18 years old the diagnosis is rarely made (Lescanne et al. 2008). Fixation of the stapes by foci located anterior to the oval window—in the so-called region of the fissula ante fenestram—is found in 96 % of ears from persons with clinical otosclerosis. In 49 % of the cases, otosclerotic foci are also present in other locations, of which the most frequent ones are the oval window

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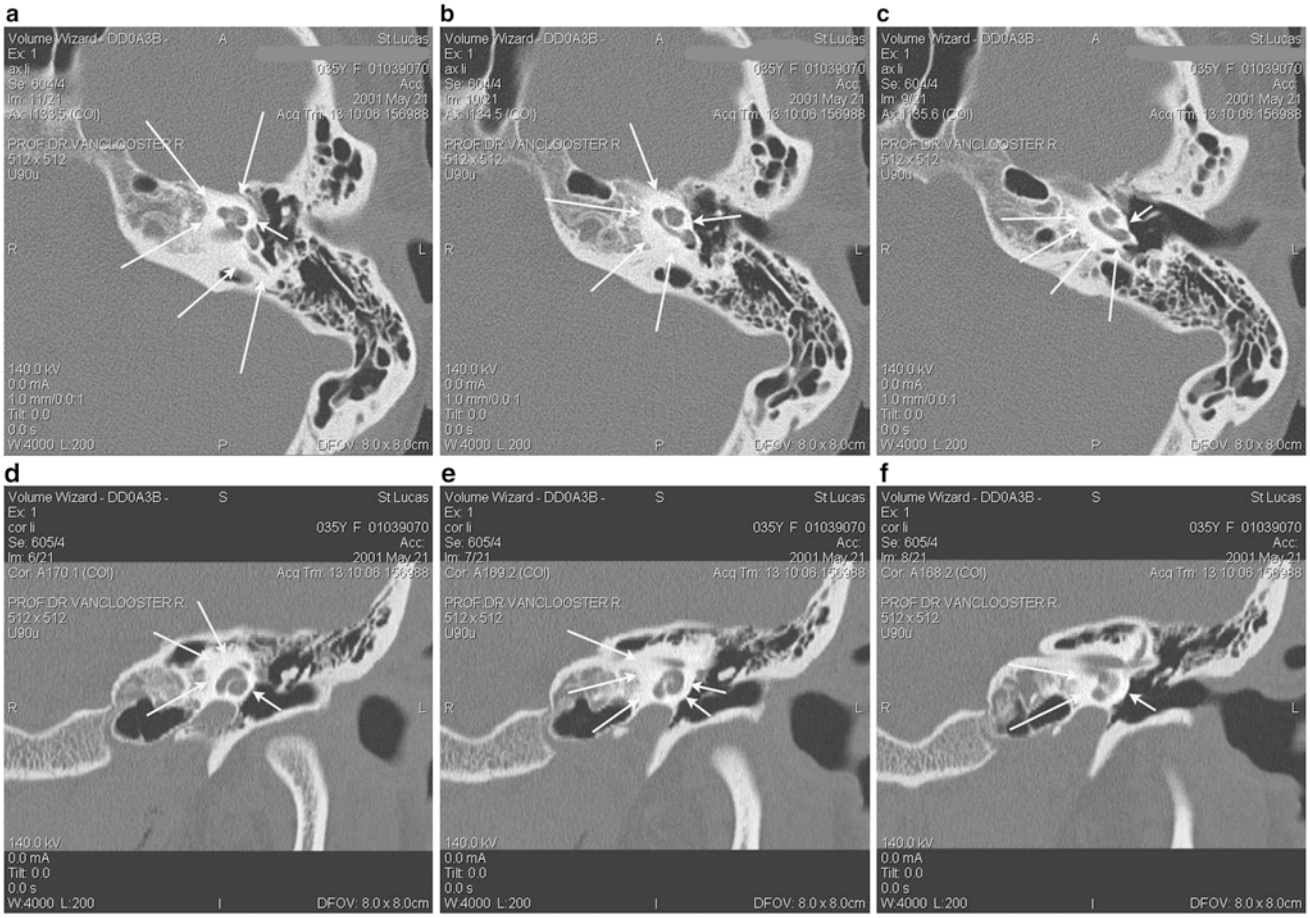

