



Rosette Forming Osteosarcoma

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Abstract

Osteosarcomas of the jaws are not uncommon entities but unusual histological variants do cause difficulties in the differential diagnosis. A recently described histological variant of osteosarcoma is the rosette forming osteosarcoma which bears close similarity to round cell tumors leading employment of a battery of immuno histo chemical markers despite the fact that osteosarcomas are best diagnosed by routine sections. In this report two cases of osteosarcoma with rosette forming features are illustrated for early recognition of this entity.

Keywords: Osteosarcoma; Round cell tumors; Rosette

Introduction

Osteosarcoma is a malignant primary bone tumor characterized by tumor osteoid and immature trabeculae of bone. It presents microscopically in various patterns in addition to the conventional variants. Osteosarcoma with rosette-like pattern is an extremely uncommon variant and is poorly recognized. The purpose of this report is to describe two jaw osteosarcomas characterized by rosette-like pattern.

Case Presentation

Case 1

A 52-year old female presented to the emergency department with a rapidly growing swelling of 1 month duration in the left mandible associated with the complaints of frequent bleeding. Her past medical and dental history was not significant. On examination, a pedunculated mass of 6 cm x 5 cm was found in the left retromolar region of the mandible. The mass was firm, non-tender and showed surface ulceration covered by a pseudo membrane which bled profusely on provocation. Panoramic radiograph showed an ill-defined osteolytic lesion in body of the mandible. As there was significant bleeding associated with the mass, a Computed Tomography scan (CT) was performed. The CT revealed a vascular lesion with feeder vessels from the lingual artery and drained by common facial and internal jugular vein. An emergency operation was performed after ligation of the external carotid artery and the left mandible was resected along with the tumor mass which was easily separable from the bone. The gross specimen was firm and the cut surface was whitish yellow in colour. Multiple soft tissue bits were processed routinely and 3.5 to 4 micron thick sections were evaluated.

On low power microscopy, the tumor mass appeared solid cellular proliferations (sheets and nodules) with interspersed vascular channels resembling hemangiopericytoma pattern associated with sparsely calcified matrix (Figure 1A). On higher magnification, the tumor cells ranged in size from small blue cells to large epithelioid cells and as well as vacuolated or clear cells. Some of the large epithelioid cells were plasma cytoid in appearance and also showed nuclear grooving. The tumor cells were characteristically arranged around eosinophilic, lace-like osteoid matrix with occasional fine calcification in a rosette-like- or hobnail pattern (Figure 1B). Multinucleated giant cells, atypical nuclei and normal mitotic figures were evident but chondroid differentiation was absent. The microscopic differential included osteosarcoma, Ewings sarcoma, peripheral neuroectodermal tumor, mesenchymal chondro-sarcoma, and metastatic carcinoma. Pertinent commercially available immunohistochemical markers such as cytokeratin, epithelial membrane antigen, vimentin, CD99, neuron specific enolase and CD56 were employed to narrow the differential, but these were negative except for focal reactivity with CD56 (Figure 1C). The immediate post-operative period was uneventful but before the pathology report issued the patient died and no autopsy was performed.

Case 2

A one-year old male infant presented with a one-month history of a rapidly growing swelling

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Received Date: 16 Aug 2018

Accepted Date: 15 Sep 2018

Published Date: 26 Sep 2018

Citation:

Ponniah I. Rosette Forming Osteosarcoma. Clin Oncol. 2018; 3: 1528.

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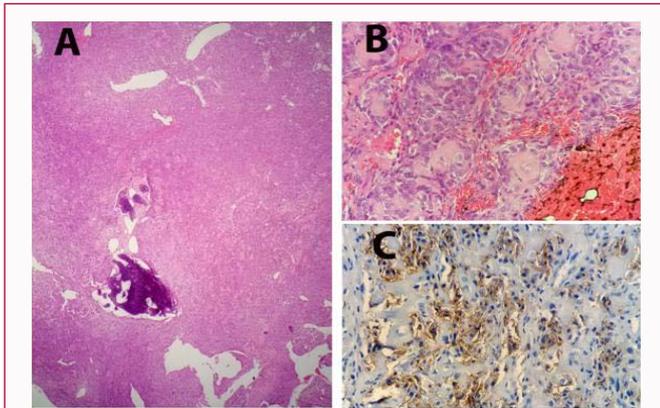


Figure 1: Shows greater part of the tumors cells with focal calcification and prominent hemangiopericytoma-like pattern in A and higher magnification of the tumor cells arranged in a characteristic rosette-like pattern around lace-like osteoid matrix in B. C, lesional cells are positive to CD 56. H & E x4, x40, CD56 x40.

