International Journal Dental and Medical Sciences Research
Volume 3, Issue 3, May-June 2021 pp 764-766 www.ijdmsrjournal.com ISSN: 2582-6018

A Rare Case of Extraskeletal Mesenchymal Chondrosarcoma of Buccal Space

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Submitted: 25-06-2021 Revised: 04-07-2021 Accepted: 07-07-2021

ABSTRACT : Conventional chondrosarcomas are low to intermediate grade tumors usually involving the limbs, pelvis, scapula and ribs. Mesenchymal chondrosarcoma is a rare, high grade pathological variant that was first described by Lichtenstein and Bernstein in 1959[1]. Extraskeletal mesenchymal chondrosarcoma (EMCS) constitutes about 1% of all chondrosarcomas[2]. We present a rare case of EMCS originating from right buccal space. The aim is to highlight the challenges in diagnosis due to rare clinical, radiographic and histologic features of this case as well as management challenges in view of malnutrition, breathing difficulties and bleeding from tumor.

KEY WORDS : Extraskeletal mesenchymal chondrosarcoma, buccal space, PET-CT, excision

I. INTRODUCTION

EMCS is a rare soft tissue sarcoma arising from soft tissues, mainly of lower extremities, followed by meninges and orbits. Only 6% of EMCS originate from other areas in head & neck region[3]. EMCS has characteristic clinical, radiological and histopathological picture. These tumors occur more frequently in young adults during the second or third decades of life, with a slight female preponderance[4]. The most characteristic histological feature is the dimorphic pattern composed of sheets of primitive undifferentiated mesenchymal cells interspersed with islands of mature cartilage[4]. Cartilagenous areas are sometimes poorly circumscribed blending in gradually with undifferentiated tumor cells[5]. CT scan shows soft tissue mass with areas of calcification, but these findings are not specific to EMCS[4]. MRI features are more characteristic and diagnostic.[3,4] These tumors need radical excision with three dimensional tumor free margins. Adjuvant chemotherapy is helpful in increasing overall survival.

II. CASE PRESENTATION

A 16 year old male presented with a big mass in right side of oral cavity for 7 months. The mass was of a size of a marble when he first noticed it. He was on liquid diet since one month due to inability to close the mouth. He had breathing difficulties in lying down position since 15 days. He couldn't open his right eye properly. The tumor was bleeding significantly even with slightest trauma. He underwent FNA at some other place before one month, which was inconclusive.

On local examination, a 15x15 cm sized swelling was seen protruding out from oral cavity with loosening of upper right sided teeth. Swelling was seen protruding in right nasal cavity also. There was no palpable cervical & supraclavicular lymphadenopathy.

A punch biopsy was suggestive of sarcoma. PET-CT revealed intense hypermetabolic FDG uptake in heterogeneously enhancing hypodense lesion in right buccal space. No metastasis was found in cervical & supraclavicular lymph nodes or other organs. MRI showed low intensity calcified & noncalcified areas in soft tissue lesion. Right sided maxilla was eroded. Lesion was reaching upto orbital plate without involving it. Lesion was highly vascular on MR angiogram.

Before definitive surgery, selective embolization of feeding artery, elective tracheostomy and feeding jejunostomy were done. Patient was transfused 2 units of PCV. Patient underwent excision of tumor with subtotal maxillectomy. Six cycles of adjuvant chemotherapy were given. Histopathology and immunohistochemistry were suggestive of extraskeletal mesenchymal chondrosarcoma. Patient came for follow up for 3 years & was disease free. After that, he lost to follow up.

III. DISCUSSION

EMCS is usually seen in young adults. It is rarely seen in children. The common site of primary tumor in head & neck region are intracranial, orbits and sinonasal tract[3]. Isolated cases of other areas like nasal cavity, eyelid, parapharyngeal space, ethmoid sinus & thyroid have been reported[6]. Diagnosis of EMCS on FNA is extremely challenging because of difficulty in

sampling both the phases of the tumor, lack of distinctive clinical and radiographic features & its rarity[3]. Immunohistochemistry is useful in diagnosis as EMCS is positive for S-100, CD99 and vimentin[5]. In present case, FNA was inconclusive. Histopathological & immunohistochemical evaluations were required for establishing the diagnosis.

Conventional radiograph like OPG is inadequate in EMCS. Three dimensional imaging is required for proper localization, grading and diagnosis of tumor[3]. 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[7], with MRI being more accurate[3,4]. PET findings in EMCS are not well documented. Hsing et al.[8] found no FDG uptake in tumor on PET-CT. Present case showed intense FDG uptake by tumor on PET-CT.

The principal metastatic site is lung. Lymph node metastases are less common. Prognosis is not related to patient's age or degree of cellular differentiation[9]. Nakashima et al.[10] have reported a 5 year survival rate of 54.6% and 10 year survival rate of 27.3%. Prognosis can be variable ranging from complete remission & long term survival to rapid local recurrence & widespread metastasis[9].

