


Maxillary mesenchymal chondrosarcoma presenting with epistaxis in a child

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ABSTRACT

Mesenchymal chondrosarcomas are a rare variant of primary chondrosarcomas and can pose a diagnostic dilemma, especially when the features on conventional imaging are equivocal for an aggressive lesion. There is very little PET-CT experience in mesenchymal chondrosarcomas as per the literature and to the best of our knowledge, we are the first to describe a maxillary mesenchymal chondrosarcoma on PET-CT imaging. We report a case where PET-CT not only complemented conventional imaging in suspecting a malignant osseous lesion, but also was indicative of the grade of the tumor.

CASE REPORT

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A 13 year-old boy presented with a 1 month history of intermittent episodes of small volume epistaxis. There was no significant past medical, surgical or family history. Physical examination revealed a left palatal fleshy mass centered at left posterior molars. A nasoendoscopic examination demonstrated a large swelling on the floor of left anterior nasal space, involving the left inferior meatus and abutting the inferior turbinate.

Computed Tomography (CT) of the paranasal sinuses showed a lobulated, expansile lytic lesion of left maxilla [Fig 1] with a lucent center and peripheral calcifications. The lesion was associated with a large soft tissue component that demonstrated mild heterogeneous contrast enhancement. The lesion was centered at the left maxillary alveolus, involving the left hard palate, inferior part of the anterior left maxillary wall and medial left maxillary wall. The lesion also involved the roots of the left maxillary molars and pre-molars and was abutting the left orbital floor, with no gross orbital invasion.

The patient subsequently underwent 18-FDG PET-CT study. The mass centered in left maxilla was hypermetabolic, with index SUVmax 9.7 [Fig 2]. There was no FDG-avid metastatic disease.

A trans-oral incisional biopsy of the left palatal mass was performed and was suggestive of a preliminary diagnosis of mesenchymal chondrosarcoma, as evidenced by a biphasic pattern featuring sheets of small round cells with hemangiopericytoma-like vascular pattern and mature cartilaginous islands.

The patient underwent left subtotal maxillectomy via lateral rhinotomy and translabial approach. The final histology was conclusive of a 5cm high-grade (Grade 3) mesenchymal chondrosarcoma. One of the resection margins was positive for tumor involvement and the patient was planned for adjuvant chemotherapy.

DISCUSSION

Etiology & Demographics

Chondrosarcomas are the third most common primary malignancy of bone after myeloma and osteosarcoma. Chondrosarcomas can be categorized on histology basis as conventional intra-medullary, clear cell, myxoid, juxta-cortical, mesenchymal and dedifferentiated types [1]. On the basis of osseous location, they can be divided into central, peripheral and juxta-cortical types. The mesenchymal variant accounts for approximately 2%-8 % of all chondrosarcomas of bone [2, 3] and is an aggressive high-grade tumor with a strong tendency to metastasize. Compared with other chondrosarcoma subtypes, there is a higher proportion of extra-skeletal occurrence of tumor [4]. Also in contrast to conventional chondrosarcomas, mesenchymal tumors most commonly involve the axial skeleton, including the craniofacial bones (especially the jaw), ribs, ilium, and vertebra, and there may be involvement of multiple bones [4]. Men and women are equally affected, most frequently in the 2nd to 4th decade of life (average age, 25 years). No precursor chondroid lesion (such as an osteochondroma or enchondroma) has been identified.

Clinical & Imaging findings

The common presenting symptoms are pain and swelling. It is not uncommon for symptoms to have been present for many months. In our case, the patient presented with epistaxis.

Mesenchymal chondrosarcomas are predominantly osteolytic lesions, often associated with large soft tissue masses. A moth-eaten to permeative pattern of bone destruction may be seen. On MR imaging, the lesions typically show a low-intermediate signal on T1w and intermediate signal on T2w images. Occasionally, low-signal intensity serpentine, high-flow vessels may be seen.

