19. Osteosarcomatosis and metastatic osteosarcoma


Definition and classification of osteosarcomatosis

Multiple secondary osseous foci that appear within 5 months after the diagnosis of symptomatic, radiographically dominant tumor are classified as synchronously appearing multifocal osteosarcoma, so-called osteosarcomatosis (Figure 19-1). Because of its rapid onset and generally symmetric distribution, several theories have been proposed as to the origin of osteosarcomatosis. Several authors (1–6) have theorized that these multiple skeletal lesions represent multiple primary osteosarcomas of varying sizes and occur from the “multipotent preosseous tissue of the periosteum” [1]. Associated primitive preosseous rests [7] and humoral and cell-mediated immunity induced by specific neoplastic antigens [8,9] have also been proposed as possible etiologies. However, other authors [9–13] have suggested that these multiple synchronously appearing bone lesions actually represent rapidly appearing metastases.

Three separate systems have been proposed to classify osteosarcomatosis [2,9,14]. These systems are compared in Table 19-1. The classification system proposed by Amstutz [14] covers all aspects of multifocal osteosarcoma and appears to be the most commonly used in the medical literature. Therefore, we chose this system to evaluate the secondary osseous foci found among 690 cases of osteosarcoma in the radiographic files of the Armed Forces Institute of Pathology (AFIP) in Washington, D.C. This system divided osteosarcomatosis into types I and II (synchronously appearing less than 5 months after diagnosis of a symptomatic, radiographically dominant primary) and metastatic osteosarcoma types IIIa and IIIb (metachronously appearing over 5 months after the diagnosis of a symptomatic, radiographically dominant primary). The major difference between Amstutz type I and type II osteosarcomatosis is the age of the patient at the time of initial diagnosis. Amstutz type I osteosarcomatosis includes those patients 18 years and under, and type II includes all patients over 18 years of age. Amstutz type IIIa (early metachronous metastatic osteosarcoma) includes any patient (regardless of age) with secondary skeletal lesions occurring more than 5 months and up to 24 months after diagnosis of a primary osteosarcoma. Amstutz type IIIb (late metachronous metastatic osteosarcoma) includes any patient (regardless of