Fig. 1 Three consecutive axial (**a–c**) and coronal (**d–f**) CT images are shown in a nonotospongiotic ear to insist on the careful inspection of the dense appearance of the normal bone in many regions. It is very important to systematically inspect the region of the fissula ante fenestram on the axial image set, situated just anterior to the oval window and anterior stapes crus, in order to be sure that fenestral otosclerosis is excluded (*small arrows* on images **a–c**). The same region can be inspected on the coronal images too (*small arrows* on images **d–f**). An intensely high density must be present in the otic capsule region surrounding the cochlea, in order to be sure that cochlear otosclerosis is excluded (*large arrows* on images **a–f**)

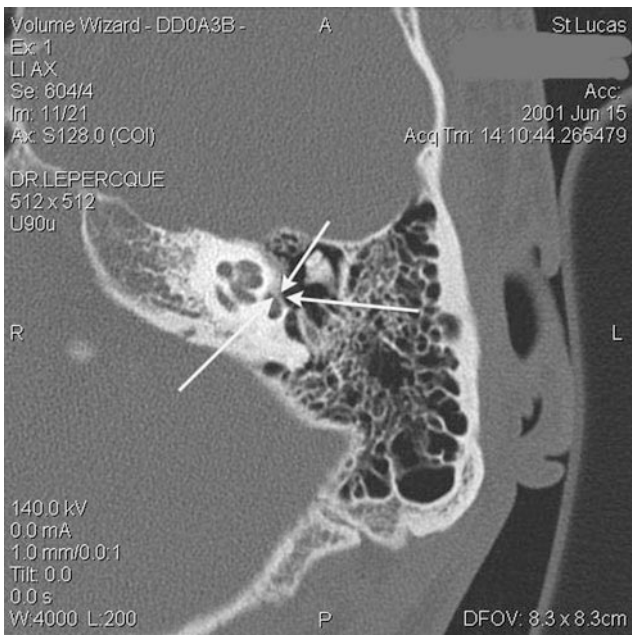

Fig. 2 The axial CT image at oval window level shows a hypodense region of otosclerotic origin in the footplate itself, and anterior to the oval window (*arrows*). Note that the footplate is thickened

