Primary Intra-Abdominal Rhabdomyosarcoma in an Adult: an Unusual Presentation and Review of Literature

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Abstract Rhabdomyosarcomas, the commonest soft tissue sarcoma in paediatric age group, represent 5–10 % of all solid tumors in childhood. These tumors are rare in adult population. There are sporadic case reports of intra-abdominal rhabdomyosarcoma, but mostly in paediatric age group. Here we are reporting an unusual case of intra abdominal rhabdomyosarcoma in an elderly which presented with acute intestinal obstruction. CECT abdomen showed a solid mass in retroperitoneum compressing adjacent bowel loops. Upon emergency exploratory laparotomy a hard mass was found to be compressing the ascending colon and infiltrating into duodenum, pancreas and liver. No specific organ of origin could be identified. Biopsy was taken, as the mass was unresectable and ileostomy was done. Histopathological examination revealed embryonal rhabdomyosarcoma. Rarity of this disease and unusual presentation prompted us to report this case.

Keywords Rhabdomyosarcoma · Soft tissue sarcoma

Introduction

Tumors arising from soft tissues are uncommon in children, accounting for about 6 % of all childhood malignancies. More than half (53 %) of these soft tissue sarcomas (STS) originate from the striated muscles and are called rhabdomyosarcomas (RMS) [1, 2] Rhabdomyosarcomas are rare in adults, and the literature regarding their management is limited. Truncal and extremity tumors are more common in adults, and the relative proportion of pleomorphic tumors increases with age [3]. Intra-abdominal rhabdomyosarcoma is still rarer in adult population. Collaborative pediatric trials have revolutionized the care for this neoplasm; approximately 70 % of children with nonmetastatic disease are currently cured using multimodality treatment [4]. Unfortunately, this success has not translated into better cure rates for adults with RMS, who continue to have a very poor prognosis (overall survival rates of only 20 to 40 %) [5]. Due to the rarity of adult RMS, information regarding its clinical and biologic characteristics is very limited; large, multi-institutional trials have not been performed, and only case reports have been published. Ours is probably the second case of intra abdominal rhabdomyosarcoma reported in literature.

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Case Summary

A 65 years old male patient was brought to our central emergency with complaints of pain abdomen, vomiting, abdominal distension and total obstipation for 4 days duration. On examination his vitals were stable, abdomen distended with diffuse tenderness but no guarding and rigidity. X ray erect abdomen was done which showed multiple air fluid levels. CECT abdomen showed a large, solid, mildly enhancing mass lesion of size 23×20×15 cm arising from the subhepatic region and extending upto the right iliac fossa, pushing and compressing

the adjacent bowel loops (Fig. 1). Patient was shifted to OT for emergency exploratory laparotomy. Upon exploration, there was a hard mass originating from retroperitoneum and occupying whole of the right half of abdominal cavity and compressing ascending colon. Mass was infiltrating into pancreas, duodenum and liver. Both kidneys were free from mass. No organ of origin could be specified (Fig. 2). Due to unresectability of the mass, incisional biopsy was taken and ileostomy was made.

Histopathological examination of tissue showed sheets of predominantly small and round cells with abundant and eccentric eosinophilic cytoplasm, small oval nuclei with prominent nucleolus. Few spindle cell myoblasts with prominent tapered fibrillar eosinophilic cytoplasm and cross striations were also present. Highly cellular areas around blood vessels alternating with parvicellular mucoid regions (resembling normal embryonal myogenesis) were seen. Cells stained for myogenin on immunohistochemistry (Fig. 3).

Discussion

Soft tissue sarcomas (STS), a rare entity in paediatric age group can arise from striated muscle cells (53 %) and are called as rhabdomyosarcomas and remaining group (47 %) consists of a heterogeneous collection of subtypes referred to as non-rhabdomyosarcoma soft tissue sarcomas (NRSTS).

Fig. 1 CT abdomen showing solid mass extending from right subhepatic region to right iliac fossa

Fig. 2 Intra operative picture showing mass

Presentation of these STSs differ significantly in paediatric and adult age group. Rhabdomyosarcoma is the commonest STS in children but it is rare in adult population. In children RMS is commonly of the embryonal histology as compared to pleomorphic variety in adults. Similarly among the NRSTS, malignant fibrous histiocytoma (MFH) comprises the most common histology in adults, but is exceedingly rare in children [1, 2].

