Case Report

Extraskeletal mesenchymal chondrosarcoma of the abdomen: A rare clinical presentation

Mohammad Habib Raza[1], Mohammad Ibrahim Raza[2], Mohd Sadik Akhtar[3], Shagufta Qadri[4]

[1]Professor, Department of Surgery, [2]Junior Resident, [3]Associate Professor, Department of Surgery, [4]Assistant Professor, Department of Pathology, Aligarh Muslim University, Aligarh, Uttar Pradesh, India

ABSTRACT

Extraskeletal mesenchymal chondrosarcoma (EMCS) is a rare malignant soft tissue tumor of chondroprogenitor cell origin. Originally, it was restricted to the bone only but that is no longer the case. Recent literature reports that 20–33% of these tumors occur at the extraskeletal sites. We report one such case, in which the tumor involved the anterior abdominal wall muscles and also had a large intra-abdominal mass that covered a large part of the peritoneal cavity. The clinical features and computed tomography findings suggested the diagnosis of a malignant desmoid tumor with intra-abdominal extension; however, the histopathological examination and the immunohistochemistry proved the tumor to be EMCS. The case is reported due to the dilemma in diagnosis, its rarity, large size, parietal, and intra-abdominal extension with multiple site involvement.

Key words: Extraskeletal mesenchymal chondrosarcoma, Desmoid tumor, Chondroprogenitor cells, Abdominal mesenchymal chondrosarcoma

Extraskeletal mesenchymal chondrosarcoma (EMCS) is a rare and aggressive pathological variant of chondrosarcoma arising from the soft tissues of mainly the extremities, meninges/dura, trunk, and orbits. Although the head and neck regions are the most common sites, EMCS has been reported from the thoracic, abdominal, and retroperitoneal organs also. It usually affects the second and third decades of life and is more common in women [1]. Histologically, mesenchymal chondrosarcoma has a characteristic dimorphic pattern composed of islands of hyaline cartilage admixed with highly undifferentiated small round cells. Radiographically, it appears as a soft tissue lobulated mass with various patterns of calcification [2]. The treatment protocol for this tumor has not been well established because of its rarity. Local recurrence and metastasis are common; hence, surgical resection with adequate margins is considered the gold standard of treatment.

CASE REPORT

A 32-year-old female patient presented with complaints of lumps in the abdomen for 9 months and pain in the abdomen for 6 months. She had been operated on for post-partum infectious pathology, which was not documented, and also underwent surgery for a fibroid uterus 2 years back.

Her general condition was poor, she had marked pallor, and the vitals were within normal range except for tachycardia. Abdominal examination showed a midline scar from the previous surgery. Two large lumps, one parietal in the lower abdomen occupying the hypogastrium and extending to the right and left iliac fossae, and the other lump was intra-abdominal extending to the right and left hypochondrium, lumbar, and umbilical regions (Fig. 1). The parietal lump was hard and fixed to the anterior abdominal wall muscles. The intra-abdominal lump was bosselated and firm in consistency. Other systems were normal. A clinical diagnosis of a malignant desmoid tumor of the abdominal wall with extension into the abdominal cavity was made.

Her white blood count was 11,800/mm, hemoglobin was 8 g/dL, platelet count was 2,98,000/mm^3, blood urea nitrogen was 17, serum creatinine was 0.57 mg/dL, and the international normalized ratio was 1.13. Serum electrolytes were:- sodium-138 mEq/L, potassium-4.4 mEq/L, and calcium-8.3 mg/dL.

