

extended from the midramus to ipsilateral canine along the inferior border of the mandible was placed.

#### Acknowledgments

University of Florida College of Dentistry, Shands Hospital

**OSTEOPATHIA STRIATA WITH CRANIAL SCLEROSIS.** R.D. JAGTAP, S. PALACIOS, M. HANSEN, D. KASHTWARI. UNIVERSITY OF FLORIDA, GAINESVILLE, FL

**Background:** Osteopathia striata with cranial sclerosis (OSCS) is a rare x-linked genetic disorder that has variable clinical findings but specific radiographic features, which include marked sclerosis of the long bones and axial skeleton, including the skull. Linear striations, commonly seen in the long bones, are the basis for the name *osteopathia striata*. Maxillofacial manifestations can include a cleft palate, high palatal vault, midface hyperplasia, hypoplastic maxillary sinuses, enlarged alveolar bone processes, temporomandibular joint (TMJ) abnormalities, and dense mandibular bone with variable striations. Dental abnormalities have been poorly documented but include delayed eruption of permanent teeth, missing teeth, short roots, and microdontia.

**Study Design:** A 44-year-old male was referred to the University of Florida College of Dentistry for evaluation and treatment of temporomandibular disorder (TMD). A pantomograph was exposed to assess TMJ dysfunction related to progressive bony overgrowth of craniomaxillofacial skeleton and ankylosis, which had previously been surgically treated. His medical history was significant for OSCS, cleft lip and palate, obstructive sleep apnea, bronchitis, asthma, disk herniation, arthritis, diabetes, and chronic kidney disease. The radiographic assessment was limited because of severe sclerosis of the cranium, displayed as multiple lobulated appearances and homogeneous radiopaque entities superimposed on the maxilla, TMJs, and the sphenoid, mastoid, and temporal bones. The visualized cortical outlines of the maxilla and the mandible appeared dense, thick, and sclerotic.

**Discussion/Conclusions:** OSCS is a rare disorder. Diagnosis can often be challenging because other diseases, such as osteopetrosis, pyknodysostosis, Paget disease of bone, and other sclerosing bone dysplasias, make this difficult to diagnose. Radiologists should be aware of the specific radiographic appearance of this condition to aid in the proper diagnosis and management of these patients.

#### Acknowledgments

The University of Florida Department of Oral and Maxillofacial Surgery

#### References

- 1 Hurt RL. Osteopathia striata: Voorhoeve's disease: report of a case presenting the features of osteopathia striata and osteopetrosis. *Bone Joint J.* 1953;35:89-96.
- 2 Nakamura T, Yokomizo Y, Kanda S, Harada T, Naruse T. Osteopathia striata with cranial sclerosis affecting three family members. *Skeletal Radiol.* 1985;14:267-269.
- 3 Daley TD, Wysocki GP, Bohay RN. Osteopathia striata, short stature, cataracts, and microdontia: a new syndrome? A case

report. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod.* 1996;81:356-360.

- 4 Heung-Chul P, Kim HG, Kim YH, Kim JH, Kim MY, Kim KW. Osteomyelitis in an osteopathia striata with cranial sclerosis patient. *Maxillofac Plast Reconstruct Surg.* 2014;36:285-291.
- 5 Goodman J R, Robertson CU. Osteopathia striata: a case report. *Int J Paediatr Dent.* 1993;3:151-156.
- 5 Gay BB, Elsas LJ, Wyly JB, Pasquali M. Osteopathia striata with cranial sclerosis. *Pediatr Radiol.* 1994;24:56-60.

**OSTEOSARCOMA IN PAGET DISEASE OF BONE.** M. ALMAHNDR, W. DRANE, A. RUPRECHT, D. KASHTWARI. UNIVERSITY OF FLORIDA, GAINESVILLE, FL

**Background:** Paget disease of bone (PDB) is a disorder of bone metabolism that involves osteoclasts. Osteosarcoma arising in Paget disease is a rare complication of PDB occurring in less than 1% of all osteosarcomas. The aim of this case report is to describe different imaging features of osteosarcomas arising in PDB and its progression.

**Clinical and Radiographic Findings:** A 72-year-old female with known arthritis and PDB presented to the emergency room with a 1-week headache and right temporal/parietal skull swelling. The erythrocyte sedimentation rate (ESR) and serum alkaline phosphatase (ALK) were elevated. Planar radiographs of the skull exhibited mixed sclerotic and radiolucent lesions throughout, giving a cottonwool appearance. Multidetector computed tomography (MDCT) of the head showed peripheral enhancement, a partially calcified right temporal region mass, and expansion of the underlying calvarium with a periosteal reaction. Intracranial extension was clear on magnetic resonance imaging (MRI). Furthermore, positron emission tomography/computed tomography (PET/CT) revealed increased fluorodeoxyglucose (FDG) avidity in the solitary right temporal/parietal lesion. Biopsy revealed osteosarcoma with chondroblastic features arising in PDB. The patient underwent right craniectomy and resection of the tumor. Six weeks later, gross recurrent disease at the resection site was found on a head MDCT that demonstrated soft tissue density underlying the cranioplasty with a significant amount of vasogenic edema involving the temporal/frontal lobe. The patient underwent right decompressive and epidural tumor resection. A day later, she was pronounced brain dead.

**Discussion/Conclusions:** PDB is often diagnosed through incidental findings in a regular blood chemistry panel, particularly ALK, or through imaging done for another reason. The planar radiograph gives the best clue to the presence of PDB. The MDCT is mandatory for better outcomes in treatment of osteosarcoma, whereas MRI is best for depicting local intracranial extension. PET identifies the region of a tumor through FDG increase. Lastly, biopsy is necessary for confirming the tumor.

#### References

- 1 Hansen MF, Seton M, Merchant A. Osteosarcoma in Paget's disease of bone. *J Bone Miner Res.* 2006;21:P5863.
- 2 Mancebo-Aragoneses L, Lacambra-Calvet C, Jorge-Blanco A, Coarasa-Cerdan A, Guadaño-Salvadores V. Paget's disease of the skull with osteosarcoma and neurological symptoms associated. *Eur Radiol.* 1998;8:1145.