CT of the Temporal Bone in a Patient with Osteopathia Striata and Cranial Sclerosis

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Summary: The authors present a case of congenital dysplasia affecting the long bones, skull, and other systems in a 7-year-old girl, with special attention to CT of the temporal bone, which showed abnormal ossicle fixation, a narrowed Eustachian canal, thickened sclerotic bone, and a small mastoid antrum and middle ear cavity. CT of the temporal bone can help one distinguish the etiologies of hearing loss associated with this disorder.

Index terms: Temporal bone, computed tomography; Sclerosis, cranial

Osteopathia striata (OS) with cranial sclerosis is a rare and potentially symptomatic form of OS (Voorhoeve disease), one of the sclerosing bone dysplasias that usually have few or even no clinical manifestations. In addition to long bone involvement, there are variable effects on the craniofacial structures including the temporal bones. Plain radiographic findings have been well described and illustrated (1–18) but, to our knowledge, only one previous temporal bone computed tomography (CT) description (data not shown) has been published (13). The current case has CT findings that correlate well with the clinical presentation.

Case Report

A 7-year-old girl, previously undiagnosed, presented to us for evaluation of bilateral conductive hearing loss. High resolution CT exam of the temporal bones (Figs. 1A–1H) revealed generalized sclerosis and thickening of the osseous structures with poorly pneumatized and underdeveloped mastoids including the antrum and aditus ad antrum on each side. The Eustachian canals appeared diminutive. On each side, the head of the malleus and the body of the incus appeared depressed in position with regard to the epitympanic recess and partially fixed to the walls of the tympanic cavity. The external auditory canals, internal auditory canals, and seventh nerve canals appeared normal as to size and position. Elsewhere, the arch of the palate was noted to be high and the mandibular condyles flat.

Skeletal radiographs (Figs. 1I–1K) and further physical exam revealed other classical features of OS with cranial sclerosis including longitudinal striations throughout dense bones, diffuse cranial sclerosis especially at the skull base, macrocrania with biparietal bossing, bifid uvula, high arched palate, micrognathia, partial hypodontia of some permanent teeth, telecanthus, and small ventricular septal defect. There was also a history of large fontanelles in infancy, recurrent otitis media, and learning disability.

Other radiographic findings in addition to these CT findings, which correlate with the patient's conductive hearing loss and recurrent otitis media, included small femoral heads (Fig. 1I) and squared broad distal phalanges. Clinical findings consisted of distinct-appearing long palms and fingers with squared-off fingertips and short, broad nails.

Discussion

OS with cranial sclerosis is an extremely rare bone disorder of autosomal dominant inheritance with apparent complete penetrance, but variable expressivity (14). The prevalence is probably less than 0.1 per million (19). The phenotype appears to be sex-influenced in that the classic craniofacial appearance, hearing loss, and cranial nerve involvement have been described predominantly, although not exclusively, in females. This form of OS is one of the few that have any clinical implications. Focal dermal hypoplasia (Goltz syndrome) is another well-reported condition in which OS is associated with significant clinical symptoms and signs (20–22). Before and since OS with cranial sclerosis was recognized as a distinct clinical entity, there have been descrip-