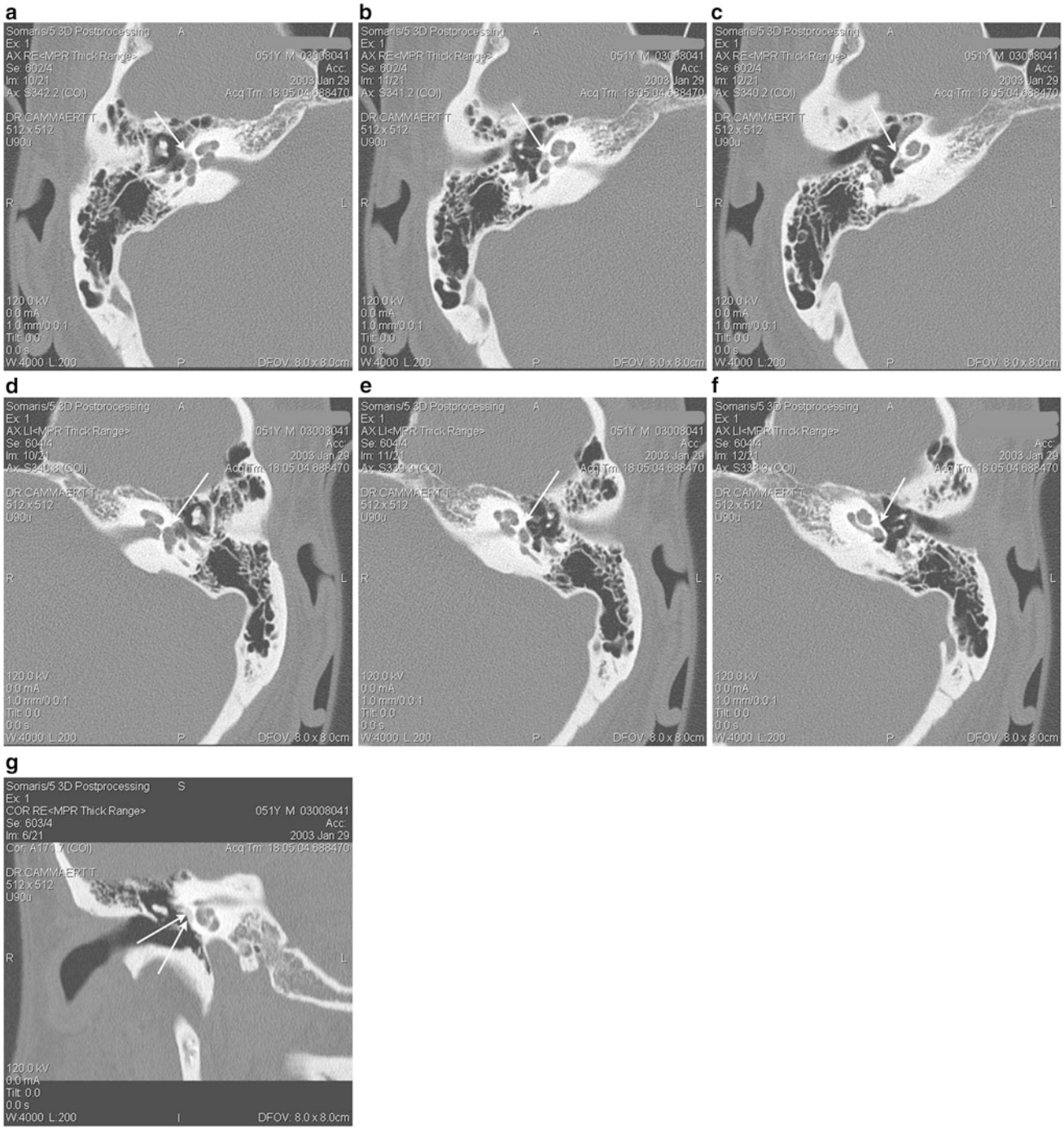

Fig. 3 In the right ear three consecutive CT images (**a**–**c**) show a lucent area just anterior to and extending to the oval window: fenestral otosclerosis (*arrow*). In the same patient, the normal contralateral ear is shown. Note the normal high density anterior to the footplate (**d**–**f**). In this same patient, a coronal reconstruction (**g**) obviously depicts the otospongiotic focus in the region of the promontory (*arrows*)

niche (30 %), the cochlear apex (12 %), and posterior to the oval window (12 %). Round window obliteration is seen in 7 % of ears with clinical otosclerosis. Other even rarer sites of involvement are the walls of the internal auditory canal, around the cochlear aqueduct, around the semicircular canals, and within the footplate (Schuknecht and Barber 1985). Incus and malleus invasion each have been reported once, and the internal auditory canal itself is never invaded. Invasion of the labyrinthine spaces rarely occurs (Schuknecht 1993a). The extension of otosclerosis as seen on CT/CBCT scans seems to generally correlate with the preoperative and postoperative hearing loss (Marx et al. 2011).

