A Rare Case of Extraskkeletal Mesenchymal Chondrosarcoma with Dedifferentiation Arising from the Buccal Space in a Young Male

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Abstract Extraskkeletal mesenchymal chondrosarcoma (EMCS) is a rare and aggressive pathological variant of chondrosarcoma arising from soft tissues of mainly the extremities, meninges/dura, trunk, and orbits. EMCS comprises only 2 % of all soft tissue sarcomas and only 6 % of them arise from soft tissue in the head and neck region. It usually affects in the second and third decade of life and is common in women. It runs a very rapid clinical course with distant metastases and has poor prognosis and survival rates. Histologically it has a dimorphic presentation of small round mesenchymal cells interspersed with foci of cartilaginous differentiation. Radiographically it appears as a soft tissue lobulated mass with various patterns of calcification. This is the first case of primary and recurrent EMCS originating in the buccal space with unusual features of dedifferentiation. The diagnostic challenges in this case were the inconclusive FNAC results,

lack of radiographic evidence of characteristic calcifications and presence of areas of myxoid material and dedifferentiation on histopathology. The clinical, cytological, histological, immunohistochemical, radiographic, PET-CT findings and management with review of literature is presented. The diagnostic and management pitfalls of this extremely rare tumor are also discussed.

Keywords Extraskkeletal mesenchymal chondrosarcoma · Myxoid · Cell dedifferentiation · Radiographic imaging · Calcification · PET CT

Introduction

Chondrosarcomas are a heterogeneous group of malignant neoplasms that produce cartilaginous matrix. Conventional chondrosarcomas are low to intermediate grade tumors usually involving the extremities, pelvis, scapula and ribs. Mesenchymal chondrosarcoma is a rare, and high grade pathological variant of conventional chondrosarcoma that was first described by Lichtenstein and Bernstein in 1959 [1]. About 30–50 % of all mesenchymal chondrosarcomas are extraskkeletal in origin [2]. However, extraskkeletal chondrosarcomas comprise only 2 % of all soft tissue sarcomas and mesenchymal variant constitutes about 1 % of all chondrosarcomas [3].

EMCS has characteristic clinical and dimorphic histological picture [4]. It is more commonly seen in women in the second and third decade of life and is a rare aggressive tumor with rapid clinical course, widespread metastasis and poor prognosis and survival rates. It is commonly found in the extremities followed by meninges/dura, trunk and orbits. Only 6 % of EMCS are found originating from other areas in the head and neck region [4, 5].

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This is a rare case of primary and recurrent EMCS originating from the buccal space in a young male showing unusual features of dedifferentiation. The clinical, cytological, histopathological, immunohistochemical, radiological and PET features of this aggressive tumor with management and review of current literature is presented. The diagnostic challenges due to rare radiographic and histopathologic features seen in this case are also discussed.

Case Report

A 21 year old male, reported with a swelling in the left cheek region since past 15 days. He gave a history of trauma with a cricket ball in the region 6 months previously. The swelling in the left buccal space was well circumscribed (4 cm × 3 cm), mobile and non-tender on palpation. The overlying skin was not indurated and was normal in color and temperature. Intra oral examination revealed a lobular well circumscribed swelling in the maxillary buccal vestibule with ulcerated mucosal surface (Fig. 1). There was no bleeding from the ulcerated area, and the adjacent teeth were non-tender and non-mobile. There was no cervical lymphadenopathy and no odontogenic cause for the swelling. The patient had no other systemic complaints. Informed written consent was obtained from the patient before further investigations. Routine hematological and serological investigations (liver function, kidney function, bone chemistry) were normal.

An orthopantomogram showed a well circumscribed homogeneously radio- opaque lesion over the left maxillary alveolar process with no evidence of bony involvement. The walls of the left maxillary antrum were intact on the orthopantomogram and para nasal sinus views. A CECT of the jaws and neck revealed a well defined lesion in the left buccal space with thick enhancing wall and both cystic

Fig. 1 Intraoral swelling in buccal vestibule with ulcerated mucosa

Fig. 2 CT image showing well defined soft tissue mass without any calcifications in primary tumor

and soft tissue attenuation. No evidence of bony involvement was seen and no evidence of calcification was noticed within the lesion (Fig. 2). There were few sub centimeter enlarged Level II lymph nodes on the left side. A fine needle aspiration cytology of the swelling revealed many stromal fragments and round to plasmacytoid cells with prominent nucleoli, scattered singly and in clusters, suggesting the possibility of skin adnexal tumor or pleomorphic adenoma.

