International Journal of Research in Medical Sciences
Gupta M et al. Int J Res Med Sci. 2023 May;11(5):1837-1840
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012

DOI: https://dx.doi.org/10.18203/2320-6012.ijrms20231369

Case Report

A case report of primary intraabdominal rhabdomyosarcoma presenting as metastasis

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Received: 01 March 2023
Revised: 07 April 2023
Accepted: 10 April 2023

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ABSTRACT

Rhabdomyosarcoma (RMS) is a rare and in adult accounts for only 1-3% of all malignant soft tissue tumors. The most frequent sites of origin is within head and neck area. One of the least common sites is the retrorectal-presacral space. This case of 61 years old female is probably the fifth well-documented case of primary abdominopelvic RMS. She presented with abdominal distensions, obstipation, vomiting for 5 days. Excision of the gut wall along with soft tissue mass was done and sent to our department for histopathological examination. On microscopic examination, a diagnosis of malignant mesenchymal tumor with closest resemblance to RMS was made. It is a rare case and needs to bring in notice as there is very few information regarding intraabdominal RMS. This case initially thought to be metastasis from gynecologic malignancy. It is important for pathologists, gynecologists and radiologists to recognize RMS as differential diagnosis of masses arising in abdomen and pelvis.

Keywords: Primary, Intestinal, RMS, Adult

INTRODUCTION

Rhabdomyosarcoma (RMS) is a soft tissue sarcoma, which resembles histologically embryonic skeletal muscle. It can be seen anywhere in the body, including tissues devoid of skeletal muscles. The cell of origin of it remains unknown though recent evidence suggests that RMS can originate from aberrant development of non-myogenic cells.[1] RMS is rare malignancy and typically observed in childhood and adolescence. Very few of RMS cases have been reported to develop in the adult population older than 20 years.[2] The most frequent sites of origin is within head and neck area, followed by the genitourinary tract, extremities, trunk, retroperitoneum. One of the least common sites for RMS is the retrorectal-presacral space.[3-5]

Primary abdominopelvic RMS is very rare tumor although soft-tissue sarcomas such as liposarcoma, leiomyosarcoma and gastrointestinal stromal tumor can be seen more commonly. At the time of diagnosis, abdominopelvic RMS is usually presents as a large mass due to its clinically silent characteristics until it compresses or invades vital organs. Thus, this kind of tumor in female adults is commonly misdiagnosed as genital organ malignancy due to similar clinical presentations. Magnetic resonance imaging (MRI) is the most appropriate modality for evaluation of tumors in the pelvis and abdomen.[6] Multi-institutional trials have not been performed, and only case reports have been published. Our case is probably the fifth well-documented case of primary abdominopelvic RMS reported in literature.

CASE REPORT

A 61 years old female patient came to outpatient department of PGIMS, Rohtak with complain of abdominal distensions, obstipation, nausea and vomiting for 5 days. She had history of weight loss since 6 months

and was a known case of carcinoma ovary. She underwent oophorectomy 1 year back and now on radiotherapy for the same. There was no significant history of diabetes mellitus, hypertension although known case of hypothyroidism. She attained menopause at age of 49 years. On examination, she was pale, temperature was normal, pulse rate was 82/min and blood pressure was 130/90 mm Hg. On abdominal examination, distension was present along with diffuse tenderness with no guarding and rigidity. No organomegaly was present.

All lab investigations were within normal limits. X-ray erect abdomen show multiple air fluid levels suggestive of intestinal obstruction. CECT abdomen showed multiple large, solid, mildly enhancing peritoneal mass lesions, largest mass of size 40×25×15 mm arising from the subhepatic region and extending upto the right iliac fossa adjoining the bowel loops (suggestive of neoplastic etiology) causing obstruction.

Patient was shifted to operation theatre for emergency exploratory laparotomy. Upon exploration, multiple soft to firm tissue masses attached to small intestine were identified on mesenteric portion. Excision of the gut wall along with soft tissue mass was done, followed by end to end anastomosis. Rest organs were free from mass. No organ of origin could be specified. Resected gut was sent to our department for histopathological examination.

We received a gut segment measuring 19 cm in length along with attached mesentry. On mesentric portion, two growths identified measuring 4×4×2 and 4×3×1 cm respectively (Figure 1). Grossly, both cut ends of the gut were free from tumor. On sectioning the masses, grey white areas along with haemorrhage and necrosis identified. No calcification seen. The tumor appears to be involving the intestinal wall grossly. Sections were taken as per protocol.

