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Review

Radiopathological evaluation of primary malignant skull tumors: A review

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ABSTRACT

Skull tumors comprise a wide variety of entities, ranging from chronic inflammatory disease to primary and secondary neoplasms. There is no valid incidence or data about the incidence of skull tumors in general. Primary malignant skull tumors are rare, with most articles reporting single cases. We would discuss some of the frequent tumors in this group and review of the literature for the same.

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1. Introduction

Metastatic carcinomas are the most common malignant tumor affecting the skeleton. Most metastases originate from common cancers namely breast, lung, prostate, kidney and thyroid gland, which account for 93% of all deposits. Out of 114 histologically evaluated lesions 44.3% involved axial skeleton, 28.8% the appendicular skeleton and 26.9% involved multiple bones [1].

Among the wide array of human neoplasms, primary tumors of bone are relatively uncommon. Not only has this contributed to the paucity of meaningful and useful data about the relative frequency and incidence rates of the various subtypes of bone tumors, but it also explains our rudimentary understanding of risk factors. In general, bone sarcomas account for only 0.2% of all neoplasms [2].

There is a wide variety of histopathological diagnosis in tumors of the skull. These neoplasms comprise tumors of bony, cartilaginous, fibrous, histiocytic or hematopological origin. The most common malignant skull tumors are osteogenic sarcoma and chondrosarcoma. The clinical presentation varies according to the site of tumor origin. However the most common symptom is painless, slowly growing epicranial mass, which may vary widely regarding size and rate of growth. Tumors of skull base may present with

Abbreviations: MDCT, multi detector computed tomography; NCCT, non contrast computed tomography; H&E, hematoxylin & eosin; SIP, Solitary intramedullary plasmacytoma; SPS, solitary plasmacytoma of the skull; CECT, contrast enhanced computed tomography; HPE, histopathological examination; MR, magnetic resonance; CNS, central nervous system; CT, computed tomography.

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cranial nerve symptoms like diplopia, visual or hearing loss, olfactory sensations, or impaired swallowing function. [3]

2. Discussion

2.1. Osteosarcoma

In a survey of the literature around 100 cases of primary osteosarcoma of the skull were found, including our cases. The occurrence of osteosarcoma in the craniofacial bones peaks in the third decade, where as that in the skeleton peaks in the second decade. The etiology of osteosarcoma is unknown, but the major risk factors for development of osteosarcoma in craniofacial bones may be similar to those of the long skeletal bones, consisting of exposure to radiation, retinoblastoma, Li–Fraumeni syndrome, and Paget's disease. The skull is a favored site for osteosarcoma arising out of Paget's disease. Other bone abnormalities, such as fibrous dysplasia, multiple osteochondromatosis, chronic osteomyelitis, myositis ossificans, and trauma, have also been proposed as risk factors [4].

Radiologically osteosarcomas commonly have osteolytic and osteosclerotic features, a cloudy pattern of mineralization either diffuse or in clusters, and illdefined borders. Radiating striations (sun-burst appearance) and Codman's triangle are signs of periosteal reaction due to periosteal elevation [5]. These can be seen in periosteal osteosarcomas and intramedullary osteosarcomas with cortical extension, but not in paraosteal osteosarcomas. High grade surface osteosarcomas tend to have a more immature and basal pattern of mineralization. Paraosteal osteosarcomas are usually dense sessile masses on the bone surface, with smooth or irregular margins. A thin radiolucent line may separate part of the tumor from the cortex. CT and MR scanning provide further details regarding tumor structure and extent, namely to adjacent soft tissue, bone itself and neurovascular structures. A chest CT scan and a bone scintigraphy can be used to evaluate the existence of lung and skeletal metastases respectively [6].

The differential diagnosis of cranial osteosarcoma to be considered includes chondrosarcoma and osteochondroma. The CT findings of new bone formation in the soft tissue mass and the characteristic matrix strongly suggest osteogenic sarcoma, but differentiation from chondrosarcoma and osteochondroma cannot be made definitely without histology. Computed tomography in cases of chondrosarcoma of the base of skull shows similar changes, but once the calvarium undergoes membranous ossification, it becomes an unlikely site for cartilaginous tumors. Osteochondromas usually are sharply defined and homogenous indensity [7].