An incisional biopsy was urgently performed under local anesthesia. The histopathological sections showed tumor tissue composed of numerous round primitive mesenchymal cells arranged in lobules, having basophilic nuclei. High mitotic activity and numerous mitotic figures (6–8 cells per HPF) were noted. Large amount of mucous material was seen underlying the cellular areas. Areas of cartilaginous differentiation with pleomorphic chondroid cells having numerous mitoses, few bizarre mitotic figures and binucleation were observed. Foci of myxoid degeneration and dedifferentiation were also noted (Figs. 3, 4). A fibrous capsule was seen at the periphery in most parts. There was positive staining of the chondroid material with Alcian Blue (Fig. 5). Immunohistochemical staining showed strong positivity for mesenchymal marker vimentin, focal positivity for S100 but was negative for CD99, epithelial, lymphoid, neural, endothelial and muscle markers. Correlating the clinicopathological and radiological findings a diagnosis of EMCS with dedifferentiation was made.

The patient sought opinion at a different hospital where the tumor was excised. The patient reported with a recurrence of the tumor at the same site within a month. In view of the aggressive behavior of the tumor, further investigations like contrast enhanced MRI of the face and neck and whole body PET-CT were carried out. MR imaging

Fig. 3 Cellular tumor with clear and myxoid areas and areas of dedifferentiation (Hematoxyline and Eosin $\times 10$)

Fig. 4 Malignant mesenchymal cells with focal cartilaginous differentiation (Hematoxyline and Eosin $\times 40$)

showed a large lobulated heterogeneously enhancing mass measuring approximately $49 \times 32 \times 47$ mm appearing hypointense on T1W and heterogeneously hyperintense on T2W/STIR images in the left buccal space (Fig. 6). The lesion extended up to the retromolar trigone posteriorly and displaced the buccinators muscle laterally with subtle indistinct fat planes. Medially the lesion was infiltrating the maxilla with extension of the mass superiorly into the left maxillary sinus through the destruction of the posterolateral and inferior wall of maxillary sinus. Few erosions were seen in the posterolateral and anterior wall of left maxillary sinus with associated periosteal reaction. Mucosal thickening was seen in the left maxillary sinus. A small area showing flecks of calcification was noted within the lesion on CT, which corresponded with hypointense areas on MR images (Fig. 7). PET-CT revealed abnormal intense hypermetabolic FDG uptake in mildly heterogeneously

Fig. 5 PAS with Alcian Blue stain shows bluish chondroid matrix ($\times 40$)

enhancing hypodense lesion in left buccal space (Fig. 8). There was physiological uptake of the tracer in bilateral cervical, supraclavicular and paravertebral region. Physiological uptake of the tracer was also noted in the heart, liver, lungs, renal, pelvicalyceal systems, urinary bladder and the gut.

The patient then underwent left subtotal maxillectomy (sparing the left orbital rim and floor) with split thickness skin graft from the left thigh to cover the mucosal defect

Fig. 6 Heterogeneous hyperintense mass on T2 weighted MR image

Fig. 7 Subtle calcifications seen in recurrent tumor mass on CT

Fig. 8 Intense hypermetabolic FDG uptake on PET-CT scanning

via modified Weber Fergusson incision with lateral extension. The histopathologic margins of the excised lesion were free of the tumor. Post-operative recovery of the patient was uneventful (Fig. 9). The patient was then treated with radiotherapy using IMRT technique with trilogy linear accelerator (total dose 58.52 Gy). Oral rehabilitation with obturator was provided in the interim period. There has been no recurrence in the 9 months post-radiotherapy.

Discussion

EMCS is usually seen in patients in the second and third decade (15–35 years of age) and very rarely seen in children. The common site of primary tumor in the head and neck region are intracranial, orbits and sinonasal tract [4]. Isolated reports of other areas like nasal cavity, eyelid, parapharyngeal space, ethmoid sinus and thyroid have been

Fig. 9 Post-operative intraoral view of surgical site

reported [6–10]. We could not find any reports of EMCS arising in the buccal space (PUBMED/MEDLINE). It has two peak age incidences in adults, 23.5 years for intracranial involvement and 43.9 years for soft tissue and/or muscular involvement [11]. This report is a rare presentation of primary EMCS in a young male patient primarily involving the left buccal space. The recurrent tumor was aggressive and spread to involve the left maxillary sinus. The etiological factors are unknown but accidental or surgical trauma has been implicated as in most sarcomas. In this case the patient also gave a history of trauma as he was hit by a cricket ball to the left side of the face 6 months previously.