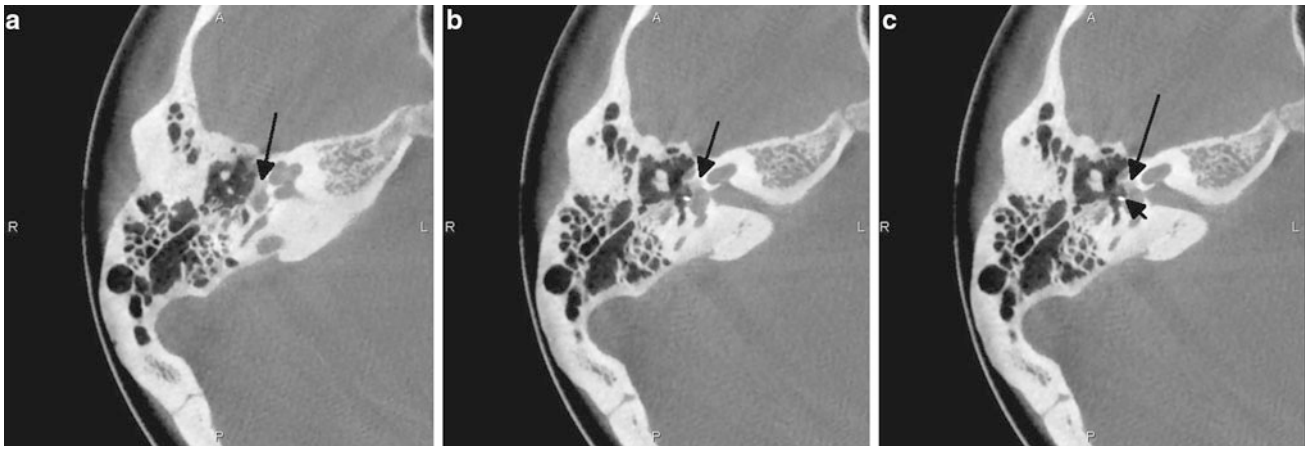

Fig. 4 On these axial CBCT images (**a**–**c**) of the right middle ear, a hypodense focus of otosclerosis is nicely depicted in the region just anterior to the oval window (*large arrow*). The patient is treated with a stapes prosthesis (*small arrow*)

