Pathologic Quiz Case

Dual Intussusceptions in the Small Intestine

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A 61-year-old woman sought medical attention for pain and swelling in her left lower leg. Radiographs revealed a large destructive lesion involving the distal two thirds of the tibia. Osteosarcoma was diagnosed at curettage biopsy (Figure 1). Four years later, a 5 × 4 × 3.5-cm, presternal, soft tissue mass was noted. Computed tomographic scan delineated a solid lesion involving the sternum and 4 intrapulmonary lesions (the largest 2 cm) involving both lungs. These tumors were clinically thought to represent metastatic osteosarcoma, but core biopsy of the presternal mass revealed low-grade leiomyosarcoma. The presternal leiomyosarcoma had doubled in size by the time the patient returned 9 months later complaining of nausea and vomiting of several months' duration. Computed tomographic scan identified an obstructing ileal intussusception. No other abdominal findings were reported, and no uterine abnormalities were described on computed tomographic scan or at laparotomy. The excised 67-cm segment of ileum contained 2 intussusceptions. Each had a 2.5-cm, pedunculated, polypoid, intraluminal lesion at its leading edge (Figure 2). Between these lead points were a third similar 2.5-cm polypoid lesion and 3 submucosal nodules, measuring 0.2 to 0.9 cm. On sectioning, each polypoid lesion was white and well circumscribed. None involved the muscularis propria (Figure 3). Histologically, all of the polypoid and submucosal tumors consisted of fascicles of spindle cells with enlarged pleomorphic nuclei and frequent mitotic figures (Figure 4).

What is your diagnosis?

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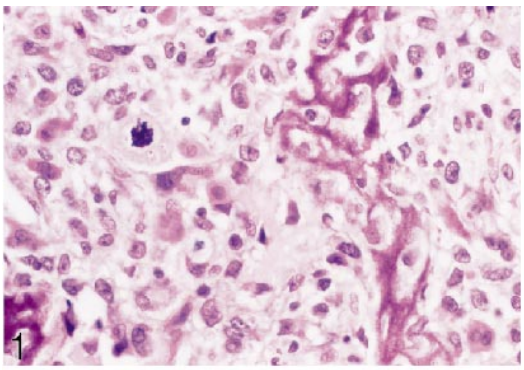

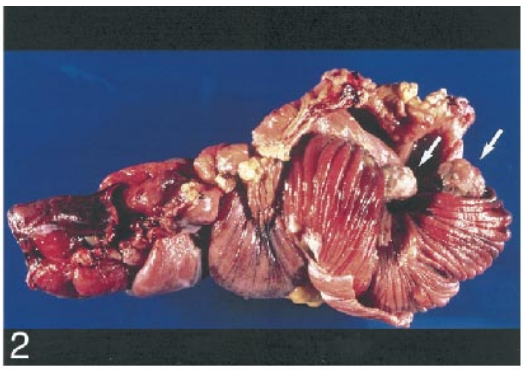

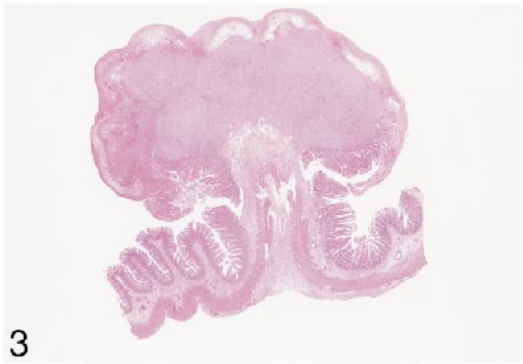

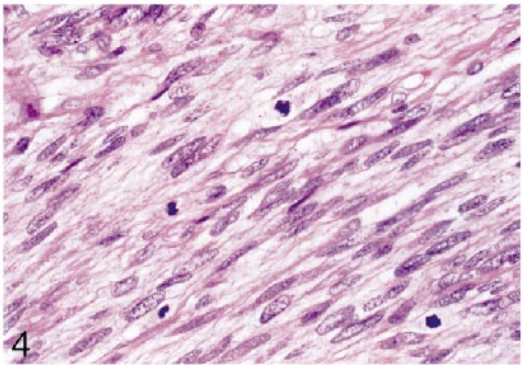

Pathologic Diagnosis: Metastatic Leiomyosarcoma as the Cause of Dual Intussusceptions

The tumor cells were strongly immunoreactive for smooth muscle–specific actin and vimentin and were focally immunoreactive for desmin. These tumors were of higher nuclear grade than the smooth muscle tumor in the prior presternal biopsy and did not resemble the previously diagnosed osteosarcoma (Figure 1).

COMMENT

Intussusception is an infrequent cause of intestinal obstruction in adults, accounting for approximately 1% of bowel obstructions.[1,2] In a review of 261 cases of small and large intestinal intussusception in children and adults, Pang[3] listed the following causes (in order of decreasing frequency): lipoma, benign intestinal polyp, adenocarcinoma, leiomyoma, metastatic carcinoma, lymphoma, and leiomyosarcoma. Nagorney et al[4] found that malignant tumors accounted for the majority (63% of 24 cases) of colonic intussusceptions in adults and that 93% of these were due to primary adenocarcinoma. In contrast, benign conditions caused the majority (71% of 24 cases) of small intestinal intussusceptions. Among the cases attributable to malignancy, 71% were due to metastatic tumor.

Leiomyosarcomas can be primary or secondary in the intestine. Primary lesions in the small intestine are characteristically single and arise from the muscularis propria. They protrude (extraluminally or intraluminally) as well-delineated lobulated lesions, but do not present as pedunculated intraluminal polyps.[5] Secondary involvement of the small intestine by leiomyosarcoma can occur by direct extension from a primary uterine lesion or by lymphatic/hematogenous spread from far less common sites, including (as in this case) the deep soft tissues. Tumors that metastasize to the intestine by direct extension present on the serosa and later show intramural or intraluminal growth. In contrast, tumors that metastasize to the small intestine via the bloodstream have been noted to arise in the submucosa and present as pedunculated intraluminal masses.[6] In a review of 107 tumors metastatic to the small intestine and colon, Myers and McSweeney[6] reported 23 cases of melanoma metastatic to the small intestine. Like the metastatic leiomyosarcoma in this patient, each metastatic melanoma presented as a pedunculated intraluminal mass arising in the submucosa. They associated multiple tumors of approximately equal size, involvement along a specific arterial distribution, and predilection for lesions along the antimesenteric border with hematogenous metastases and postulated that the unusual appearance of these polypoid tumors was dependent on such factors as the degree of vascularity, rate of growth, and limited desmoplastic response elicited by the tumor.

We are aware of one other reported case in which metastatic leiomyosarcoma presented as an intraluminal polypoid lesion causing intussusception in the small intestine.[7] The patient was a 56-year-old woman who presented with endobronchial and small intestinal metastases 8 years after total abdominal hysterectomy for leiomyosarcoma. The solitary intestinal tumor was the lead point for an intussusception. Leiomyosarcomas that arise in the deep soft tissues tend to be large and aggressive, and metastases are reported in 30% to 50% of cases.[8] To our knowledge, this is the first report of metastases from an extrauterine leiomyosarcoma presenting as synchronous intraluminal polypoid tumors that served as the lead points of 2 intussusceptions in the ileum.

References

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