Surgery involving wide resection with 3dimensional tumor free margins followed by adjuvant chemotherapy is the mainstay of treatment. Addition of chemotherapy has been reported to improve overall survival[4]. Role of adjuvant radiotherapy is not well defined[4].

IV. CONCLUSION

This is a rare presentation of primary EMCS in the buccal space of a young male. FNA is not adequate in diagnosing EMCS. Preoperative course in this case was challenging because of poor nutrition, difficulty in breathing and plethora of tumor on MRI. Early diagnosis, detection of distant metastasis and early aggressive resection followed by adjuvant chemotherapy are important steps in management of EMCS. Although rare, EMCS should be considered in differential diagnosis of soft tissue neoplasms with calcification in orofacial region.

REFERENCES

[1]. Lichtenstein L, Bernstein D. Unusual benign and chondroid tumors of the bone. A survey of some mesenchymal cartilage tumors and a malignant chondroblastic tumor including a few multicentric ones, as well as many atypical benign chondroblastomas and chondromyxoid fibromas. Cancer. 1959;12:1142–1157.

[2]. Bertoni F, Picci P, Bacchini P, Capanna R, Innao V, Bacci G, Campanacci M. Mesenchymal chondrosarcoma of bone and soft tissues. Cancer. 1983;52(3):533–541.

[3]. Shalini Gupta, Ravinder Saran, Pankaj Sharma, Aadithya Urs. A rare case of Extraskeletal Mesenchymal Chondrosarcoma with dedifferentiation arising from the buccal space in a young male. Journal of Maxillofacial and Oral Surgery. 2015 Mar;14(Suppl 1):293-299.

[4]. Komal Arora. Extraskeletal Mesenchymal Chondrosarcoma. Arch Pathol Lab Med (2018) 142(11):1421-1424.

[5]. Weiss SW, Goldblum JR. Cartilaginous soft tissue tumors. In: Weiss SW, Goldblum JR, editors. Enzinger and Weiss's soft tissue tumors. 5. Amsterdam: Mosby Elsevier; 2007. pp. 1031–1036.

[6]. Liu J, Guo HX, Yuan L, He ZY. Extraskeletal mesenchymal chondrosarcoma of nasal cavity: report of a case. Zhonghua Bing Li Xue Za Zhi. 2009;38:204–205.

[7]. Hashimoto N, Ueda T, Joyama S, Araki N, Beppu Y, Tatezaki S, Matsumoto S, Nakanishi K, Tomita Y, Yoshikawa H. Extraskeletal mesenchymal chondrosarcoma: an imaging review of ten new patients. Skeletal Radiol. 2005;34:785–792.

[8]. Hsing CT, Oh SY, Lee S, Kwon HC, Kim SH, Park TH, Woo JS, Na SH, Kim HJ. Extraskeletal mesenchymal chondrosarcoma of the heart responded to systemic chemotherapy: a case report. Cancer Res Treat. 2007;39:131–133.

[9]. Huang KF, Tzaan WC, Lin CY. Primary intraspinal mesenchymal chondrosarcoma: a case report and literature review. Chang Gung Med J. 2003;26:370–376.

[10]. Nakashima Y, Unni KK, Shives TC, Swee RG, Dahlin DC. Mesenchymal chondrosarcoma of bone and soft tissue: a review of 111 cases. Cancer. 1986;57:2444–2453.

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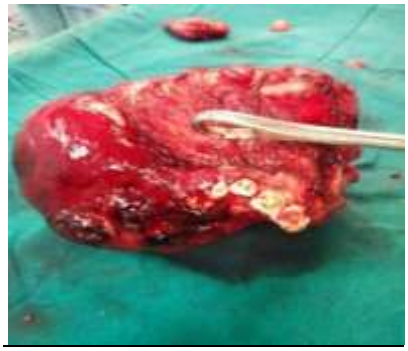
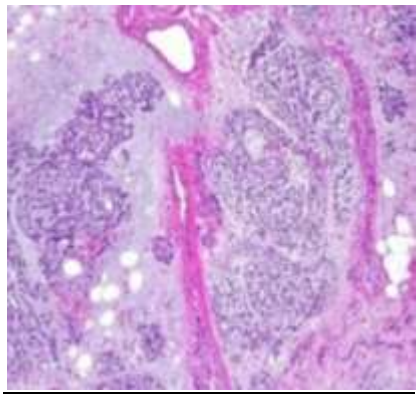
Volume 3, Issue 3, May-June 2021 pp 764-766 www.ijdmsrjournal.com ISSN: 2582-6018

PREOPERATIVE PICTURE



TUMOR SPECIMEN

POSTOPERATIVE PICTURE



HISTOPATHOLOGICAL FINDINGS