Contrast-enhanced CT findings in our patient were equivocal for an aggressive lesion. The lesion was a lobulated, expansile lytic lesion with the rim formed by markedly thinned out cortex and peripheral calcifications. The soft tissue component demonstrated mild heterogeneous enhancement and no central calcifications (specifically, there were no features of a typical ring-and-arc chondroid matrix). At this point, the differential diagnoses included a fibro-osseous lesion such as an ossifying fibroma, fibrous dysplasia and a sarcomatous lesion.

Mesenchymal chondrosarcomas are known to lack the typical cartilaginous septal and peripheral enhancement seen with a conventional chondrosarcoma on contrast CT/MRI studies [5]. The contrast enhancement is variable from homogeneous to heterogeneous, often diffuse. Subtle chondroid (ring-arc) matrix calcifications are seen in about two-thirds of cases and the rest one-third of cases do not have the typical ring-arc calcifications (as in our case) and hence, pose a diagnostic dilemma.

Bone scintigraphy, though highly sensitive for detecting intra-osseous lesions, cannot reliably differentiate between

benign and malignant causes of increased uptake of radiopharmaceutical [6].

There is limited PET-CT experience in mesenchymal chondrosarcoma.

PET imaging provides information on the biological activity of a lesion. FDG PET has been proposed as a diagnostic tool for distinguishing benign from malignant osseous lesions [7]. Several studies have found malignancies to be significantly more FDG-avid than benign lesions. A study by Feldman et al. [8] suggested 18FDG PET-CT had a high degree of sensitivity (~91%), specificity (100%), and accuracy (~95%) when using Standard Uptake Values (SUV) to predict chondrosarcoma (SUV > 2.0) versus enchondroma (SUV < 2.0). This concept particularly holds true when comparing tumors of the same histologic type. For example, for cartilage tumors, benign cartilage lesions typically have lower FDG-uptake than chondrosarcomas and lower-grade cartilage tumors have lower uptake compared to higher-grade chondrosarcomas. [8, 9]. A study by Brenner et al. [10] that involved 31 patients with chondrosarcoma correlated pre-therapeutic tumor SUV to tumor grade as well as patient outcome in terms of local relapse or metastatic disease. The tumor SUV was 3.38 +/- 1.61 for grade I (n = 15), 5.44 +/- 3.06 for grade II (n = 13), and 7.10 +/- 2.61 for grade III (n = 3) lesions in this study. Mesenchymal chondrosarcomas tend to be high grade tumors and therefore, are typically hypermetabolic masses on PET-CT.

Also, PET-CT has higher sensitivity and specificity in staging malignant disease. A study by London et al. [11] that involved 314 lesions on 86 scans in children with metastatic osteosarcoma and Ewing sarcoma, comparing PET/CT with conventional imaging (CT, MRI, ultrasound and bone scan) showed that PET/CT had a higher sensitivity (98% vs 83%) and specificity (97% vs 78%) than conventional imaging for detecting distant metastases, with the exception of pulmonary nodules.

The final histology diagnosis in our patient was that of a grade 3 mesenchymal chondrosarcoma. In retrospect, the tumor SUV was 9.7, consistent with the histology diagnosis of a high grade malignant tumor. Also, there was no hypermetabolic nodal or visceral metastatic disease on the PET-CT, obviating the need for other modalities to stage the disease.

Treatment & Prognosis

Mesenchymal chondrosarcomas are aggressive tumors requiring wide local surgical excision.

Limited literature on chemotherapy for mesenchymal chondrosarcoma suggests potential survival benefit. A retrospective series of 26 patients with mesenchymal chondrosarcoma treated at the Rizzoli Institute found that chemotherapy and complete surgical remission were associated with improved survival rates [12].

The tumor has a tendency towards both local and distant recurrences, which could occur as late as 20 years following

the initial diagnosis [13]. The overall prognosis is poorer than with conventional chondrosarcomas and the reported 10-year survival rates are 10 to 20 percent [12, 13].