Abdominal ultrasound showed multiple large, irregular lobulated heterogeneous hypoechoic lesions with cystic areas within it, completely filling the abdominal cavity and displaying the bowel loops and bilateral kidneys. Contrast-enhanced computed tomography (CECT) of the abdomen revealed a large ill-defined heterogeneous enhancing lobulated mass lesion centered

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Correspondence to: Prof. Mohammad Habib Raza, Department of Surgery, JN Medical College, Aligarh Muslim University, Aligarh, Uttar Pradesh - 202 002, India. E-mail: prof.mh.raza@gmail.com

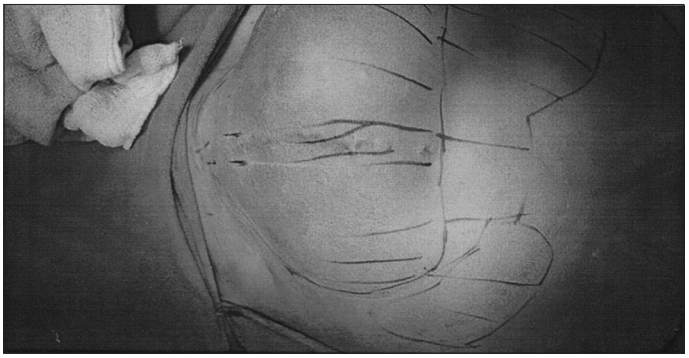

Figure 1: Extent of the abdominal mass. The lower marking represents the parietal extent, the upper marking represents the intra-abdominal lesion, and also midline scar of previous surgery is seen

in the anterior abdominal wall in midline involving the recti and adjacent aponeurotic fascia growing up to the skin with extension into the pre-peritoneal and intra-abdominal space involving the omentum and displacing most of the small bowel loops and abutting antimesenteric border of the jejunum and also forming separate soft tissue density deposits along mesenteric folds in the right and left iliac fossa. The CECT delineated the extent of the tumor and revealed its relation to the adjacent structures. A diagnosis of a desmoid tumor with malignant transformation was made on the basis of clinical and radiological findings.

She was given four units of blood transfusion and evaluated by the anesthesia team for fitness for major abdominal surgery. After adequate preoperative preparation and informed consent, she was subjected to exploratory laparotomy. A large midline incision was given, which included the previous surgical scar. On operation, two large tumors, one intra-parietal in the lower abdomen and the other intra-abdominal occupying almost the whole of the peritoneal cavity but not infiltrating into the small bowel, colon, or solid organs, were found. There were small cystic tumors about 5–10 cm in size arising from the small bowel mesentery at multiple sites. The parietal tumor appeared to arise from the abdominal muscles of the lower abdomen, which was hard in consistency near its attachment to the abdominal muscles. Wide excision of the parietal tumor followed by excision of the intra-abdominal masses and local excision of the small cystic lesions attached to the mesentery was done. The abdomen was closed in layers with two drains, one in the abdominal cavity and the other in the subcutaneous plane.

The excised specimen was weighed, measured, and sent for histopathological examination. The weight of the excised specimen was 7 Kg. The tumor measured approximately 34 × 18 cm in its greatest dimension (Fig. 2).

The patient was kept in the high-dependency unit for 48 h. She had an uneventful post-operative recovery and was discharged in good general condition on the seventh postoperative day after the removal of the drains. She was advised for a regular follow-up.

The histopathological report showed alternating nodular hypo and hypercellular areas with intervening fibrous, myxoid, osteoid, and chondroid areas in between. The lobules contain undifferentiated small cells with minimal atypia, hyperchromasia, inconspicuous nuclei, and scant cytoplasm. Occasional cells were

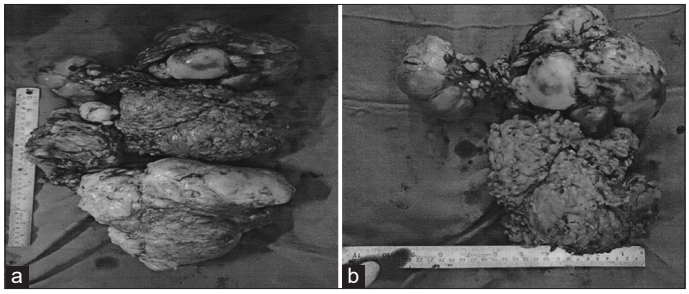

Figure 2: (a) Excised specimen, the tumor consisted of solid and cystic areas; (b) excised tumor, the solid component is from the parietal mass and the cystic component is from the intra-abdominal mass

