Chondrosarcoma
A rare case report with unusual pathology

Zahra khaffafpour. MD

Pediatric hematologist oncologist

Shahid Beheshti Medical University

Mofid Children’s hospital

Palliative care center

- A 4-year-old girl, was referred to pediatric oncology clinic with pain and two palpable lumps in labia majora on the right side, since the age of two, for which she underwent FNA and core needle biopsy.

- The pathologic results: Enchondroma without any malignant features.

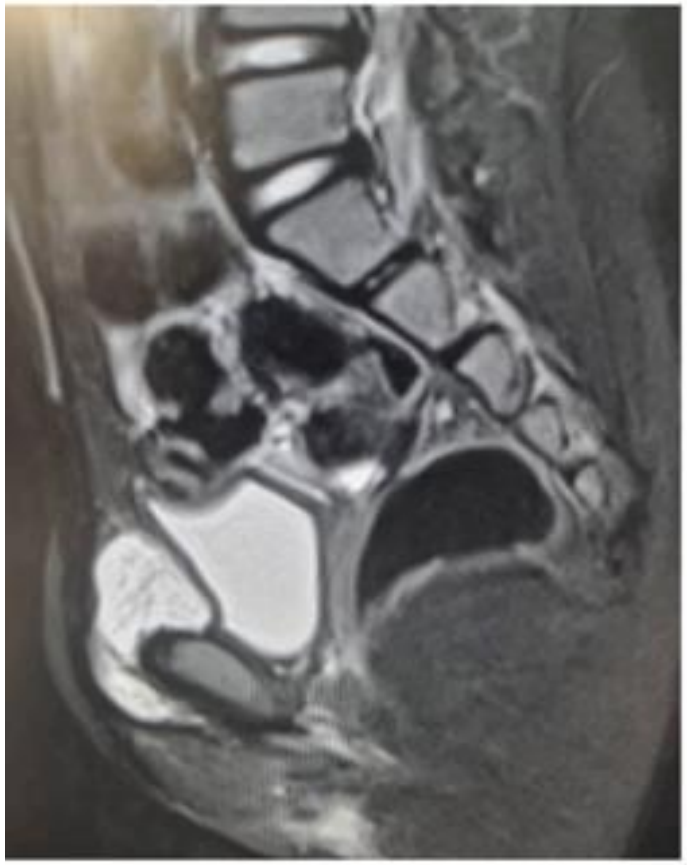
- After 2 years, she returned with recurrence of pain and firm masses in the genitalia.

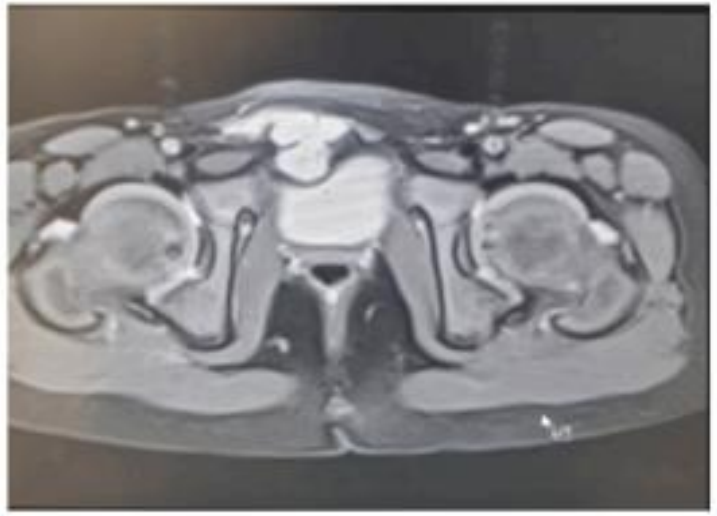
- physical examination: An ill-defined mass with firm consistency was identified within the skin fold of right labia majora.