The cytological features of EMCS on FNA are less well documented. The smears are cellular consisting of small, spindle or polygonal cells with high nuclear cytoplasmic ratios embedded in a dense metachromatic matrix [4, 12]. Doria et al. reported the first FNA report from retroperitoneal EMCS and described the two cell population of the tumor comprising of a poorly differentiated monomorphic, small cells and malignant chondroid component. A myxoid matrix was also seen by them as was also reported by Trembath et al. [12, 13]. The differential diagnoses based on FNA findings would include Rhabdomyosarcoma, Ewings sarcoma, Hemangiopericytoma, Synovial sarcoma and other variants of chondrosarcoma as these also exhibit small round blue cells [12]. The diagnosis of EMCS on the basis of FNA is extremely challenging; this is because of the difficulty in sampling both the phases of the tumor, lack of distinctive clinical and radiographic features and its rarity. Buchon et al. [14] suggested wide sampling of the

tumor mass so as to detect the cartilaginous component that is diagnostic of EMCS. Ancillary studies like immunohistochemistry can be useful as EMCS is positive for S-100, CD99 and vimentin [4]. Trembath et al. suggest “*the triple test*” approach to diagnosis as used in breast cytology where in the clinical, imaging, cytopathology or histopathology features should be used to make a definitive diagnosis [12]. Early diagnosis by FNA can be useful in early treatment of EMCS as they are aggressive in nature. The cytological findings in this case were not conclusive and histopathological evaluation was required.

The most characteristic histological feature of EMCS is the dimorphic pattern composed of sheets of primitive undifferentiated mesenchymal cells interspersed with islands of mature cartilage. Central calcification and ossification are frequently seen. The cells are round, oval or spindle shaped with ovoid or elongated hyperchromatic nuclei and scanty poorly outlined cytoplasm. The cells are sometimes seen in aggregates around sinusoidal vascular channels lined by single layer of endothelium which is described as hemangiopericytoma-like pattern. The cartilaginous areas are sometimes poorly circumscribed blending in gradually with the undifferentiated tumor cells [4].

The cartilaginous areas are strongly positive for S-100 protein while focal positivity may be seen in the undifferentiated areas. S100 positivity is not specific for chondrosarcomas and may be seen in other tumors like melanomas, malignant peripheral nerve sheath tumors, schwannomas, histiocytoma and others. The undifferentiated cells may also show positivity for CD99 and neuron-specific enolase and Leu-7. Scattered positivity may also be seen for desmin, myogenin or myoD1. Actin, cytokeratin and EMA are usually negative [15, 16]. In the absence of cartilaginous foci the histological picture is very similar to other small round blue cell tumors like ES/PNET, synovial sarcoma and rhabdomyosarcoma. Hoang et al. have found Sox9, a transcription factor thought to be a “master regulator” of chondrogenesis to be consistently positive in both the cartilaginous and primitive mesenchymal cells in EMCS thus helping in differentiating it from other small round blue cell tumors [17].

In this case the typical dimorphic histological picture was helpful in arriving at the diagnosis of EMCS. The large amounts of mucoid material and focal areas of myxoid degeneration seen posed a diagnostic dilemma as the tumor had to be differentiated from extraskeletal myxoid chondrosarcoma especially the cellular variant which is also hypercellular with less myxoid stroma. However the tumor cells in the myxoid variant are large with vesicular nuclei and prominent nucleoli unlike EMCS [4]. Areas of dedifferentiation with EMCS is very rare, reported before in few cases and associated with poor survival rates [18]. Dedifferentiation into anaplastic lesions may be seen in 11 % of chondrosarcoma.

Dedifferentiation in parent tumor is usually responsible for accelerated growth and aggressive nature of the tumor as was seen in this case. It also implies more wider or radical margins during surgical resection to prevent local recurrence.

Conventional radiography may reveal non-specific soft tissue masses with various patterns of calcification. Invasion and erosion of underlying bone with periosteal reaction is unusual but may be seen. The various patterns like flecks, punctuate, granular, stippled, flocculent, ring- and arc like, coarse and dense calcification are best seen on CT images especially when the calcifications are subtle and when the tumors are deep seated and in complex anatomical areas [19, 20]. Matrix mineralization is more common in EMCS but calcifications may not be evident in rare instances [21]. On contrast enhancement the tumor appears lobulated with peripheral enhancement and sometimes with central low attenuation areas that probably represent necrosis [5, 22]. The uncalcified areas have low attenuation similar to muscle on CT [20]. On MR imaging EMCS appears as a lobulated mass with intermediate to high signal intensity on T2 weighted images and shows heterogeneous enhancement of both calcified and non calcified areas. Serpentine high flow vessels corresponding to hemangiopericytoma like areas seen histologically may also be seen. Low signal- intensity focal areas may be seen on contrast enhanced T1 weighted images [5].