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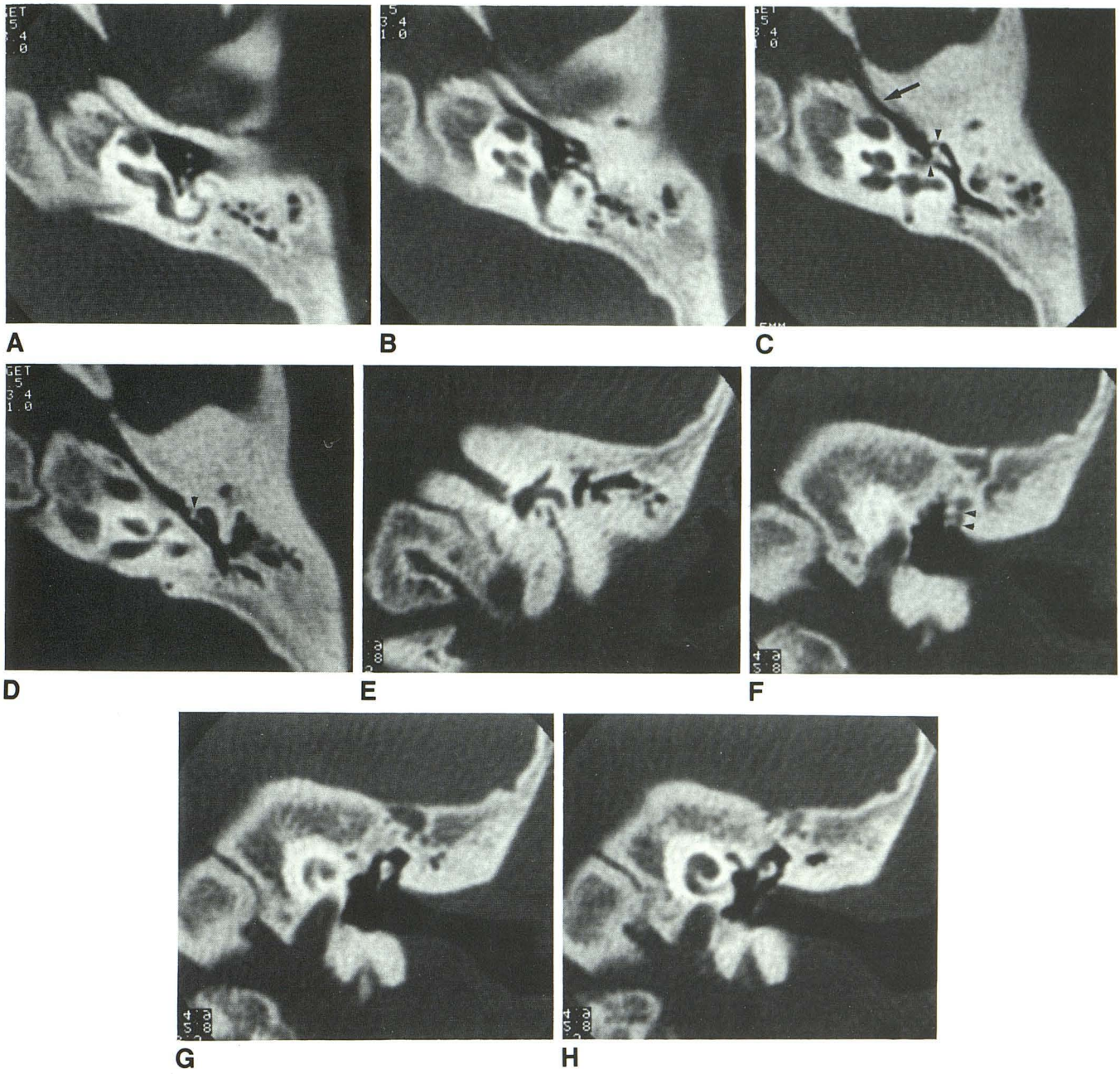

Fig. 1. *A–D*, Axial CT scans of left temporal bone demonstrate thickened and sclerotic bone with small mastoid antrum and middle ear cavity. The ossicles are abnormally fixed anteriorly and medially (*arrowheads*). The Eustachian canal is narrow (*arrow*). Bony labyrinthine inner ear structures appear normal. Identical findings were noted on right.

E–H, Coronal CT scans of left temporal bone demonstrate abnormal ossicular fixation posterolaterally (*arrowheads*). The external auditory canal, internal auditory canal, and descending facial nerve canal are patent. Identical findings were noted on right.

tions of mixed/combined or crossover conditions between OS and osteopetrosis (1, 2, 3, 6, 23) as well as other sclerosing bone dysplasias including osteopoikilosis (1, 2, 23, 25), melorheostosis (23–27), fibrous dysplasia (27), craniometaphyseal dysplasia (Pyle disease) (28), and hyperostosis corticalis generalisata/endosteal hyperostosis (Van Buchem disease) (29, 30). Descriptions of biochemical findings and biopsy changes in patients having OS with cranial sclerosis are scarce (9, 16).

The most striking clinical symptom resulting from the disorder is conductive hearing loss especially at low frequencies, or a mixed deafness. Proposed and previously described findings have included bony atresia or stenosis of the external

