

# CASE REPORT

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## UNUSUAL PRESENTATION OF EXTRASKELETAL MESENCHYMAL CHONDROSARCOMA OF ABDOMEN IN 10 YR OLD GIRL- A CASE REPORT

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**ABSTRACT:** Mesenchymal chondrosarcoma (MSC) is rare form of chondrosarcoma which usually arises in bone. Extraskeletal mesenchymal chondrosarcomas are far less common and accounts approximately 30–50% of all mesenchymal chondrosarcomas. We report a rare case of intra-abdominal extraskeletal MSC in a 10 yr old girl who presented with biliary vomitings and abdominal pain. Ultrasound abdomen showed 42x24 mm hypoechoic mass in right iliac fossa. CT abdomen showed calcified granulomas in spleen, Soft tissue dense lesion in right iliac fossa, suggestive of lymphadenopathy. Histopathology and immunohistochemistry confirmed the diagnosis of MSC.

**KEYWORDS:** Mesenchymal chondrosarcoma (MSC), extraskeletal, intra-abdominal.

**INTRODUCTION:** Mesenchymal chondrosarcoma (MSC) is a rare variant of chondrosarcoma constituting approximately 1% of chondrosarcomas. It is characterized microscopically by a dimorphic pattern in which areas of well-differentiated cartilage alternate with undifferentiated stroma. The undifferentiated element is composed of small cells and can be confused with malignant lymphoma, hemangiopericytoma and Ewings sarcoma.

Most patients are females in the second or third decade of life. The bones most commonly affected are the jaw, pelvis, femur, ribs, and spine. One third of these neoplasms involve the extraosseous structures, such as orbit, paraspinal region, meninges, or soft tissues of the extremities. The prognosis is generally poor, although there is great variability in the clinical course.

**CASE REPORT:** A 10 yr old girl presented with bilious vomiting and abdominal pain of 10 days duration. O/E there was mild abdominal distention, no mass palpable per abdomen, clinically diagnosed as intestinal obstruction. Routine hematological and biochemical investigations were done and they were within normal limits. Ultrasound abdomen showed 42x24mm hypoechoic mass in the right iliac fossa, suspicious of lymph nodal mass or SOL. CT abdomen showed calcified granulomas in spleen, Soft tissue dense lesion in right iliac fossa was suggestive of lymphadenopathy. At surgery, exploratory laparotomy, debulking and appendectomy were

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done. Small bowel obstruction due to omental band was noticed. Nodular mucinous pelvic mass and mesenteric lymphadenopathy was noted.

## **HISTOPATHOLOGICAL EXAMINATION:**

### **GROSS:**

We received multiple grey white, slimy, soft tissue masses, soft in consistency and omentum measuring 7× 3.5 cm with multiple tumor nodules, cut section showing grey white slimy areas. Appendix with normal serosa measuring 5 cm, cut surface showed faecolith. Another specimen sent as mucinous material was grey white, soft tissue bits measuring 3x3 cm.

**MICROSCOPY:** Multiple sections from pelvic tumor mass showed poorly differentiated small cells arranged in a hemangiopericytoma pattern along with islands of well differentiated cartilage. Diagnosis of mesenchymal chondrosarcoma was made. Immunohistochemistry (IHC) marker S-100 was done to support the diagnosis. Sections from omentum showed deposits similar to the MSC tumor deposits. Sections from appendix show histopathological features of chronic appendicitis with *Enterobius vermicularis* in the lumen.

**DISCUSSION:** Mesenchymal chondrosarcoma was first described as a distinct entity by Lichtenstein and Bernstein in 1959. Chondrosarcomas are a heterogeneous group of malignant tumors of cartilaginous origin. Although most chondrosarcomas arise from cartilaginous or bony structures, they may also develop in extraskkeletal locations such as in soft tissues where cartilage is normally not found<sup>1</sup>. Compared with conventional chondrosarcomas, mesenchymal chondrosarcomas commonly have an extraskkeletal location, earlier age at presentation, female predominance, high chances of recurrence or metastases, and have poorer prognosis<sup>1</sup>. Extraskkeletal MSC most often occur in the head and neck region, including the orbit, meninges and in the lower extremities, thigh being the most common site. However, the tumor has also been reported in a variety of unusual sites, including the heart, femoral vein, abdomen, lung and kidney<sup>3</sup>. Presenting symptoms of extraskkeletal mesenchymal chondrosarcoma vary with the sites of involvement. The common signs are localized pain, swelling and neurological symptoms. The duration of symptoms vary<sup>2</sup>.