Conventional radiographs are inadequate in case of soft tissue tumors like EMCS as was seen in this case on the OPG where the tumor was seen as a well demarcated soft tissue mass. Three-dimensional imaging is required for proper localization of the tumor, grading and diagnosis. EMCS shares its imaging features with many benign and malignant cartilaginous tumors but they are not usually found in extraskeletal sites. Although imaging features of EMCS are not characteristic, the two component structures of EMCS with differentiated cartilage islands interspersed within vascular undifferentiated mesenchyme are best appreciated with CT and MR imaging [5, 23]. PET findings in EMCS are not well documented. Hsing et al. [24] found no FDG uptake in the tumor mass on PET CT. In this case the primary tumor did not show any evidence of calcification but the recurrent tumor was more aggressive causing erosion of walls of maxillary antrum and also showed some subtle calcifications within the tumor mass. CT and MR findings were similar to those reported in literature although PET imaging showed intense FDG uptake in the tumor.

There are relatively few cytogenetic studies in EMCS and although some complex cytogenetic alterations have been found, not one of them has been found to be characteristic and consistent. Sainati et al. [25] found t (11;22) (q24;q12) typical of ES/PNET family of tumors in a case of osseous mesenchymal chondrosarcoma raising the possibility that EMCS may be related histogenetically to this

group of tumours. Nuamann et al. [26] have found an identical Robertsonian translocation involving chromosomes 8 and 20 and gain of all or a certain portion of chromosome 12 in both skeletal and extraskeletal mesenchymal chondrosarcoma which supports a genetic as well as histopathologic relationship between these anatomically distinct neoplasms. Gatter et al. have reported Trisomy 8 as the sole cytogenetic abnormality in a single case of EMCS [27]. Park et al. [28] found over expression of p53 protein in substantial fraction of mesenchymal chondrosarcomas although the genetic alteration of p53 gene was rarely seen. Cytogenetic studies were not done in this case due to lack of facilities.

EMCS is a very aggressive tumor with a rapid clinical course and metastasizes by hematogenous and lymphatic routes in high percentage of cases. The principal metastatic site is the lung and lymph node metastases are less common. Prognosis is not related to the patient's age or degree of cellular differentiation [4, 29]. Nakashima et al. [2] have reported a 5 year survival rate of 54.6 % and 10 year survival rate of 27.3 %. Prognosis in EMCS can be variable ranging from complete remission and long term survival to rapid local recurrence and widespread metastasis [29]. In this case primary excision with conservative margins was not successful and local recurrence was seen within a month. However no distant metastases were seen on PET-CT.

Early aggressive surgical resection with wide margins is the only effective treatment for EMCS. Adjuvant radiotherapy has been used although EMCS may be radio resistant. However preoperative radiotherapy may reduce the tumor bulk before radical resection and prevent further extension and micro metastasis and thus improve survival rates [30]. Adjuvant chemotherapy can be used in aggressive, high grade tumors with metastasis and rapid recurrence. The regimen is same as for ES/PNET consisting of vincristine, dactinomycin, cyclophosphamide and doxorubicin (VACD) either used alone or alternating with ifosfamide and etoposide (VACD-IE) [30]. Targeting active signaling pathways in chondrosarcoma like phosphoinositide-3 kinase and MEK- extracellular signal-regulated kinase (ERK) can improve control of tumor growth and recurrence as has been reported in intracranial chondrosarcoma [30]. Due to aggressive clinical, radiographic and histological features but lack of distant metastases, in this case the recurrent tumor was treated with wide excision followed by post-operative radiotherapy. There has been no recurrence in the 9 months following radiotherapy.

The clinical presentation, radiographic and FNAC findings of the primary tumor suggested a benign lesion. The rapid growth of the tumor suggested otherwise and the true nature of the tumor was confirmed on histopathologic evaluation only. This emphasizes the need for relying on histopathology to arrive at a correct diagnosis and also

determining the prognosis as was seen in this case. Conservative margins during resection of the primary tumor was ineffective in eliminating the tumor and this was followed by a more aggressive and rapid growth of the recurrent tumor within a short period of time.

Conclusions

This is a rare presentation of primary and recurrent EMCS with dedifferentiation in the buccal space of a young male with aggressive clinical, radiographic and histological features. The diagnostic challenges in this case were the inconclusive FNAC results, lack of radiographic evidence of characteristic calcifications and presence of areas of myxoid material and dedifferentiation on histopathology. Early diagnosis by histopathological evaluation, detection of distant metastases and early aggressive resection with wide margins followed by adjuvant radiotherapy/chemotherapy is important in diagnosis and management of EMCS. EMCS although rare can be considered in the differential diagnosis of soft tissue neoplasms with calcification in the orofacial region.

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