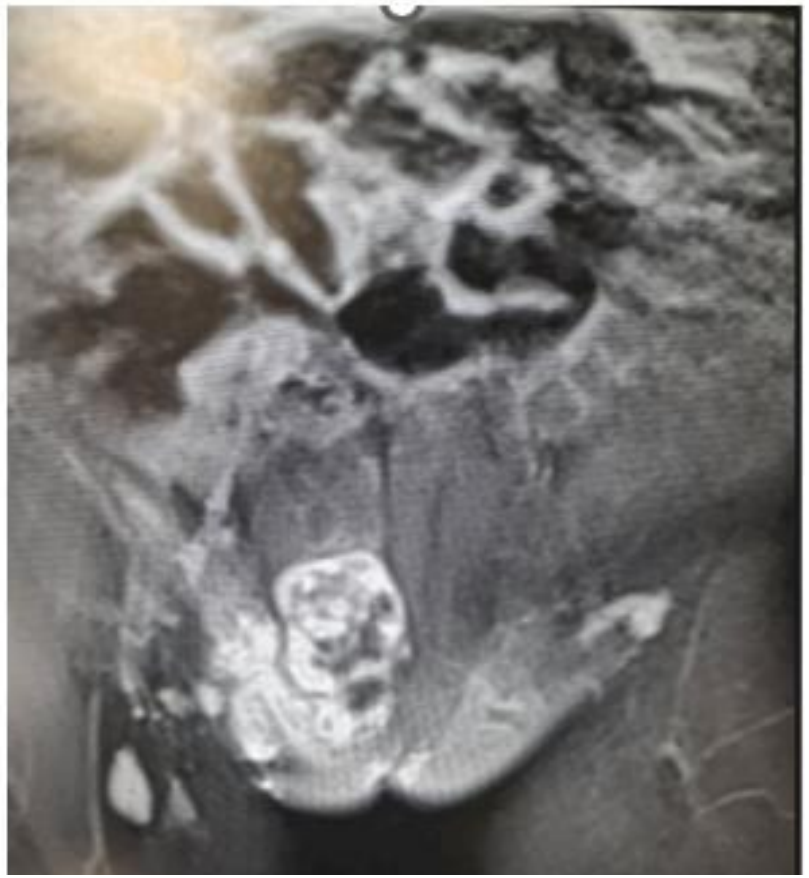
- A whole-body scan was performed for the patient which was negative for increased bone activity.

Magnetic resonance imaging

A lobulated lesion measuring 35 × 20 × 19 mm, on right side of prepubic space with extension into the suprapubic area and retzius space without evidence of bone involvement (Fig. 1 A, B). T1 image showed patchy and peripheral enhancement of the prepubic mass after contrast administration (Fig. 2).







Chondrosarcoma

- **Wide surgical excision** of the tumor was carried out under general anesthesia.

- **A soft tissue mass was found at the right side of the prepubic space with extension into the suprapubic area and deeply into the retzius space.** Biopsies were taken from ramus and symphysis of pubis.

- The **pathologic examination** revealed a **cartilaginous neoplasm composed of chondroid nodules** of varying sizes permeating through fibrous stroma.

The lesion exhibited **atypical chondrocytes** with hyperchromatic vesicular nuclei, with some binucleated or multinucleated forms, accompanied by **myxoid areas**. There were also **foci of atypical chondrocytes in the periphery with spindle and epithelioid pattern**. The **neoplastic chondrocyte nodules** were encapsulated by a thin fibrous capsule.

