

A Case of Chondromyxoid Fibroma-Like Osteosarcoma in a 13-Month-Old Girl

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Abstract

Chondromyxoid fibroma-like osteosarcoma is a rare type of osteosarcoma with only a handful of cases reported in the literature. We report a case of chondromyxoid fibroma-like osteosarcoma involving the sinonasal region in a 13-month-old girl, with discussion of its radiological and pathological features and differentials.

Introduction

Malignant bone tumors account for around 6% of childhood cancers, of which Osteosarcoma is the most common type [1]. Chondromyxoid fibroma-like osteosarcoma is a rare type of osteosarcoma that can easily be misdiagnosed as a benign lesion [2]. If treated inadequately, it may recur with an increased histological grade [2]. This report documents a rare case of sinonasal chondromyxoid fibroma-like osteosarcoma in a 13-month-old girl.

Case report

A 13-month-old girl was referred to the pediatrics and ophthalmology outpatient clinic for divergent squint. She was born full-term via a normal spontaneous vaginal delivery and enjoyed an unremarkable perinatal history. She was first noticed to have a right, divergent squint and subtle facial asymmetry at 6 months of age. Otherwise, there was no delay in her developmental milestones. Upon examination, her right face appeared more prominent than the left. Limited adduction of her right eye and optic disc pallor suggesting right optic atrophy were noted.

An MRI examination was performed, and revealed a multilobulated soft tissue mass involving the right maxillary sinus and nasal cavity. It was isointense on T1-weighted images and heterogeneously hyperintense on T2-weighted images, showing heterogeneous contrast enhancement (Figure 1). It displaced the nasal septum to the left. It abutted the right orbit, displaced the right medial rectus, and caused extrinsic compression of the intra-canalicular portion of the right optic nerve (Figure 2). The mass eroded the bony walls of the maxillary sinus, reaching the pterygopalatine fossa, pterygomaxillary fissure, the retro maxillary fat space, and pre-maxillary subcutaneous tissue. The alveolar process of the maxilla and part of the hard palate was involved. Superiorly, the right cribriform plate was also eroded. A CT scan (Figure 3) was performed and revealed a soft tissue mass with mild contrast enhancement and internal calcifications. Extensive adjacent bone erosions were noted. No suspicious lesions in the thorax, abdomen and pelvis were detected. Bone scans showed no abnormal tracer uptake.

Endoscopic biopsy of the tumor mass was performed. Histology of the specimen was reported as chondromyxoid fibroma of sinonasal region. The mass was eventually excised endoscopically. Intraoperatively, the tumor was found to involve the whole right maxillary antrum, anterior and posterior ethmoid sinuses and sphenoid sinus. The lamina papyracea, medial orbital floor and part of the bony cribriform were eroded. The dura was intact.

Histology of the tumor showed a distinctly lobulated pattern, consisting of lobules of polygonal tumor cells with a prominent Chondromyxoid appearance (Figure 4a). In these areas, the cells were loosely scattered in a highly Chondromyxoid stroma and formed lobules separated by fibrovascular septa of varying thickness. The histomorphology was akin to chondromyxoid fibroma. In other areas, however, the tumor cells assumed a plump spindle form, possessing moderately pleomorphic hyperchromatic nuclei and small to moderate amount of eosinophilic cytoplasm (Figure 4b). They were fairly closely packed and demonstrated prominent osteoid deposition in their vicinity. Mitosis was infrequent, lamellar trabecular bony fragments were not seen. Immunohistochemical staining revealed strong positive staining of the spindle tumor cells for CKD4 and MDM2 but only focal staining for S100. Mib1 showed about 10% nuclear staining. The overall features are those of chondromyxoid fibroma-like osteosarcoma.

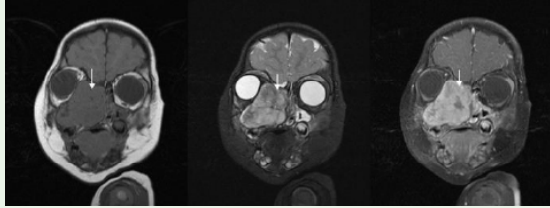


Figure 1: Coronal MR Images showing a soft-tissue mass (arrow) involving the right maxillary sinus and nasal cavity demonstrating isointense signals on T1W images, heterogeneously hyper intense signals on T2W images, and heterogeneous contrast enhancement.

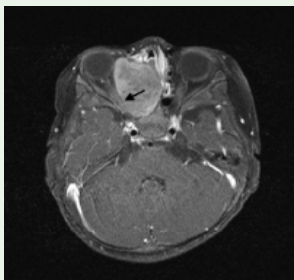


Figure 2: Gadolinium-enhanced axial MR image showing compression of the right optic nerve by the tumor (arrow).



Figure 3: Coronal CT image showing calcification within the tumor (arrow) and bony erosions.

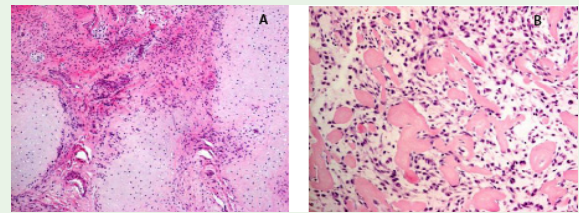


Figure 4: Figure 4: (A) The tumor shows a prominent chondromyxoid lobulated pattern, consisting of lobules of loosely scattered polygonal cells supported in a highly chondromyxoid stroma (B) In areas, the tumor cells assume a plump spindle form with prominent deposition of osteoid in their vicinity (Hematoxylin and eosin stain, magnification A 100x, B 200x).