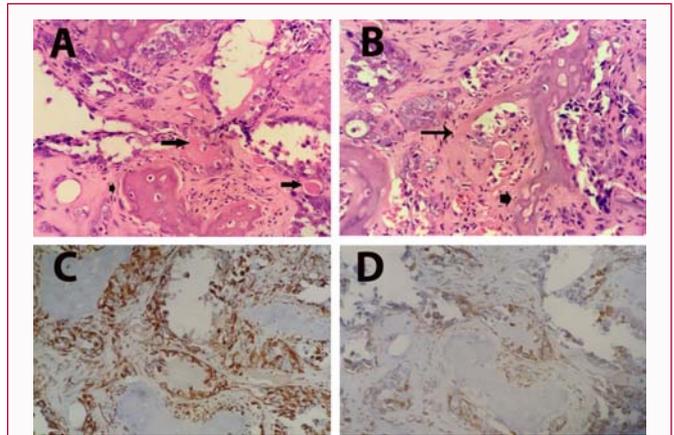


Figure 2: The photomicrograph in A) shows epithelioid appearing tumor cells with fibrillar matrix (arrows) and immature trabeculae of bone (arrow head). B) shows clusters of epithelioid appearing tumors cells with loss of cohesion and lace-like osteoid (arrow) and immature trabeculae of bone (arrow head). C & D) shows positive reaction of tumor cells to vimentin and NSE. H & E x40, Vimentin x40 and NSE x40.

of the right lower jaw. No significant medical history could be elicited for the infant or the mother. On examination, a firm swelling of 3 cm x 2 cm in size was noted in the region of the right mandibular incisor to the left mandibular first molar with expansion of buccal and lingual cortical plates. CT showed an osteolytic lesion in the left parasymphysis region with expansion of the buccal cortical plate. The clinical differential diagnosis included central giant cell granuloma and desmoplastic fibroma. An incisional biopsy was obtained for histopathological examination. Microscopic examination revealed a cellular tumor characterized by clusters of epithelioid cells and focal spindle cells interspersed between immature trabeculae of bone and lace-like osteoid matrix. In some regions the tumor cells were peripherally arranged over a central fibrillar matrix in a rosette-like pattern with loss of cohesion of the cellular tumor clusters (Figure 2). The microscopic differential included small round cell tumors and epithelioid osteosarcoma. A panel of immunohistochemical markers were employed which included cytokeratin, EMA, S100, vimentin, α SMA, desmin, CD99, LCA, TdT, CD34, NSE, CD56, synaptophysin and chromogranin. None of the markers were positive except vimentin and NSE (Figure 2). Urinary vanillylmandelic acid and homovanillic acid were within normal range. The lesion was finally categorized as epithelioid osteosarcoma with rosette-like features. Induction chemotherapy was initiated and lesion responded well to the chemotherapy.

Discussion

Osteosarcoma is a primary intra medullary high-grade malignant tumor in which the neoplastic cells directly produce osteoid. It can affect patients of all ages, in children less than 5 years of age to adults in the 10th decade of life [1]. However, it represents only 2% of patients with osteosarcoma fall under 5 years of age, while greater majority occur in the first two decades of life [1]. Osteosarcoma slightly involves older age group when it occurs in the craniofacial region [1]. Osteosarcoma of the jaw bones constitutes 5% to 13% of all cases of skeletal osteosarcoma with predilection for the mandible, where the body is the most common location followed by angle, ramus and symphysis, while the alveolar ridge is the most commonly affected site in the maxilla [1]. Radio graphically; osteosarcoma has an aggressive appearance with extensive bone destruction and propensity for extension into the soft tissues. It usually manifests as lytic, blastic or mixed density lesion on roentgenogram. Histologically,