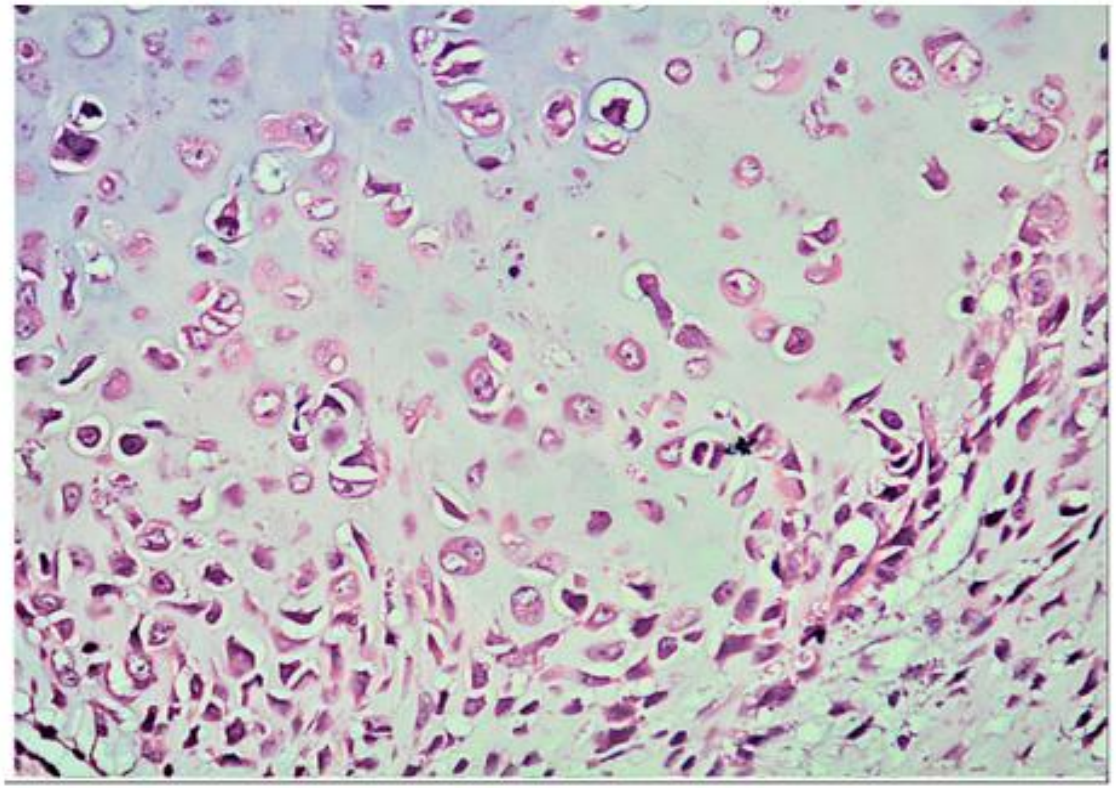


Fig. 5. Atypical chondrocytes with hyperchromatic and vesicular nuclei (H&E, X400).

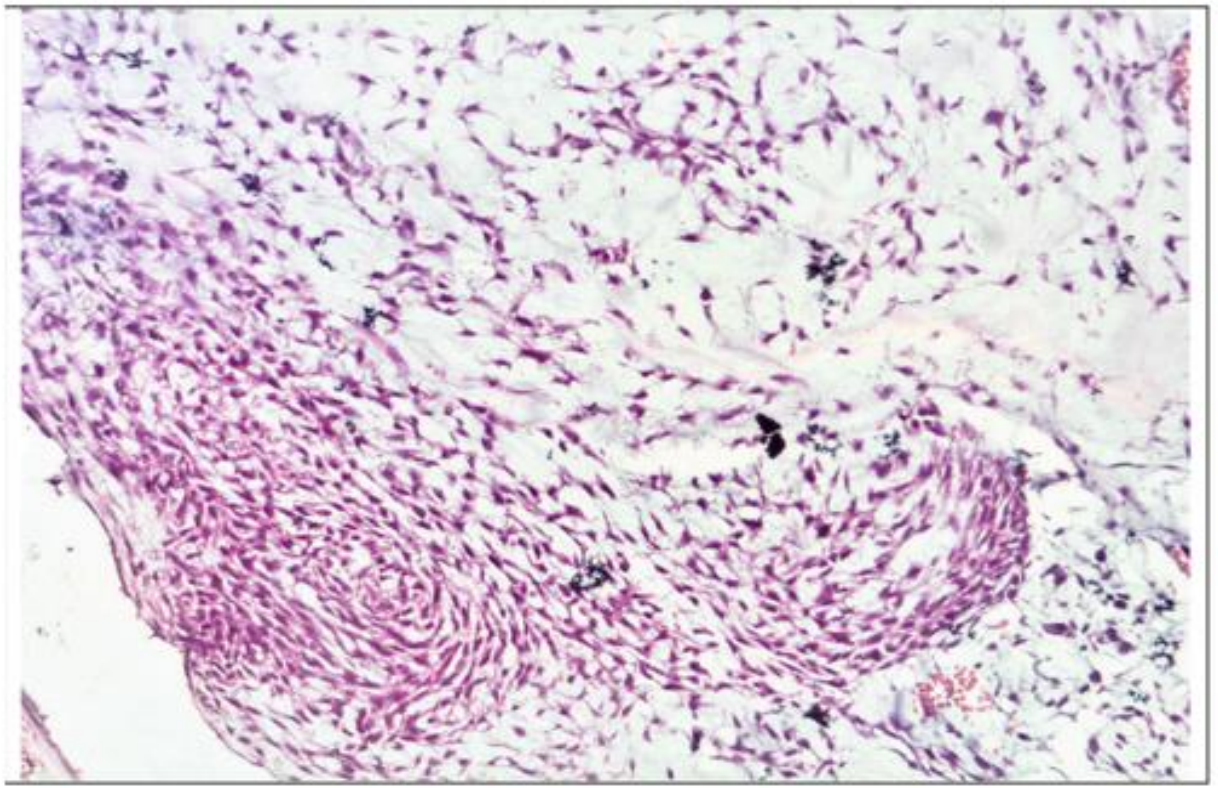


Fig. 4. Chondroid stroma with myxoid areas and atypical chondrocytes with spindle pattern (H&E, X200).

Subtype	Key Features	Behavior
Conventional	Hyaline cartilage, graded 1–3	Variable, grade-dependent
Clear Cell	Epiphyseal, clear cytoplasm	Low-grade, indolent
Dedifferentiated	Mix of low-grade + high-grade sarcoma	Very aggressive
Mesenchymal	Small round cells + cartilage	Aggressive, metastasis-prone
Juxtacortical (Periosteal)	Surface lesion	Usually better prognosis

Conventional Chondrosarcoma

All **surgical margins** and biopsy from **bony structures** were reported to be negative.