Differential Diagnosis

The differential diagnoses based on osteolytic lesions with foci of internal calcifications include conventional chondrosarcoma, osteosarcoma, fibro-osseous lesions (such as fibrous dysplasia, ossifying fibroma) and calcified/treated metastases. Apart from the clinical setting, biopsy may be essential to differentiate and confirm the diagnosis.

Conventional chondrosarcoma: The "rings and arcs" chondroid matrix is often seen and the septal and peripheral enhancement is helpful in making the diagnosis.

Osteosarcoma: The tumor demonstrates aggressive sunburst/lamellated periosteal reaction with characteristic "cloud-like" or "fluffy" matrix calcifications.

Fibro-osseous lesions: Fibrous dysplasia (FD) and ossifying fibroma (OF) are well-circumscribed lesions lacking a periosteal reaction. FD can vary in appearance from lucent to a densely sclerotic lesion and typically demonstrates ground-glass matrix. OF has a soft tissue density (fibrous) central area surrounded by a sclerotic rim.

TEACHING POINT

18FDG-PET can help in identifying malignant cartilage lesions, when the findings are equivocal on conventional cross-sectional imaging. The increased radiotracer uptake by chondrosarcomas has been correlated well with SUV readings and tumor grade. Hypermetabolic lesions, in the appropriate clinical context, would prompt the physician to obtain a histopathological diagnosis. In addition to aiding in the primary diagnosis of malignant osseous lesions, PET-CT is very useful in staging the malignant disease and follow-up for tumor recurrence.

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FIGURES



Figure 1: A 13-year-old boy with mesenchymal chondrosarcoma of the maxilla. Findings: Axial CT images in bone window (A and B) and soft tissue window (C) demonstrate a lobulated, expansile lytic lesion of left maxilla (arrows) with lucent center and peripheral calcifications. The lesion is centered at the left maxillary alveolus and is associated with a large soft tissue component which shows mild heterogeneous contrast enhancement. Coronal CT in bone window (D) shows the lesion abutting the left inferior orbital wall (arrowhead), with no gross invasion into the orbit. Technique: Siemens Sensation 64 scanner, 2mm Slice thickness, 120 kVp and 100 mAs; Intravenous contrast administered: Omnipaque 300, Volume 50 ml.