osteosarcoma is classified as osteoblastic, chondroblastic, fibroblastic depending upon the presence of malignant osteoblasts with osteoid matrix, chondroid matrix and fibrous spindle cell areas. The latter type generally lack osteoid or tumor bone and require examination of many sections to find the diagnostic osteoid, if it is only in smaller quantity [1]. Other rare histological variants include telangiectatic, chondroblastoma-like, clear cell, small cell, epithelioid, and malignant fibrous histiocytoma-like. Recently, Okada et al. [2] described from his retrospective analysis of osteosarcoma that certain osteosarcoma resemble rosette-forming tumors. These osteosarcomas were designated as rosette-forming osteosarcoma, which are rare high grade epithelioid appearing osteosarcoma that characteristically show central, amorphous, eosinophilic nidus around which tumors cells are arranged in a rosette-like pattern [2]. In contrast, the conventional osteosarcomas usually lack osteoblast rimming and the tumor cells are present in the intertrabecular spaces.

Review of the literature regarding epithelioid osteosarcoma or rosette-forming osteosarcoma reveals that the epithelioid osteosarcoma reported by Rinaggio et al., [3] was made up of sheets of epithelioid cells with eccentrically placed nucleus resembling plasmacytoid cells and foci of osteoid matrix, whereas the case reported by Kaveri et al., [4] was characterized by sheets of epithelioid and spindle cells amidst zones of osteoid matrix. In comparison, others have reported epithelioid osteosarcoma with plasmacytoid features in a trabecular or rosette-like pattern [5-7]. In the latter pattern, in only two case reports, [5,7] the tumors cells were either arranged around fibrillar or osteoid matrix.

In the present report, the tumor in the adult patient was consistent with the rosette-forming osteosarcoma described in the literature, [2,5,7,8] whereas the pediatric case manifested epithelioid cells amidst lace-like osteoid or immature bone trabeculae and also characteristically showed cells around fibrillar matrix in a rosette-like pattern, with apparent loss of cohesion of the tumor cells giving an appearance of alveolar pattern. Therefore, the latter case is neither typical of the rosette-like osteosarcoma nor the epithelioid osteosarcoma described by others [2-8]. Further, the immuno histo chemical findings did not support the round cell tumors or metastatic disease. Therefore, the presence of lace-like osteoid matrix and

immature bone trabeculae and reaction to vimentin and NSE are consistent with osteosarcoma regardless of sub typing.

In a review of 131 cases of osteosarcoma excluding the jaw bones, only 14% (18 cases) manifested the rosette-forming pattern on routine sections. This variant of osteosarcoma typically lacks the prominent osteoid matrix elaborated by the tumor cells in conventional osteosarcoma [2,5,7]. However, osteoid matrix can still be discernible in at least focal regions of the tumor. According to Okada et al. [1], the rosette forming variant usually express EMA, CD56 and CD99, but the literature reveals no clear cut immuno profile of tumors reported either as epithelioid osteosarcoma or rosette forming osteosarcoma [2,3,5-7]. Therefore, the diagnosis can easily be identified on the routine sections based on the morphological appearance of the tumors cells around amorphous, eosinophilic, osteoid matrix, with or without the presence of fine granules of calcifications associated with hemangiopericytoma-like pattern [2,9]. Surgery is the standard treatment modalities for osteosarcoma of the jaw with or without adjuvant radiation or chemotherapy [10]. However, the biologic behaviour of osteosarcoma of the jaws differs from osteosarcoma of long bones [11]. The prognosis of rosette-forming osteosarcoma is considered to be dismal even when neoadjuvant chemotherapy is effective.

In conclusion, the present report emphasize the early recognition of osteosarcoma with rosette-like feature is important in view of its aggressive nature and the potential to lead to a diagnosis of primary or metastatic neural neoplasms of bone.

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