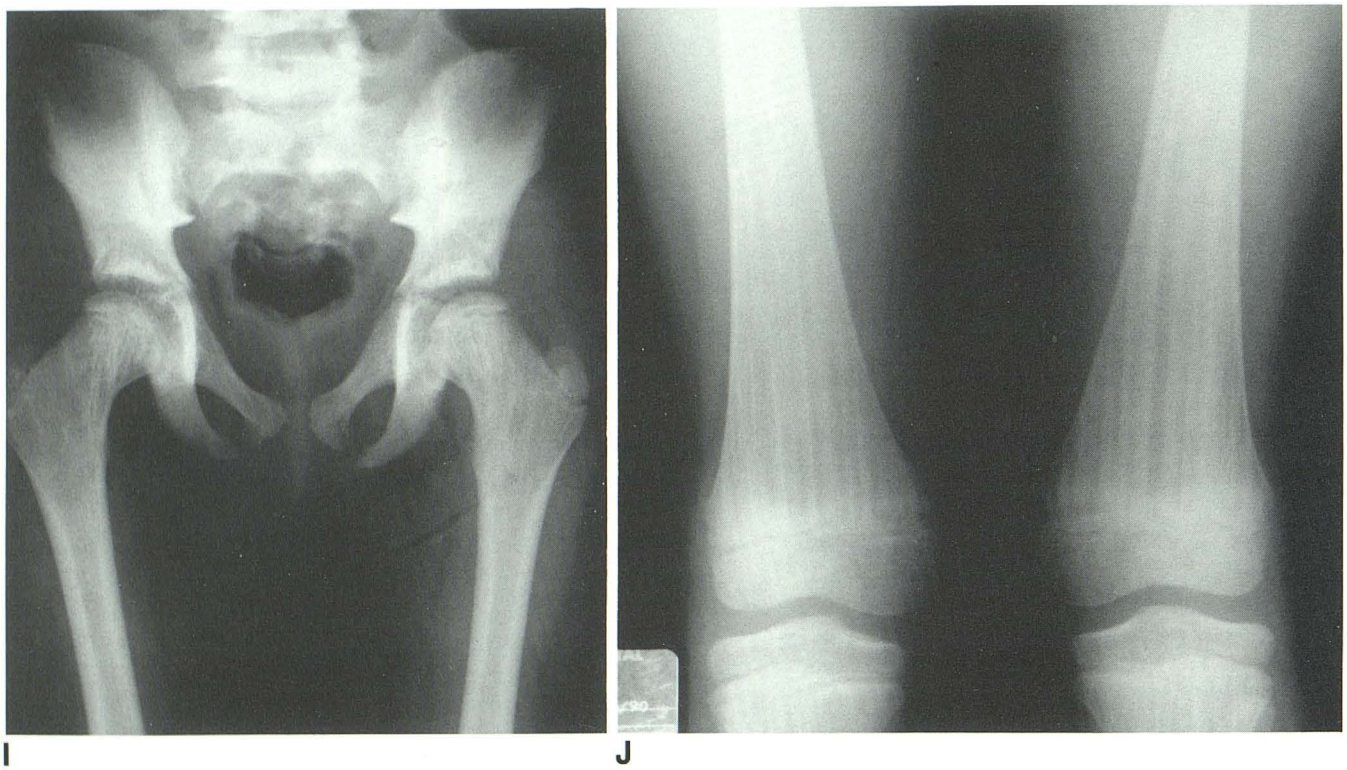

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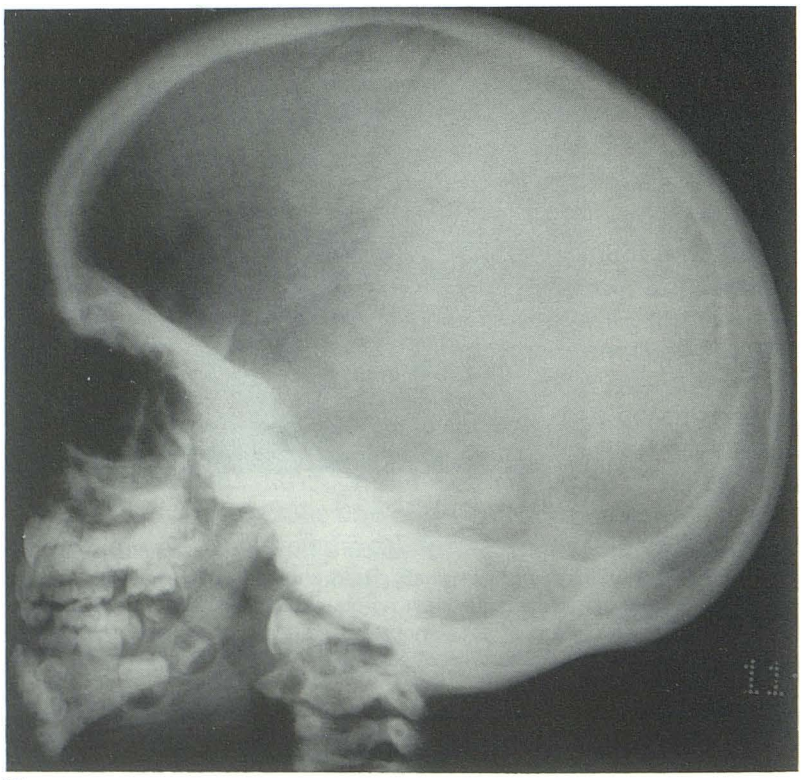
Fig. 1. —*Continued. I–J*, Anteroposterior views of the pelvis and femora demonstrate the longitudinal sclerotic striations typical of OS. The femoral heads are small and there are mild modeling abnormalities of the long bones.

K, Lateral view of skull demonstrates severe sclerosis and thickening especially at skull base.



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auditory canals, middle ear (tympanic) cavities, and internal auditory canals, as well as abnormal ossicular chain fixation. To our knowledge, this is the first reported case in which the diagnosis of sclerosing bone dysplasia was made by CT evaluation for conductive hearing loss and in which CT evaluation corroborated smallness of the middle ear cavities and abnormal ossicular

fixation. This is presumed to be secondary to mural bone overgrowth and encroachment with subsequent impaired mobility of the ossicles. Optic, facial, and trigeminal (maxillary division) cranial nerve deficits have also been described (3, 9, 11, 13).

Conclusion

In conclusion, temporal bone CT is an excellent technique to evaluate and distinguish among the various etiologies of hearing loss in OS with cranial sclerosis. In theory, CT could evaluate a variety of potential cranial nerve deficits in this condition. Moreover, this case underscores that anyone performing temporal bone CT on a regular basis should be aware that conductive hearing loss may be the first clinical presentation of an underlying bone dysplasia. In this way the CT exam can be the springboard for further clinical assessment and radiographic studies which, in combination, may indicate the final diagnosis.

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