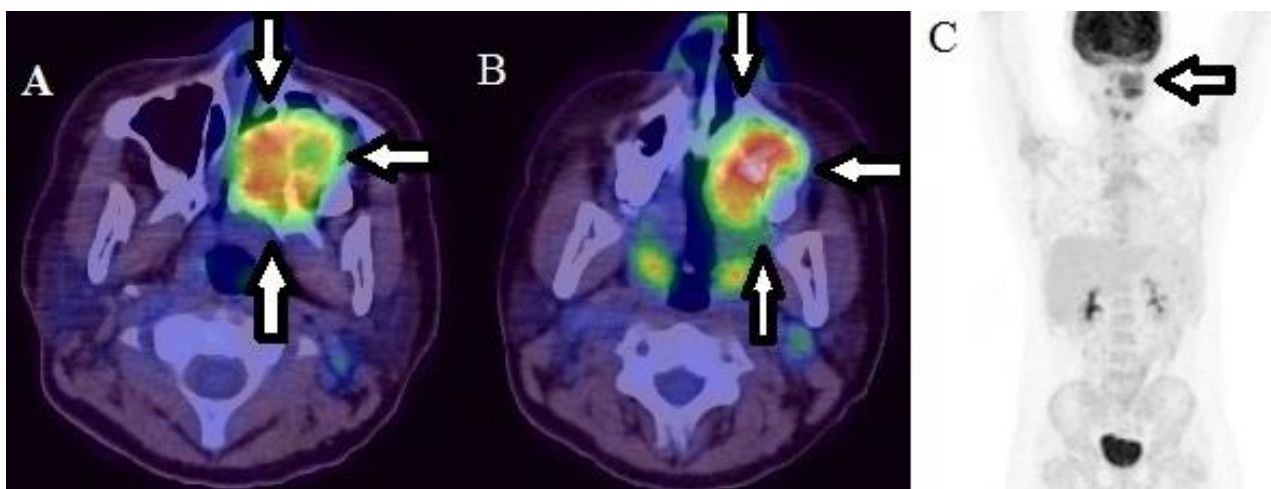


Figure 2: A 13-year-old boy with mesenchymal chondrosarcoma of the maxilla. Findings: Axial PET-CT (A and B) and Coronal PET (C) images show a moderately FDG-avid mass (index SUVmax 9.7) arising from left maxilla (arrows). No other abnormal focus of increased FDG-uptake is seen in the coronal PET image. Technique: Performed using Siemens Biograph 64 scanner, 96 minutes after injection of 5.2mCi of 18-FDG tracer. Patient's blood glucose was 95mg/dl at the time of FDG injection. CT scan settings were 5 mm slice thickness, 120 kVp and 191 mAs.

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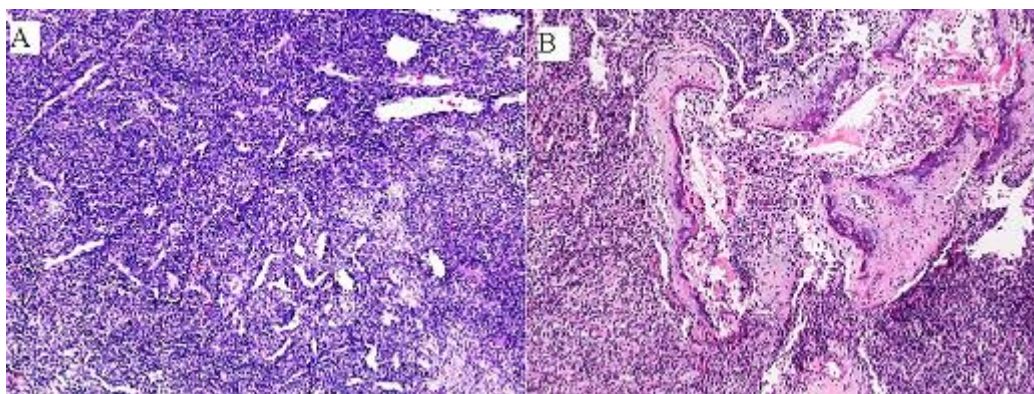


Figure 3: A 13-year-old boy with mesenchymal chondrosarcoma of the maxilla.
(A) Hematoxylin and eosin-stained section (100x) shows non-chondroid component of mesenchymal chondrosarcoma featuring small round cell morphology with hemangiopericytoma-like vascular pattern.
(B) Hematoxylin and eosin-stained section (100x) shows biphasic pattern of mesenchymal chondrosarcoma featuring hyaline cartilage islands admixed with small round cells.

Etiology	Unknown
Incidence	0.24% among bone neoplasms and 2-8% of all chondrosarcomas
Gender predilection	Males and females are equally affected
Age predilection	10 – 40 years
Risk factors	Recurrence from inadequate surgical resection
Special features	In contrast to conventional chondrosarcoma: - affects younger patients - no precursor chondroid lesion such as an osteochondroma or enchondroma - commonly involves the axial skeleton (non-long bone sites) and also has a high proportion of extra-skeletal incidence.
Treatment	Surgical resection with wide margins; Adjuvant chemo-radiation therapy when the margins are positive or complete resection is not possible.
Prognosis	Poor prognosis compared to most other chondrosarcomas. 10-year survival cited as 10-20%. Tend to have late local recurrence and metastasis.
Findings on Imaging	<ul style="list-style-type: none"> • Predominantly osteolytic with aggressive bone destruction on radiographs and CT. They are often large with extensive extra-osseous soft tissue components. Subtle chondroid (ring-arc) matrix calcifications seen in two-thirds of cases. • Variable pattern of contrast-enhancement on contrast CT/MR, with lack of typical cartilaginous septal and peripheral enhancement seen with conventional chondrosarcomas. • Hypermetabolic masses on 18 FDG PET-CT as the tumors tend to be of high-grade.