Follow-up MRI 7 weeks after the surgery showed residual tumor, which was later confirmed by endoscopic biopsy. Further operation was not performed in view of the extensive involvement of the tumor and the high risk of further radical surgery. Serial MRI performed in the following two years showed no significant interval change in the residual tumor at the surgical bed.

Discussion

Chondromyxoid fibroma-like osteosarcoma is a rare type of osteosarcoma. It is classified by the World Health Organization as a subtype of conventional osteosarcoma [3]. It is also considered as a subtype of low-grade osteosarcomas [2]. Histologically, it is characterized by loose aggregates of cells supported in a highly myxoid stroma and segregated into lobules by fibrovascular septa, with osteoid production by the tumor cells suggesting osteosarcoma [2]. Two cases were described by Chow et al. [2]. The first case was a 39-year-old woman with tumor of sinonasal origin. The second case was a 28-year-old man with a tumor at the left iliac crest. Both cases were initially misdiagnosed and recurred after surgical resection with increase in histological grade. Radiological features of chondromyxoid fibroma-like osteosarcoma were described by Chow et al. as expansile osteolytic mass with bone erosions and soft tissue infiltration [2].

Our case is unique in that it demonstrated that chondromyxoid fibroma-like osteosarcoma can occur in children as young as 13 months old. And to the best of our knowledge, the MRI features of chondromyxoid fibroma-like osteosarcoma have not been described in the literature before. In our case, CT and MRI demonstrated an expansile soft tissue mass involving the right maxillary sinus and nasal cavity with erosions of adjacent bones and internal calcifications representing matrix mineralization within the tumor. MRI signal characteristics were non-specific, showing isointense signals on T1-weighted images, heterogeneously hyper intense signals on T2-weighted images, and heterogeneous contrast enhancement.

The extensive soft tissue mass and adjacent bone erosions noted in our case was suspicious of a malignant tumor. However, the overall radiological findings did not permit the differentiation of chondromyxoid fibroma-like osteosarcoma from other types of osteosarcoma or other sinonasal malignancies.

A recent review by Shapiro et al. [4] showed that rhabdomyosarcoma is by far the most common pediatric sinonasal malignancy, followed by lymphoma, sarcoma, and olfactory neuroblastoma. Radiologically, all of them can present with soft tissue mass with erosion of the adjacent bones. However, calcifications are rare in rhabdomyosarcoma and lymphoma, whereas osteoid matrix and cartilaginous matrix calcifications may be seen in osteosarcoma and chondrosarcoma respectively [5]. Olfactory neuroblastoma are usually centered at the superior nasal cavity. Calcifications within olfactory neuroblastoma have been frequently observed, but could be due to remaining fragments of eroded bone [5]. Malignancies arising from the skull base with anterior extension may also demonstrate similar radiological findings, but the site of origin should be helpful in the differential diagnosis. Although it may be difficult to predict the histology of the tumor based on imaging findings, a detailed description of the tumor extent is crucial in guiding the further investigation and management and in monitoring of disease. MRI and CT are complementary to each other in the assessment of sinonasal tumors as MRI, is superior in delineating tumor extent while CT is superior in detecting calcifications and delineating the extent of bony involvement.

The main pathological differential diagnosis of chondromyxoid fibroma-like osteosarcoma includes chondromyxoid fibroma, ossifying fibromyxoid tumor, and chondrosarcoma. The single most important distinguishing feature is the presence of fairly closely packed plump spindle tumor cells associated with direct osteoid deposition. Such features, not seen in chondromyxoid fibroma

and chondrosarcoma, coupled with a chondromyxoid fibromatous pattern in other areas are diagnostic of chondromyxoid fibroma-like osteosarcoma as described by Chow et al. [2]. The absence of peripheral lamellar trabecular bone formation in the present case also distinguishes it from an ossifying fibromyxoid tumor. While rare ossifying fibromyxoid tumors have atypical or overtly malignant features including high nuclear grade, high cellularity, increased mitotic activity and the presence of hyalinised collagen simulating osteoid, they are not seen in typical cases and such atypical ones have been regarded as low grade malignant tumors [6]. In addition, it is of interest to note the recent finding that MDM2 and CDK4 immunohistochemistry has been shown to be of value in the differentiation of low-grade osteosarcomas from other primary fibro-osseous lesions of the bone [7]. Indeed, our case demonstrated strong immunostaining for both MDM2 and CDK4, a finding that greatly supports if not confirm our diagnosis of chondromyxoid fibroma-like osteosarcoma.

In conclusion, chondromyxoid fibroma-like osteosarcoma is a rare type of osteosarcoma with only a handful of cases reported in the literature. It can occur in both pediatric and adult age groups. Radio logically; it appears as an aggressive soft tissue mass with bony erosions and matrix mineralization. Careful histological evaluation is mandatory to avoid misdiagnosis.

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