BENIGN BONE-FORMING TUMORS: APPROACH TO DIAGNOSIS AND CURRENT UNDERSTANDING OF PATHOGENESIS

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KEYWORDS

• Osteoma • Osteoid osteoma • Osteoblastoma • Osteosarcoma

ABSTRACT

he diagnosis of benign bone-forming tumors continues to be based on the traditional approach to bone tumor diagnosis using knowledge of the spectrum of histopathologic features seen in these tumors in combination with clinical and radiological correlation. This review emphasizes the pathologic features and the clinical and radiological features that the surgical pathologist must have a working understanding of to make an accurate diagnosis and avoid confusion with other lesions, particularly osteosarcomas. New and persistent challenges facing the practicing pathologist and our current understanding of the cytogenetic and molecular abnormalities involved in the pathogenesis of these tumors are discussed.

OVERVIEW

It has been imperative that the practicing surgical pathologist has knowledge no only of the histopathologic features of a bone tumor, but also of the clinical presentation and imaging features in order to arrive at a correct diagnosis. The diagnosis of osteoma, osteoid osteoma, and osteoblastoma

continues to be based on this approach. As such, this review focuses on the clinical, radiological, and pathologic features that surgical pathologists must have working knowledge of to accurately classify these tumors as part of



Key Points Bone-Forming Tumors

- The diagnosis of benign bone-forming tumors continues to be based on the knowledge of the spectrum of histopathologic features in combination with clinical and radiological correlation.
- Distinguishing osteoblastomas from osteosarcomas, recognizing pseudosarcomatous change in an osteoblastoma, and working with limited tissue in trying to diagnose osteoid osteomas are still the greatest challenges for the pathologist.
- There is limited understanding of the cytogenetic and molecular abnormalities that result in the pathogenesis of benign bone tumors and that could be used for ancillary diagnostic studies.

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a multidisciplinary approach. In addition, the authors comment on challenges and practical issues pertaining to making a diagnosis and highlight new developments in the management of these neoplasms that impact the practicing pathologist. The authors also discuss the current understanding of the molecular and cytogenetic abnormalities that pertain to the pathogenesis of these neoplasms and that may one day serve as ancillary diagnostic tools.

OSTEOMAS

Skeletal osteomas are benign tumors composed of dense cortical bone that arise on the surface of bones. They generally can be placed into 2 major categories: (1) sinonasal/skull osteoma and (2) long bone osteoma (parosteal osteoma). The exact incidence of osteomas is uncertain because most cases are incidentally discovered. The incidence of sinonasal osteomas has been reported to be 3% in patients undergoing computerized tomography (CT) imaging for sinonasal symptoms. Long bone osteomas are exceedingly rare.

LOCATION OF OSTEOMAS

Sinonasal/skull osteomas usually arise on the surfaces of the cranial vault, jaw, and orbit, or within the paranasal sinuses where they most frequently involve the frontal sinus.^{1,4} Up to 86% of long bone osteomas arise in the lower extremity but may also

affect the humerus and vertebral column.^{3,5} Osteomas are typically isolated lesions, but multiple osteomas can be a manifestation of Gardner syndrome.^{2,4,6}

CLINICAL FEATURES OF OSTEOMAS

Osteomas affect all age groups, but are most commonly diagnosed in adults around the fourth and fifth decade of life. The male-to-female ratio has been reported to range from 1.7 to 2.6: 1.0 for sinonasal osteomas. There is not a definite sex predilection of long bone osteomas. They are often asymptomatic; but if they are larger and situated in a vital location, osteomas can cause sinusitis, visual disturbances, spinal cord compression, otitis media, and headaches. Osteomas of the pelvic bones and long bones often present with pain. 9.

RADIOLOGICAL FEATURES OF OSTEOMAS

On plain films or CT imaging, an osteoma appears as a round to lobular, homogeneous radiodensity with a smooth contour on the cortical surface usually measuring less than 3 cm with no involvement of the underlying bone (Fig. 1). 1.3 In the sinonasal region, osteomas tend to form polypoid intracavitary growths. Some sinonasal osteomas may have central areas of radiolucency that correlate with microscopic features indistinguishable from osteoblastomas. 10 In long bone, it can be



Fig. 1. CT of an osteoma of the frontal sinus demonstrates characteristic homogeneous density of cortical bone, smooth contour, intracavitary growth, and no involvement of the underlying bone. The central areas of lucency correspond to cancellous bone or osteoblastoma-like areas within the tumor.

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