Figure 1: Two growths (tumor masses), measuring 4×4× 2 and 4×3×1 cm respectively on mesentric portion of gut.

On microscopic examination, tumor cells were arranged in lobules and nests. These cells have high N:C ratio, fine chromatin, prominent eosinophilic nucleoli along with moderate amount of cytoplasm (Figure 2). Few polynuclear cells and cells with bizarre enlarged nuclei could be detected. Few mitosis seen (2-3/HPF). No necrosis was identified. These tumor cells were reaching upto to muscularis propria. CK, inhibin, CA125 were negative, thus ruling out possibility of metastasis from ovarian cancer or any other carcinoma while vimentin, Myogenin, Myo d1 came positive favouring mesenchymal tumor (Figure 3). A diagnosis of malignant mesenchymal tumor with closest resemblance to RMS was made.

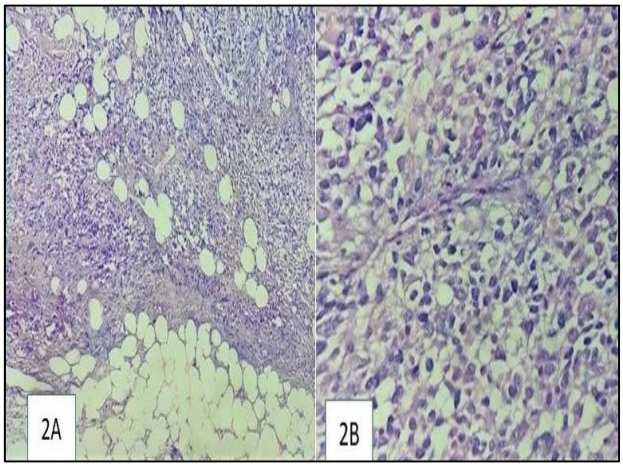

Figure 2 (A and B): Microphotograph of haematoxylin and eosin stained section of tumor masses (100X), microphotograph of haematoxylin and eosin stained section of tumor masses (400X).

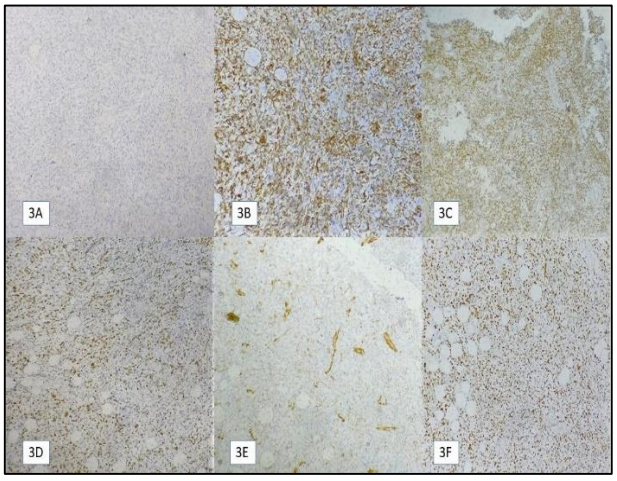

Figure 3 (A-F): Tumor cells are negative for CK (100X), tumor cells showing bright positivity for vimentin (100X), tumor cells showing bright positivity for Myo D1 (100X), tumor cells showing bright positivity for myogenin (100X), tumor cells are negative for SMA (100X) and tumor cells showing positivity for ki 67 in >20% cells (100X).

DISCUSSION

RMS is most common sarcoma in paediatric age group. RMS is very uncommon in adults and is thought to arise from immature mesenchymal cells that are committed to skeletal muscle lineage.[3]

The WHO divided RMS into four distinct subtypes: embryonal, alveolar, pleomorphic and spindle cell/sclerosing. The embryonal type is seen more commonly in childhood whereas pleomorphic type occur in older adults with a mean age of 51 years at diagnosis and alveolar tumors affect all age groups. This age distribution is in accordance with the histologic maturity of the RMS subtype as embryonal types resemble embryonic tissues and pleomorphic represent aggressive adult carcinomas with malignant fibrous histiocytoma like features. RMS cells express desmin along with myogenic transcription factors MyoD1 or myogenin. Due to the high sensitivity and specificity of MyoD1 in the diagnosis of RMS, it is an essential tumor marker to identify RMS from other non-RMS.[11,12]