Pathologically the diagnosis of osteosarcoma is predicated on the accurate identification of osteoid. Histologically, osteoid is a dense, pink, amorphous intercellular material, which may appear somewhat refractile [8].

Surgery and chemotherapy have been recommended for the management of skull osteosarcoma [9]. The identification of prognostic factors has been an additive process in which factors have been investigated, identified and incorporated into an overall therapeutic strategy [10]. Traditionally, age, gender, location, tumor size, stage, and the results of various laboratory tests have been used in an effort to predict prognosis. However, response to pre-operative therapy is currently the most sensitive indicator of survival. At the same time, it is recognized that a single system does not apply to all cases. Unique biological aggressiveness, coupled with an inability to completely resect the tumor at certain sites (e.g., skull, spine) is one example (Figs. 1 and 2).

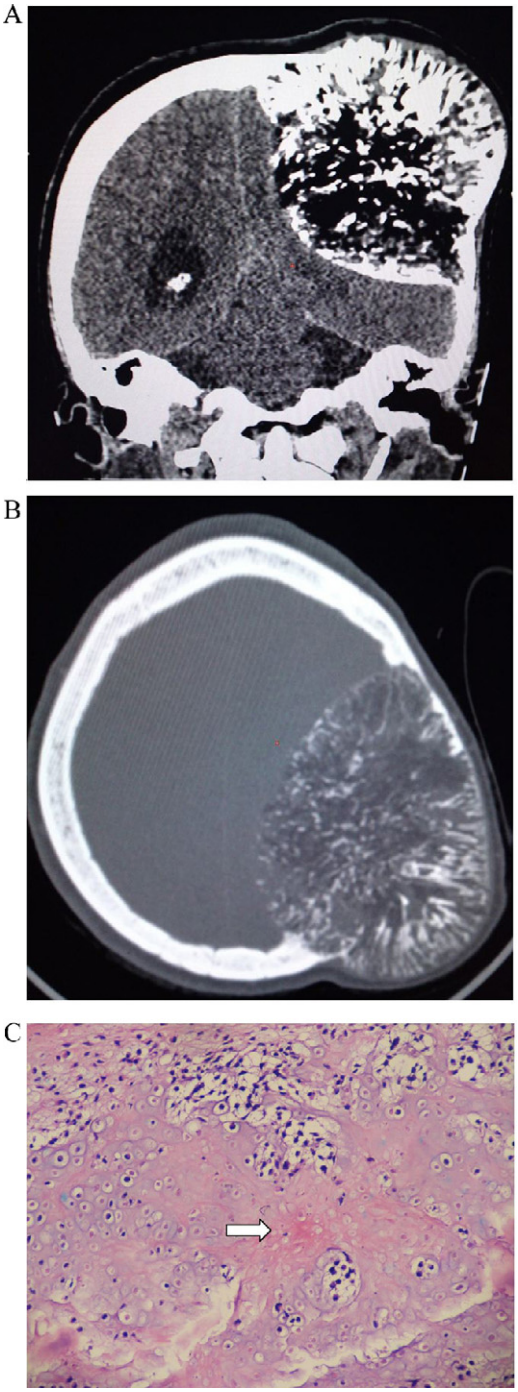

Fig. 1. (a) Coronal NCCT and (b) axial bone window settings of cranium showing a giant geographic sclerotic/lytic lesion involving parietal bone on left side with dense matrix mineralization (sunburst appearance) showing patchy central hypodense necrotic foci with gross mass effect over left cerebral hemisphere (parietal lobe) and large outer extracranial scalp portion. (c) H&E of chondroblastic osteosarcoma showing lobules of malignant-appearing cartilage with bone formation (arrow) in the center of the lobules.

2.2. Plasmacytoma

Solitary intramedullary plasmacytoma (SIP), a sub-type of plasma cell tumors, is a malignant neoplasm that may occur as a primary focal entity or as the first presentation of systemic multiple myeloma. Plasma cell tumors are classified as either multiple myeloma (systemic), intramedullary plasmacytoma (involving bone), or extramedullary