showed vacuolation. These tumor cells are suspended in copious amounts of the myxoid matrix. Mitotic rate was 2 × 10 HPF, necrosis was less than 50%, and histological grade (FNCLCC) grade was 2 5-1+1. The margin could not be assessed, and lymphovascular invasion was not identified (Fig. 3). Regional lymph nodes were not submitted. On immunohistochemistry (IHC), tumor cells show strong, diffuse cytoplasmic positivity. Histopathological diagnosis and IHC findings were suggestive of the diagnosis of EMCS TNM- pT_4N_0. After the confirmation of the diagnosis on the basis of histopathological examination and IHC, the patient was subject to chemotherapy. The patient was advised six cycles of ifosfamide + doxorubicin combination.

DISCUSSION

EMCS is a rare malignant soft tissue tumor of chondroprogenitor cell origin. Mesenchymal chondrosarcoma was first described by Lichtenstein and Bernstein in 1959 [1]. Originally, it was considered restricted to the bone, but that is no longer the case. More recent literature reports that 20–33% of these tumors occur at the extraskeletal sites. It is a high-grade malignancy with a strong tendency for distant metastasis. Most frequently, the tumor occurs in young adults during the second or third decades of life with a slight preponderance in females. In our case also, the patient was a female in the third decade of life.

The tumor is found more commonly in the head and neck regions, including the face, orbit, brain, meninges, and lower extremities [3,4]. Based on tumor location, two separate categories of EMCS have been previously described: one occurring in the skeletal muscle and soft tissue and the other in the central nervous system and spinal cord. Although the abdomen is an important site for its occurrence, the extent and the unique presentation mimicking the desmoid tumor have not been described earlier.

Chen and Chou have reported a case of unusual presentation with a pancreatic tail mass, a first distant metastasis from a palpable chest wall mass which was diagnosed as mesenchymal chondrosarcoma [5]. Lee *et al*. have reported a case of EMCS of the uterus in a 33-year-old female. This was the second case to be reported in the literature [6]. Chu *et al*. have reported a case of intracranial extraskeletal chondrosarcoma which was diagnosed only after the histopathological examination as imaging was not clear in establishing the diagnosis [7]. Chatzipantelis *et al*.

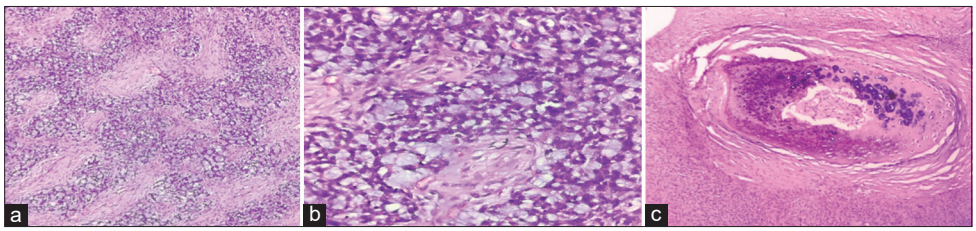

Figure 3: Histopathological section shows (a) a tumor with hypercellular nodules with ovoid cells embedded within the chondromyxoid stroma with hypocellular areas with fibrocollagenous tissue (H and E ×40); (b) section shows ovoid to round tumor cells in a chondromyxoid matrix. The cells show mild-to-moderate pleomorphism with vesicular nuclei and scant cytoplasm (H and E ×40); (c) section shows areas with cartilage and foci of ossified bone formation (H and E ×40)

have reported two rare cases of mesenchymal metastatic tumors in the pancreas [8]. Sun *et al*. reported a case of mesenchymal chondrosarcoma with pancreatic and adrenal metastasis [9].