RMS are well-circumscribed un-encapsulated firm, nodular masses on gross appearance and of variable size and consistency. However, they often tend to infilterate extensively into adjacent tissues. On light microscopy, diagnosis of RMS is based on the identification of characteristics of skeletal muscle i.e., crossstriations or rhabdomyoblasts. Histologically embryonal RMS (ERMS) is composed of rhabdomyoblasts and small round cells. Rhabdomyoblast is the more mature of the embryonal component and characterized by bright eosinophilic cytoplasm. In pleomorphic RMS, anaplastic cells are present in large aggregates or as diffuse sheets. It occurs in the extremities and the trunk. The diagnostic electron microscopic features of RMS are visible z-bands. Skeletal muscle or muscle-specific proteins, like antidesmin, muscle-specific actin and Myo D can be identified by immunohistochemical staining.[13]

Because there are very few reported cases of intestinal RMS so it is not possible to speculate on its behaviour, response to treatment, or prognosis. However, RMS outcomes are considerably less favourable for adults than for children.[14,15]

The optimal management of RMS in adults is unknown due to its rarity and the absence of any standard treatment protocol or guidelines. The current treatment standards were proposed by the intergroup RMS studies (IRS). These standards include multimodal therapy (MMT: resection, chemotherapy, and radiation). Surgery is the mainstay of treatment for adult RMS. Though as per protocol all RMS patients should undergo radiotherapy to achieve long-term local control of the tumour.[16]

Among the differential diagnosis, following options were considered. Neuroendocrine carcinoma consists of uniform small-to-medium-sized cells, with indistinct cytoplasmic boundaries and round regular nuclei. These cells are normally arranged in nested patterns. Areas of necrosis and presence of >2 mitotic figures/HPF are highly indicative of malignancy. Tumor cells are positive for chromogranin A, synaptophysin, CD56, NSE and negative for MyoD1, myogenin, myoglobin, and desmin. Biopsy from poorly differentiated adenocarcinoma reveals small tumor cells with a loose glandular nest structure of fusiform or irregular cancer cells. There may be 1-5 mitotic figures/HPF. The tumor is positive for CKpan, CK8/18, CK19, CK20, EMA, and CEA, negative for MyoD1, myogenin, myoglobin, desmin, and actin. Gastric epithelioid malignant melanoma is usually located in the lamina propria of the mucosa in the early stages and shows an adenoid, solid, nest-like pattern with little fibrous connective tissue. These cells have richly basophilic cytoplasm with numerous melanin particles. A large, strongly eosinophilic nucleolus occupies ≥80% of the nucleus. These tumors are strongly positive for HMB45 and MART-1, negative for MyoD1, myogenin, myoglobin, desmin, and actin. Gastrointestinal stromal tumor (GIST) is the most common mesenchymal tumor of the gastrointestinal tract. They commonly develop in the 60-65 years. Histological appearance varies from the spindle cell type (most common) to epithelioid cell type. The pleomorphic cell type exhibits sarcomatoid characteristics, with a large number of atypical nuclei and mitotic figures. Most gastric GISTs are positive for CD117 and DOG1 and partially positive for CD34 and S1-00. They are negative for MyoD1, myogenin, myoglobin, desmin, and actin. Plasmablastic lymphoma is mostly composed of large cells resembling B immunoblasts, but the morphology may vary widely. The tumor cells form nested, adenoid structures in some areas. Mitotic figures are present, as also scattered multinucleated giant cells and macrophages that phagocytose dyeable small bodies. Large necrotic areas may be present. Tumor cells are positive for positive CD138, CD38, and IRP4/MUM1. About 50-80% of cases are positive for CD79a. There is no expression of MyoD1, myogenin, myoglobin, desmin, or actin. Myeloid sarcoma in the stomach is a localized tumor formed by extramedullary proliferation and infiltration of myeloid primordial cells or immature myeloid cells. These tumors are positive for MPO, lysozyme, CD68, and CD117, but negative for CD3, MyoD1, myogenin, and myoglobin.[17-23]

CONCLUSION

The RMS in peritoneal mass including intestine is a rare case and needs to bring in notice as there is very few information regarding intraabdominal rhabdomyosarcoma. This case initially thought to be metastasis from a gynecologic malignancy. It is important for pathologists, gynecologists and radiologists to recognize RMS so, it can be listed in the differential diagnosis of masses arising in the abdomen and pelvis. Better awareness of its features and the differential diagnoses will help in early diagnosis and treatment.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Gupta M, Kaushik N, Prasad SSP, Singh S. A case report of primary intraabdominal rhabdomyosarcoma presenting as metastasis. Int J Res Med Sci 2023;11:1837-40.