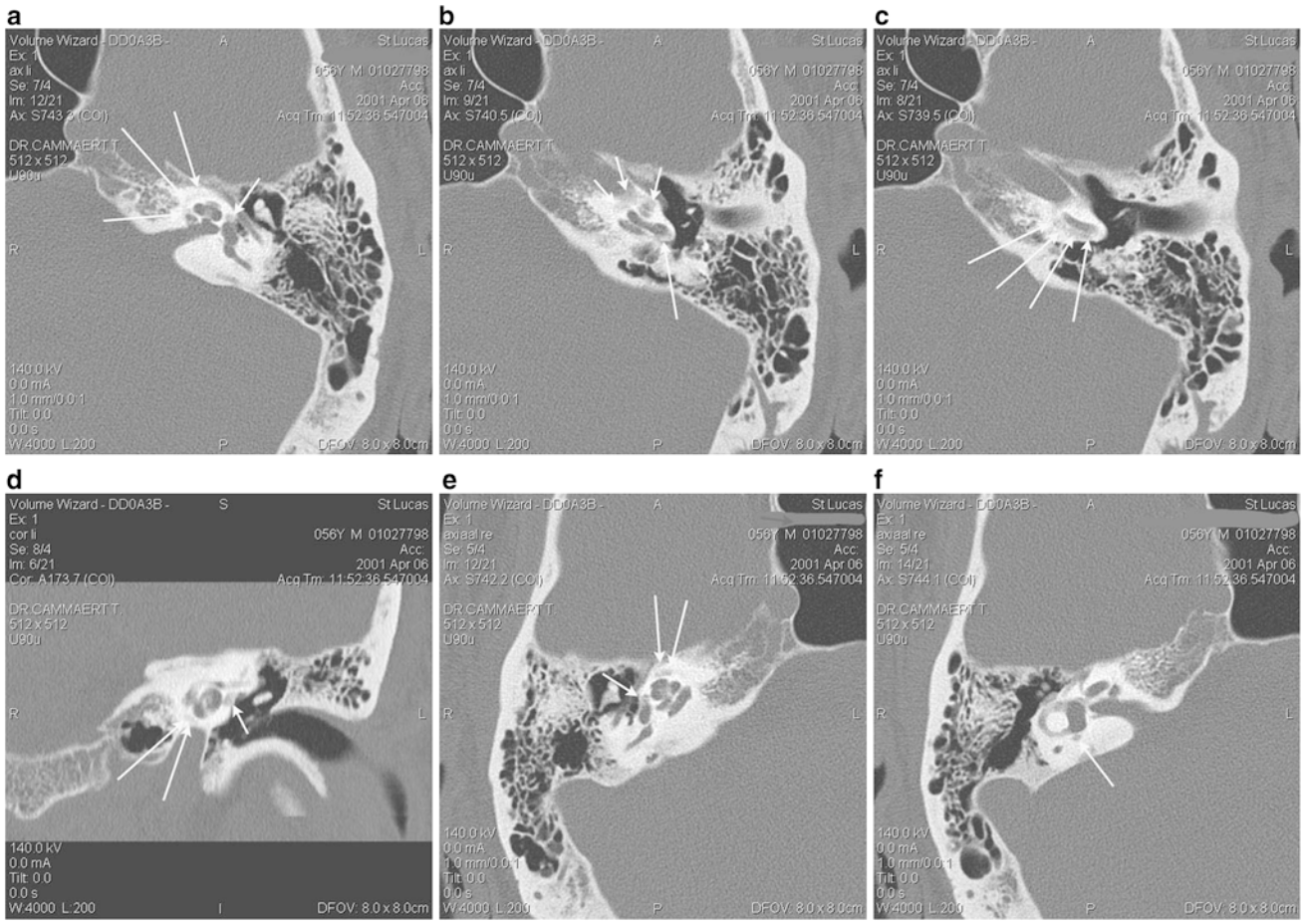

Fig. 5 Multiple CT images are shown from a patient with bilateral fenestral and retrofenestral otosclerosis. On the most cranial axial image on the left side (**a**) lucencies of otosclerotic origin are seen around the cochlea (*large arrows*) and anterior to the oval window (*small arrow*). On the image below (**b**) cochlear otosclerosis is seen medial to the apical and middle cochlear turns (*small arrows*), but also around the basal cochlear turn, and in the round window region (*large arrows*). On the slice more caudally (**c**) more otosclerotic foci are obviously present around the basal turn of the cochlea. The coronal image from the same left ear confirms the presence of fenestral (*small arrow*) and retrofenestral (*large arrows*) foci of otosclerosis (**d**). In the contralateral right ear, the axial image performed at footplate level (**e**) shows more otosclerotic lucencies at the fissula ante fenestram (*small arrow*) and in the pericochlear otic capsule (*large arrows*). On the slice performed 2 mm more caudally (**f**) otosclerosis is seen posterior to the vestibule (*arrow*)

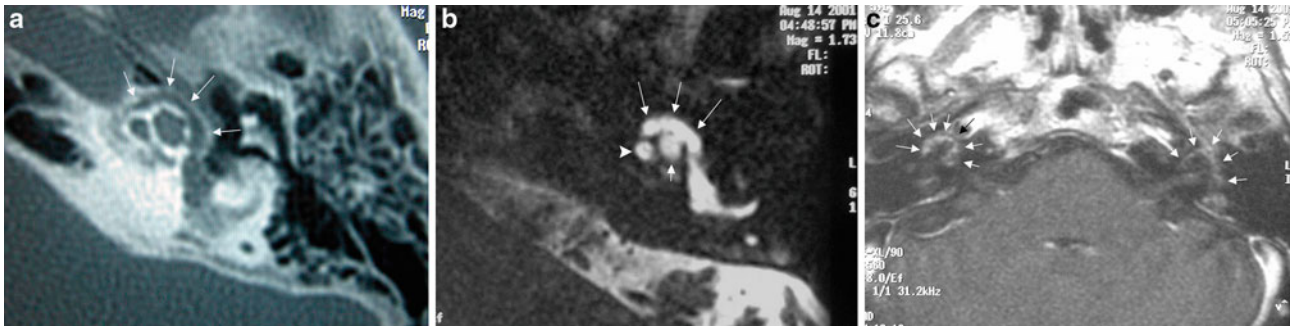

Fig. 6 The axial CT image at the level of the middle and apical turns of the left cochlea (**a**) shows otosclerotic changes around the cochlea: the so-called fourth ring of Valvassori is seen (*arrows*). The axial 3D FSE T2-weighted image at the same level (**b**) again demonstrates the fourth ring of Valvassori (*large arrows*), now seen as a semicircular hyperintensity around the middle (*arrowhead*) and apical (*small arrow*) turns of the cochlea. On the axial contrast-enhanced T1-weighted image through both inner ears (**c**) diffuse semicircular enhancement is noted around the cochlea bilaterally: the diagnosis of bilateral severe retrofenestral otosclerosis in the active phase can be made (Courtesy by Dr. B. Defoer, Antwerp, Belgium)

Sensorineural hearing loss can be seen in patients with otosclerosis, and is believed to be caused by the accumulation in the inner ear of products liberated by the growth of the otosclerotic foci that are present in the pericochlear region (Schuknecht 1993a). In patients with pericochlear otosclerotic foci, sensorineural hearing loss is present with a higher degree (Guneri et al. 1996).

