Pediatric intestinal leiomyosarcoma

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ABSTRACT

The paper reports an infant presenting with leiomyosarcoma of the small intestine. The patient presented with intermittent abdominal pain. Examination revealed a hard and mobile intraperitoneal mass. The tumor arose from the mid-ileum with regional lymphadenopathy. Excision of the tumor along with the involved bowel was performed followed by three cycles of chemotherapy. Histological diagnosis was that of a low-grade malignant leiomyosarcoma of the small intestine. Surgical excision was followed by three cycles of chemotherapy. After surgery and three cycles of chemotherapy, the patient was followed up for four years with no evidence of recurrence or metastasis. Surgery followed by chemotherapy was curative for leiomyosarcoma in our patient. Intestinal leiomyosarcoma should be kept as a differential diagnosis for mobile solid intraabdominal tumors in childhood.

KEY WORDS: Intestinal tumors, leiomyosarcoma

INTRODUCTION

Intestinal leiomyosarcoma is an uncommon tumor in the pediatric age group and the presentation of this condition varies considerably. This paper describes an infant with leiomyosarcoma of the ileum, which presented with unexplained abdominal pain and an intraperitoneal mass. Treatment by excision and chemotherapy was curative.

CASE REPORT

A six month-old male child was admitted with a complaint of abdominal pain of two months' duration. Examination revealed a hard, irregular, mobile intraperitoneal mass in the umbilical region. Ultrasound examination and computed tomography (CT) scan of the abdomen showed a soft tissue mass which was displacing the bowel loops; the organ of origin was, however, not clear. Hematological parameters and serum alphafetoprotein were within normal limits. At laparotomy, a large solid tumor (size 8 cm x 7 cm) was found arising from the mid-ileum with satellite nodules proximally and distally [Figure 1]. The lymph nodes adjacent to the bowel were enlarged. The tumor was resected together with all of the involved intestine and mesentery. The child made an uneventful postoperative recovery. The tumor along with the involved lymph nodes was sent for histopathology and immunohistochemistry. Grossly, the tumor was firm in consistency with a grayish-white cut surface. A histological examination showed elongated or fusiform cells arranged in thin interlacing bundles. The tumor cells showed moderate cellular atypia characterized by anisonucleosis, hyperchromatism and raised mitotic index [Figure 2]. Immunohistochemistry was positive for whole muscle actin and desmin. The involved nodes were also positive for malignant cells. Histopathological diagnosis was of leiomyosarcoma (low-grade malignancy) of the small intestine. Chemotherapy (vincristin, actinomycin D and cyclophosphamide) was administered for three cycles. There has been no evidence of recurrence or metastasis at the 4 years' follow-up visit.

DISCUSSION

Intestinal leiomyosarcomas in children are extremely rare with 28 cases previously reported till 1997 in (English) literature. The tumors present mostly in infancy and the cecum and colon are the predominant sites of involvement though origin from the ileum and stomach have also been reported. Most common signs and symptoms were abdominal pain (62%), gastrointestinal bleeding (40%) and / or abdominal mass (38%). Other presentations include intussusception, intestinal obstruction and perforation. Preoperative
imaging studies were not very helpful in establishing the diagnosis in our patient. Both ultrasound and CT scan showed a solid tumor but were unable to determine the site of origin. The only clinical feature of significance was mobility of the mass, which suggested origin from the intestinal mesentery. Diagnosis of leiomyosarcoma of the small intestine was made on the basis of histopathology and immunohistochemistry.

Immunohistochemically, these tumor cells are positive for alpha-smooth muscle actin but not for CD34 or CD117, by which the tumor can be differentiated from gastrointestinal stromal tumors.\(^1\)

Adult intestinal leiomyosarcomas have been graded according to their histology and prognostic factors.\(^4\) In a study by Ng et al.,\(^6\) on intestinal leiomyosarcomas, four factors were identified that were associated with significantly better outcome: complete resection without tumor rupture, localized lesions, low grade of tumor and tumor smaller than 5 cm. A staging system incorporating these prognostic factors of significance was evaluated using TGM system: T (tumor) T\(_1\) (< 5 cm), T\(_2\) (≥ 5 cm), T\(_3\) (contiguous organ invasion or peritoneal implants), T\(_4\) (tumor rupture); G (tumor grade): G\(_1\) (low grade), G\(_2\) (high grade); M (metastasis): M\(_0\) (no metastasis), M\(_1\) (metastasis present).\(^6\) The corresponding overall survival for stages I, II, III, IVA and IVB were 75, 52, 28, 12 and 7% respectively.\(^6\) Surgery remains the primary modality of treatment for patients with gastrointestinal leiomyosarcomas.\(^6\) Most of the cases reported in children have been low-grade malignancies where surgery alone has been curative and no adjuvant treatment has been offered.\(^7\) McGrath et al.,\(^8\) however, report one child with grade 2 leiomyosarcoma dying of sarcomatosis seven years after incomplete resection despite chemotherapy and Gamoudi et al. have reported recurrence following incomplete chemotherapy.\(^9\) Our patient, though having a low-grade tumor on histology, received chemotherapy due to the presence of satellite tumors in the adjacent bowel and enlarged mesenteric lymph nodes. Until more experience with this tumor is reported and the number of patients becomes large enough for statistical analysis, we feel that postoperative chemotherapy is a safer option. In conclusion, intestinal leiomyosarcoma is a differential diagnosis for mobile solid tumors in the abdomen. The prognosis for children seems to be better than these tumors in adults.

REFERENCES


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