Table 1: Summary table of key aspects and imaging findings of mesenchymal chondrosarcoma.

	Mesenchymal chondrosarcoma	Conventional chondrosarcoma	Osteosarcoma	Fibro-osseous lesions – FD, OF
Radiography	<ul style="list-style-type: none"> • Predominantly osteolytic lesion with a moth-eaten to permeative pattern, usually with large extra-osseous components. • May show ring-and-arc chondroid calcification, often not extensive. 	<ul style="list-style-type: none"> • Expansile, mixed lytic-sclerotic lesion with variable endosteal scalloping. • Characteristic ring-and-arc calcifications are often seen. 	<ul style="list-style-type: none"> • Destructive bone lesion with wide transition zone and aggressive sunburst/lamellated periosteal reaction with “cloud-like” or “fluffy” matrix calcifications. 	<ul style="list-style-type: none"> • FD and OF are well-circumscribed lesions with variable bone expansion. FD can be lucent or sclerotic, typically with ground-glass matrix. OF demonstrates intra-cortical osteolysis with a sclerotic band (osteoblastic rimming). The lesions lack a periosteal reaction
CT	<ul style="list-style-type: none"> • Typically an aggressive osteolytic lesion with a large associated soft tissue mass. The “ring-and-arc” chondroid mineralization is often subtle and not extensive. 	<ul style="list-style-type: none"> • The mineralized portion is seen as “rings and arcs” chondroid matrix. 	<ul style="list-style-type: none"> • Features are largely similar to plain radiographs, except that small areas of mineralization are better apparent. 	<ul style="list-style-type: none"> • Features are largely similar to plain radiographs. • CT is superior in evaluating the matrix and cortical integrity.
MRI	<ul style="list-style-type: none"> • Low-intermediate signal on T1w and intermediate signal on T2w images. Occasionally, low-signal intensity serpentine, high-flow vessels. 	<ul style="list-style-type: none"> • Low-intermediate T1w and heterogeneously T2w hyperintense signal. 	<ul style="list-style-type: none"> • Mineralized/ossified components are of low-signal on T1w and T2w sequences. The non-mineralized components are of intermediate T1w and high T2w signal intensity. 	<ul style="list-style-type: none"> • FD has a heterogeneous appearance on T1w and T2w sequences. • OF has a low T1w signal and iso – high T2w signal.
Contrast enhancement on CT/MRI	<ul style="list-style-type: none"> • Variable from homogeneous to heterogeneous, often diffuse. 	<ul style="list-style-type: none"> • Septal and peripheral pattern of enhancement. 	<ul style="list-style-type: none"> • Solid components demonstrate contrast enhancement. 	<ul style="list-style-type: none"> • The lesions show variable contrast enhancement of the fibrous components.
PET-CT	<ul style="list-style-type: none"> • Typically are hypermetabolic masses 	<ul style="list-style-type: none"> • High-grade (grade 2 or 3) chondrosarcomas are typically hypermetabolic, while the low-grade lesions cannot be differentiated from benign cartilaginous lesions. 	<ul style="list-style-type: none"> • Typically are hypermetabolic masses 	<ul style="list-style-type: none"> • FD and OF can be variably hot on FDG PET and correlation with cross-sectional imaging should help differentiate.

Table 2: Differential diagnosis table for mesenchymal chondrosarcoma.
FD = Fibrous dysplasia, OF = Ossifying fibroma

ABBREVIATIONS

CT = Computed tomography
 FD = Fibrous dysplasia
 FDG = Fluoro-deoxy-glucose
 MRI = Magnetic Resonance Imaging
 OF = Ossifying fibroma
 PET = Positron emission tomography
 SUV = Standard uptake value

KEYWORDS

Maxilla; mesenchymal; chondrosarcoma; PET-CT; hypermetabolic; epistaxis

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