Many authors have used CT grading systems of otosclerosis in their studies. Some authors grade on the basis of disease site and progression (Valvassori 1993), or on the basis of disease site with subdivision of these different sites (Shin et al. 2001b), whereas others categorize the presence of otosclerosis on the basis of the location and appearance of the disease (Marshall et al. 2005). We use a simple system with two categories of otosclerosis described on the basis of where the anomalies are seen: fenestral and retrofenestral otosclerosis. As the name suggests (fenestra is the latin word for window) fenestral otosclerosis affects the lateral labyrinthine wall, including the promontory, facial nerve canal, and both the oval and round window niche (Swartz and Harnsberger 1992). Retrofenestral otosclerosis involves the pericochlear otic capsule and is almost always present together with fenestral otosclerosis. For this latter reason it is better to use the term retrofenestral otosclerosis than cochlear otosclerosis, another term also in use.

2 CT/CBCT and MRI Appearance

CT or CBCT is the tool of choice to investigate for the eventual presence of otosclerotic lesions (Weissman 1996), and its sensitivity to make the diagnosis was recently estimated as high as about 90–95 % (Lagleyre et al. 2009). Both axial and coronal images are performed in order to give a detailed description of the extent and location of the lesions. During the reading of the CT/CBCT images, inspection for contralateral otosclerotic foci is always mandatory, even in case of unilateral clinical findings. CBCT allows to make images with a higher spatial resolution than does CT, and this is achieved with a much lower radiation dose.

In normal circumstances, the bone of the otic capsule is homogeneously dense (Fig. 1). The otosclerotic foci contain spongy new bone, which appears lucent on CT/CBCT (Valvassori and Dobben 1985; Miura et al. 1996). These are seen in the medial wall of the labyrinth in case of fenestral otosclerosis (Mafee et al. 1985a). It is important to inspect the region of the fissula ante fenestram very carefully, since otosclerotic foci can be very small. Especially, the axial images will be helpful to do so. In some cases, footplate thickening is noted (Veillon et al. 2001) (Figs. 2, 3, and 4). Additional foci of retrofenestral otosclerosis can be seen in the otic capsule around the cochlea (Mafee et al. 1985b) (Fig. 5). It is important to notice round window localizations, since these can obliterate the window and cause problems to insert a cochlear implant. In extensive cases of retrofenestral otosclerosis, a so-called 'fourth ring of Valvassori' is described (Figs. 6 and 7).

MRI is sporadically used in case of otosclerosis. Otosclerosis very often leads to severe hearing loss in a chronic progressive manner. Initially an otospongiotic phase takes place and causes an inflammatory osteolytic process in the otic capsule. During this 'active' phase, the otospongiotic foci will enhance on T1-weighted SE MRI images performed after IV injection of gadolinium (Ziyeh et al. 1997; Stimmer et al. 2002). On the T1-weighted images before gadolinium injection, a ring of intermediate signal can be present in the pericochlear and perilabyrinthine regions, and an increased signal may also be seen on the T2-weighted images (Goh et al. 2002) (Fig. 6). MRI has also proven its usefulness in the investigation of complications of stapes surgery performed for otosclerosis. Some of these complications, such as reparative intravestibular