The pathogenesis of mesenchymal chondrosarcoma is unknown, but its origin from primitive mesenchymal cells rather than from preformed cartilage seems most likely<sup>4</sup>. Microscopically, mesenchymal chondrosarcoma exhibits a characteristic pattern composed of sheets of undifferentiated round, oval, or spindle-shaped cells with an abrupt transition by small well-defined nodules of well-differentiated, benign appearing hyaline cartilage, frequently with central calcification and ossification. Immunohistochemically, the cartilaginous portion of the tumor typically shows strong S-100 protein positivity, whereas very few isolated cells in the undifferentiated areas stain for this antigen<sup>6</sup>.

In our case, tumor mass showed islands of well differentiated cartilage surrounded by poorly differentiated small cells arranged in a hemangiopericytoma like pattern. IHC with S-100 showed strong positivity in cartilaginous areas and isolated staining in small round cells. More recently, Wehrli and colleagues reported on the utility of Sox9, a transcription factor thought to be a master regulator of chondrogenesis, as helpful in distinguishing mesenchymal chondrosarcoma from other small round blue cell tumors<sup>5</sup>. Mesenchymal chondrosarcoma is a fully malignant tumor that pursues an aggressive clinical course and metastasizes in a high

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percentage of cases. The principal metastatic site is the lung. Combined radical surgery and chemotherapy or radiotherapy appears to be the treatment of choice.

**CONCLUSION:** Mesenchymal chondrosarcoma though rare, should be considered in the differential diagnosis of soft tissue lesions, as one third of the cases occur extraskeletally. In our case it presented intra abdominally. In the absence of cartilaginous foci its differentiation from other small round cell tumors can be difficult especially in centers which handle pediatric cases, in such instances IHC can be done for confirmation. Due to its aggressive course it is treated by surgery followed by chemotherapy. On follow up our patient has been doing well.

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Figure 1: Gross showing multiple soft tissue bits.

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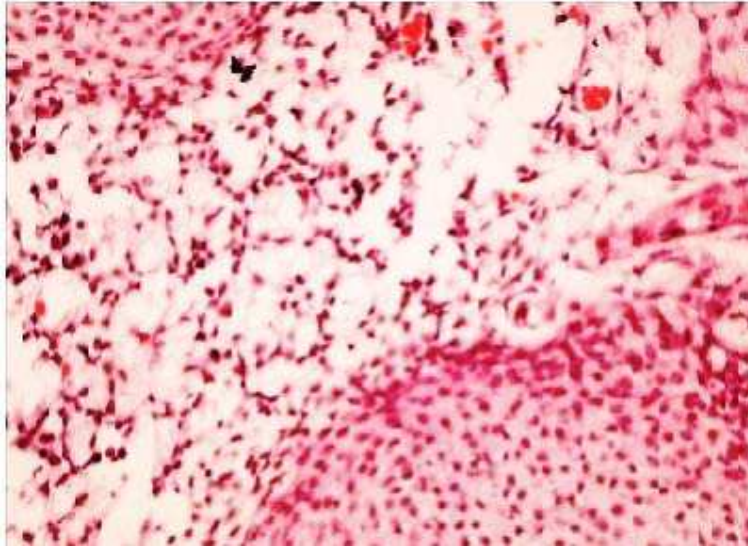


Figure 2: H&E showing well differentiated cartilage with undifferentiated stroma.

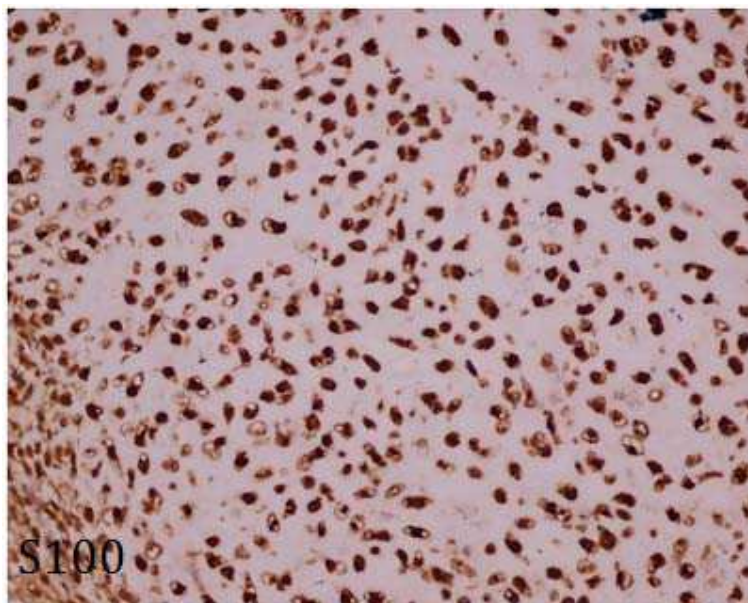


Figure 3: Showing S-100 positivity in cartilaginous area.