Rhabdomyosarcoma is thought to arise from immature mesenchymal cells that are committed to skeletal muscle lineage, but these tumors are also known to arise in tissues in which striated muscle is not normally found, such as urinary bladder. The standard classification is still the one proposed by Horn and Enterline [7] in 1958 which divided the tumor into four subgroups: embryonal, alveolar, botryoid and pleomorphic and noted that botryoid was actually a subtype of embryonal [6]. There seems to be a discrete age range characterizing RMS, so that embryonal types occur in infants and toddlers, pleomorphic types arise in adults, and alveolar tumors seem to affect all age groups. Histologically, this age distribution corresponds to the histologic maturity of the RMS subtype, because embryonal types resemble embryonic tissues and pleomorphic types appear as aggressive adult cancers with malignant fibrous histiocytoma like features. However, exceptions do occur in occasional adults, and tumors with the histology of embryonal RMS or its spindle cell variant are occasionally reported [7, 8].

Rhabdomyosarcomas are grossly firm, nodular and of variable size and consistency. They are well circumscribed but not encapsulated and often tend to infiltrate extensively into adjacent tissues. Light microscopy diagnosis of RMS is based on the identification of crossstriations, characteristic of skeletal muscle, or characteristic rhabdomyoblasts. Cross-striations

Fig. 3 Histopathological pictures

are seen in 50–60 % of the cases. Histologically embryonal rhabdomyosarcoma (ERMS) is composed of rhabdomyoblasts and small round cells. Rhabdomyoblast, the more mature of the embryonal component, is characterized by bright eosinophilic cytoplasm. pleomorphic RMS, which is extremely rare in children show anaplastic cells present in large aggregates or as diffuse sheets. It occurs in the extremities and the trunk. Electron microscopy and immunohistochemical analysis of tumors are now useful tools for demonstrating characteristics of RMS, especially when light microscopy is inconclusive. The diagnostic EM 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. Monoclonal antibodies, like those to desmin, musclespecific actin, sarcomeric actin and myoglobin have also been used to confirm the myogenic lineage with very good specificity and sensitivity [9]. Monoclonal antibodies against Myo D can be used in frozen section analysis also.

There are three modalities of treating patients with RMS. These are surgery, radiation therapy for control of residual bulk or microscopic tumor and systemic combination chemotherapy for primary cytoreduction and eradication of gross and micrometastases. Surgery is the most rapid way to ablate the disease and should always be done if subsequent function or cosmesis will not be greatly impaired. Surgery includes complete resection of the primary tumor with surrounding margins of uninvolved tissue during the initial surgery and any subsequent operation. If microscopic residual disease is found after initial resection, re-excision of the area is indicated before any other nonsurgical management. Debulking procedures have no value as initial biopsy and neo-adjuvant therapy results in shrinkage of the tumor allowing complete resection at second look operation [10].

According to IRSG protocols all RMS should get RT to achieve longterm local control of tumors [11]. All patients with RMS must receive combination chemotherapy as soon as diagnosis is made or after resection. It improves survival significantly. IRS-IV recommends a combination chemotherapy approach consisting of vincristine, actinomycin-d, etoposide or ifosfamide and cyclophosphamide. Three year failure free survival ranges from 78 to 92 % with these regimens [12]. Phase II trials from Europe and USA have shown that ifosfamide, as a single agent, is an active drug against RMS. In combination with other drugs the response rate is even better [13]. Irinotecan, a topoisomerase I inhibitor also appears to have promising activity against RMS, with minimal hematopoietic toxicity [14]. Most of these studies are done in paediatric age group.

Iyad sultan et al. in there study reported that adults with rhabdomyosarcoma had significantly worse outcome than children (5-year overall survival rates, 27 %±1.4 % and 61 %±1.4 %, respectively; $P<.0001$). Tumors in adults were more likely to be at an unfavorable site (65 % v 55 %; $P<.0001$) and to have histologies that are unusual during childhood, particularly the pleomorphic subtype (19 %) and not otherwise specified (43 %) [15].

Andrew M. Kaplan et al. reported probably the first well-documented case report of non-hepatobiliary, adult, intra-abdominal embryonal rhabdomyosarcoma in the English language literature in a 57 years old women in 1999 [16]. Extensive search of literature could not yield any other such case report. This makes our case a rare presentation of rhabdomyosarcoma and thus need for reporting.

Conflict of Interest Authors declare no conflict of interest.

Consent from patient's attendant taken for use of patient data for academic purposes.

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