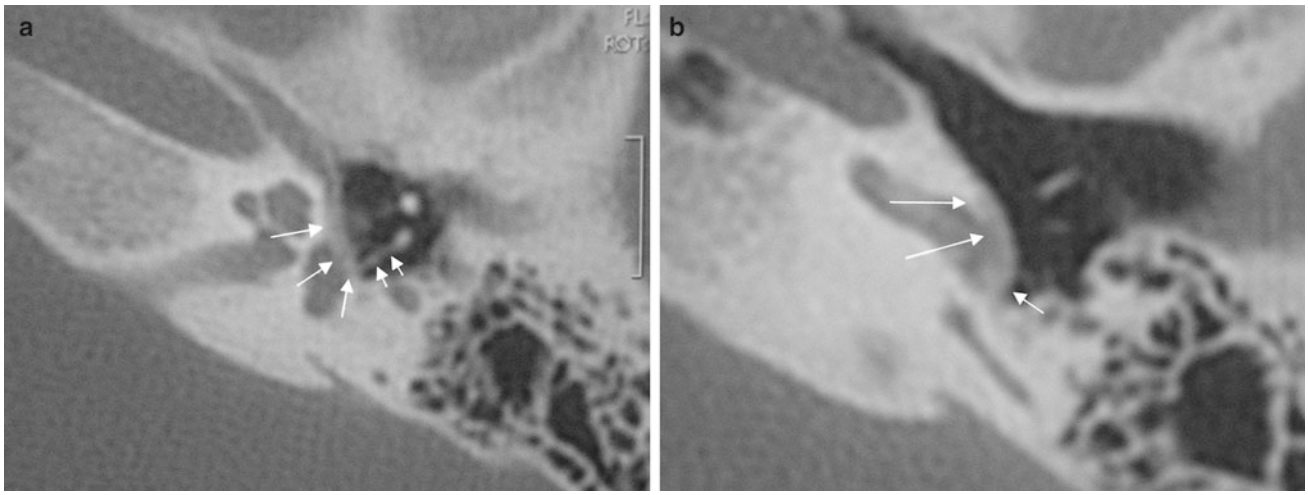

Fig. 7 The axial CT image at the level of the oval window (**a**) shows an otosclerosis focus at the fissula ante fenestram extending over the oval window, and with diffuse and severe thickening of the footplate (*large arrows*). Note the postoperative status with stapedectomie and replacement by a piston, posteriorly displaced (*small arrows*). The axial CT image at the level of the basal cochlear turn and round window (**b**) demonstrates the extension of the otosclerotic changes over the promontory toward the round window niche (*large arrows*). There is extension of the otosclerotic changes to the round window with total obliteration of the access to the round window (*small arrow*), thus making a later cochlear implantation virtually impossible (Courtesy by Dr. B. Defoer, Antwerp, Belgium)

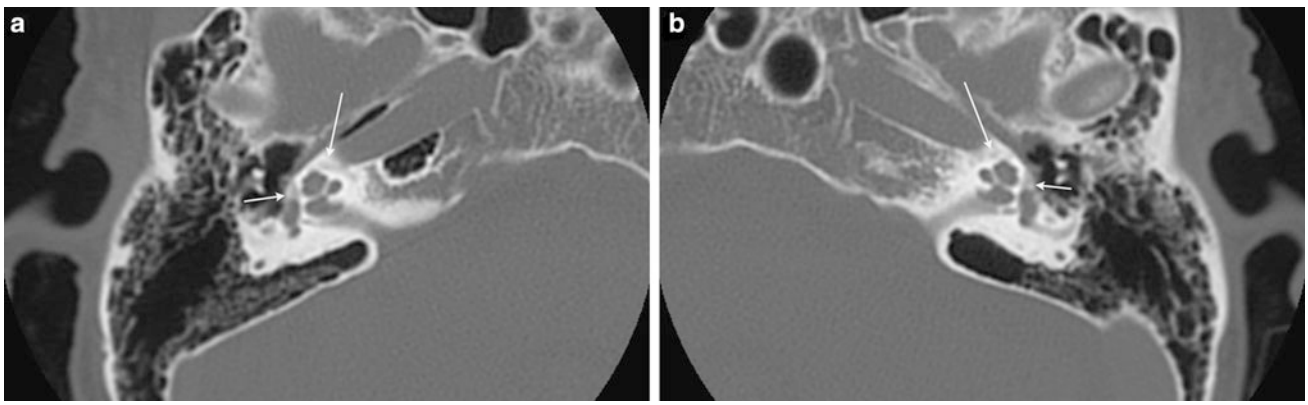

Fig. 8 The axial CT images at footplate level are shown in both temporal bones from a patient with proved osteogenesis imperfecta. Note that the bone in the region of the fissula ante fenestram (*small arrow*) and in the pericochlear region (*large arrow*) has become lucent. The CT findings do not differ from those seen in patients with combined fenestral and retrofenestral otosclerosis (Courtesy by Dr. B Defoer, Antwerp, Belgium)

granuloma formation, intralabyrinthine hemorrhage, and bacterial labyrinthitis are detectable with MRI, while they pass unrecognized on CT/CBCT examinations (Rangheard et al. 2001).

3 Differential Diagnosis

Osteogenesis imperfecta is a rare disease and is often associated with hearing loss. It is an inherited generalized disorder of type-I collagen synthesis (Zajtchuk and Lindsay 1975; Berghstrom 1981; Berger et al. 1985; Schuknecht 1993b). The classic triad of blue sclerae, spontaneous fractures, and hearing loss is known as the Van der Hoeve and De Kleyn syndrome (Schuknecht 1993b; Czerny and Temmel 1999). The CT/CBCT appearance can be undistinguishable from otosclerosis, with lucent bone anterior to the oval window and in the pericochlear otic capsule (Fig. 8). On MRI similar findings have been described as those seen in the active phase of otosclerosis: pericochlear soft tissue signal intensities and enhancement after contrast injection (Ziyeh et al. 2000).