On computed tomography scan, EMCS appears as a well-circumscribed soft tissue mass with areas of granular ring, arc, or irregularly shaped calcifications. The non-calcified areas of the tumor are hypodense compared to the surrounding soft tissue. These findings are non-specific to EMCS and can be seen in other benign cartilaginous tumors [10]. In the present case, the CECT of the abdomen revealed a large ill-defined heterogeneous enhancing lobulated mass lesion centered in the anterior abdominal wall in midline involving the recti and adjacent aponeurotic fascia growing up to the skin with extension into the pre-peritoneal and intra-abdominal space involving the omentum and displacing most of the small bowel loops. On the basis of clinical examination and imaging, a provisional diagnosis of a desmoid tumor with malignant transformation was made.

Usually, EMCS ranged from 5 to 12 cm. Our tumor was 34 × 18 cm and weighed 7 Kg. Such an extensive tumor and large size have not been reported in the literature. Surgical resection with adequate margins is considered the gold standard of treatment in EMCS; however, in our case, considering the huge size and complex nature of the tumor, it was not possible to achieve a tumor-free margin; hence, the patient was advised six cycles of chemotherapy.

The present case is unique because the tumor was involving the anterior abdominal wall muscles and extensive intra-abdominal extension; portions of the tumor, which were parietal, were hard in consistency at places; whereas, the intra-abdominal lesion was soft and cystic in nature; both extra-abdominal and intra-abdominal portions were fixed together in the lower abdomen; the weight of the excised specimen was approximately 7 Kg, and the size was 34 × 18 cm; such a large EMCS has not been reported earlier. Histopathological examination and IHC were required for confirmation of the diagnosis, which was at variance with the imaging studies.

CONCLUSION

Clinical examination and imaging included suggested the diagnosis of a desmoid tumor in our case; however, the exact diagnosis of EMCS was made only after surgical excision, histopathology, and IHC. We suggest that EMCS should be included in the differential diagnosis of desmoid tumors having a large size and intra-abdominal extension. Confirmation of diagnosis should be done by histopathology and IHC. EMCS is highly aggressive and requires a different approach to management.

REFERENCES

1. Lichtenstein L, Bernstein D. Unusual benign and malignant chondroid tumors of bone. A survey of some mesenchymal cartilage tumors and malignant chondroblastic tumors, including a few multicentric ones, as well as many atypical benign chondroblastomas and chondromyxoid fibromas. Cancer 1959;12:1142-57.
2. Weiss SW, Goldblum JR. Cartilaginous soft tissue tumors. In: Enzinger and Weiss's Soft Tissue Tumors. 5th ed. Amsterdam: Mosby Elsevier; 2007. p. 1031-6.
3. Nakashima Y, Uni KK, Shives TC, Swee RG, Dahlin DC. Mesenchymal chondrosarcoma of bone and soft tissue. A review of 111 cases. Cancer 1986;57:2444-53.
4. Shaked RJ, Geller DS, Gorlick R, Dorfman HD. Mesenchymal chondrosarcoma: Clinicopathologic study of 20 cases. Arch Pathol Lab Med 2012;136:61-75.
5. Chen JJ, Chou CW. A rare case report of mesenchymal chondrosarcoma with pancreatic metastasis. Medicina (Kaunas) 2022;58:639.
6. Lee Y, Choi S, Kim HS. Extraskeletal mesenchymal chondrosarcoma of the uterus. Diagnostics (Basel) 2022;12:643.
7. Chu J, Ma H, Wang Y, Li K, Liao C, Ding Y. CT and MRI findings of intracranial extraskeletal mesenchymal chondrosarcoma-a case report and literature review. Transl Cancer Res 2022;11:3409-15.
8. Chatzipantelis P, Karvouni E, Fragoulidis GP, Voros D, Pafiti A. Clinicopathologic features of two rare cases of mesenchymal metastatic tumors in the pancreas: Review of the literature. Pancreas 2006;33:301-3.
9. Sun J, Zhang W, He T, Wang H, Tian R. 18F-FDG PET/CT imaging of pancreatic and adrenal metastases in a patient with mesenchymal chondrosarcoma. Clin Nucl Med 2021;46:231-2.
10. Arora K, Riddle ND. Extraskeletal mesenchymal chondrosarcoma. Arch Pathol Lab Med 2018;142:1421-4.

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