IHC: positive staining for S-100, desmin, and vimentin, negative staining was observed for CD99, FLI-1, myogenin, NKX2.2

The histopathological findings were indicative of a **conventional chondrosarcoma (grade II) of soft tissue**, since the lesion was located in the **soft tissue of the labia majora**.

Chondrosarcoma (CS) is considered the **second most common** sarcoma of bone following osteosarcoma, accounting for **20–30 %** of all skeletal sarcomas.

CS have an estimated incidence of 1 in 200,000 per year in the United States.

CS is a heterogeneous malignant tumor that produce cartilage matrix showing hyaline cartilage differentiation.

CS are categorized as **primary** when arise de novo; and if they arise from preexisting benign cartilaginous neoplasms such as enchondroma or osteochondroma are referred to as **secondary** chondrosarcomas.

chondrosarcoma

The risk of chondrosarcoma arising in a **solitary osteochondroma** has been reported to be **<1 %,** risk increases to 5 % in **multiple osteochondromatosis.**

CS occurs mostly in adult with the peak of incidence occurring in the fifth to seventh decades of life. **Chondrosarcomas in children and adolescents are uncommon and constitute <5 % of bone sarcomas**.

The **pelvis**, followed by the **proximal femur** is the most common site of skeletal involvement in CS irrespective of the age group.

CS most commonly presents in the bony skeleton, although a small percentage present as a primary soft tissue mass.

chondrosarcoma

- The 2020 WHO classification categorizes malignant chondrosarcoma as grade 1 to 3 based on the **nuclear size, mitotic activity, and degree of cellularity.**

- The majority of chondrosarcomas are **conventional** and are classified as **grade 1 or 2.**

Conventional chondrosarcomas are locally aggressive tumors that constitute the most common form of CS observed **only as skeletal tumors** (85 % of cases).

Extraskeletal chondrosarcomas account for about 1 % of all chondrosarcomas with the majority being of the myxoid (most common) or mesenchymal subtypes.

Our case was a 4-year-girl with an **extraskeletal chondrosarcoma** of genitalia (vulva) which did not fulfill either criteria for mesenchymal or myxoid chondrosarcoma but showed the pathologic features of **the conventional type.**

This pathologic subtype has not been reported in the extraskeletal CS so far.

Extraskeletal chondrosarcomas

Extraskeletal chondrosarcomas comprise 1 % of all chondrosarcomas and 1–2% of soft-tissue sarcoma.

Extraskeletal chondrosarcomas tend to be of higher grade than conventional skeletal chondrosarcomas, with the majority being of the myxoid (most common) or mesenchymal varieties.

Our case was a very young child with extraskeletal chondrosarcoma of conventional type not reported in the literature.

Pediatric chondrosarcomas: a retrospective review of 17 cases

- **Extraskeletal mesenchymal chondrosarcoma** typically affects young adults between 15 and 35 years of age, with a female predilection.
- In a study from Italy, the clinical, radiological, and pathological features of patients with chondrosarcoma referring to a single institution has been reported. There were 17 patients ranging from 13 to 17 years (median 15 years).
- The tumors were central, periosteal and peripheral, the latter arising from multiple exostoses.
- They involved the femur, tibia, pelvic bones, humerus and metacarpal bones.

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Chondrosarcomas in children and adolescents

Cartilaginous tumors can be quite challenging to diagnose. It is best for these lesions to be discussed in a multidisciplinary meeting which includes a radiologist and a pathologist specializing in bone tumors.

Treatment principles are similar to those in adults, with adequate surgical excision respecting oncologic principles being the mainstay of treatment.

Extremity Grade I chondrosarcomas may be managed with extended intralesional curettage without increasing the risk for local recurrence or metastatic disease, but case selection is critical and should be based on clinical, imaging and histological characteristics.

Chondrosarcomas are resistant to chemotherapy and relatively radioresistant. For mesenchymal chondrosarcomas, there may be a role for chemotherapy, though data on this is limited.

Prognosis and rate of recurrence correlate directly to the adequacy of the surgical resection.

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Case report

Extraskeletal conventional chondrosarcoma of genitalia in a child- An unusual pathology for a rare tumor

Gholamreza Ebrahimisaraj [a], Samin Alavi [b,*], Maryam Kazemi Aghdam [c], Mitra Khalili [d], Zahra Khaffafpour [b], Manouchehr Ebrahimian [a]

Palliative Care in Pediatric Patients with Sarcomas

Pediatric patients with sarcomas experience symptoms such as pain, muscle weakness, difficult walking, constipation, loss of appetite, nausea, vomiting, confusion leading to significant morbidity and compromised quality of life throughout their course.

These times could be viewed as opportunities for increased subspecialty palliative care (PC).

Supportive and Palliative Care Ward in Mofid Children's Hospital