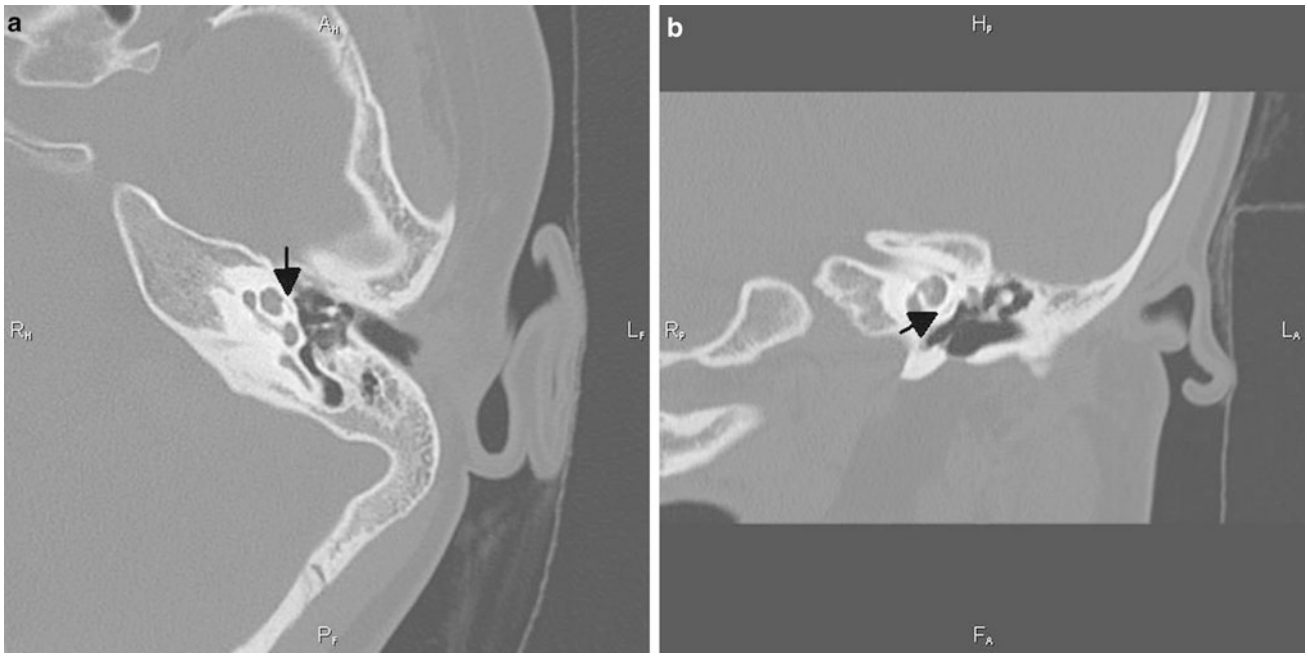

Fig. 9 On the axial CT image (**a**) in a 10-year-old child performed in the region of the fissula ante fenestram, the hypointense C-shaped cochlear cleft is noted in the otic capsule. On the corresponding coronal image (**b**) the cochlear cleft often has a more linear shape

Paget disease is another condition to be excluded if a decreased attenuation is seen in the otic capsule (Mafee et al. 1985b).

4 The Cochlear Cleft: Potential Imaging Pitfall in Children

The so-called 'cochlear cleft' is a hypodense cleft in the cochlear otic capsule seen on CT/CBCT examinations in the region anterior to the oval window. It is particularly seen in children with an average incidence of about 40 % (age group from 0.5 to 19.3 years old) (Chadwell et al. 2004). Another group of investigators noted an incidence of 32 % in a pediatric population aged between 0 and 9 years old (Pekkola et al. 2004). Its incidence decreases with age. The finding is believed to be a space in the interface between the endosteal and outer layers of the otic capsule or that it is related to the fissula ante fenestram (Fig. 9). The finding does not have any pathological implication in the absence of a clinical evidence for otosclerosis or osteogenesis imperfecta.

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