CASE REPORT

Epithelioid angiosarcoma of the small intestine after occupational exposure to radiation and polyvinyl chloride: A case report and review of literature

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(Received 7 March 2005)

Abstract
Angiosarcomas represent 1–2% of soft tissue sarcomas and most frequently occur in the subcutis. They may affect internal organs, such as the heart, liver, and spleen, and only rarely do they emerge in the gastrointestinal tract. The association between angiosarcomas and certain toxic chemical substances or previous external-beam radiation therapy is well documented.

Keywords: Abdominal pain, CD31, CD34, capsule endoscopy, angiosarcoma/etiology/diagnosis/pathology/surgery, intestinal neoplasms/diagnosis/pathology/surgery, intestine, small, neoplasms, radiation-induced

Introduction
The following case, the fourth report of an epithelioid variant of angiosarcoma of the ileum highlights the possible relation between this extremely rare tumor and occupational exposure to radiation and chemicals such as polyvinyl chloride.

Report of a case
Clinical history
This 68-year-old restaurateur, who used to work as a chemist with 30 years history of heavy occupational exposure to radiation and polyvinyl chloride developed GI bleeding and melena in January 2000. Esophagogastroduodenoscopy and colonoscopy were both negative and the symptoms disappeared.

In April 2003 the patient developed right lower quadrant and epigastric recurrent crampy abdominal pain, associated with melena. He was evaluated with upper and lower endoscopies and then capsule endoscopy, which showed a probable mass at the ileocecal junction (Figure 1).

Abdominal and pelvic CT scan showed a 5 x 4.5 x 4.5-cm right lower quadrant mass, which seemed to arise in the iliac wing of the right iliac fossa. It showed central low attenuation suggesting necrosis and stranding of fat. An isotopic scan for Meckel's diverticulum was inconclusive.

On 16 July 2003 the patient was taken to the operating room for laparoscopic-assisted small bowel resection and partial abdominal wall resection. At surgery he was found to have a loop of small bowel adherent to the right lower quadrant anterior and anterolateral abdominal wall just outside of the pelvis, well above the ureter and iliac and gonadal vessels. The mesenteric loop had a mass in it and the small bowel had a mass as well. The mass was resected together with an adherent portion of abdominal wall. He subsequently recovered from the surgery. The pathology report at that time showed a high-grade angiosarcoma with extensive necrosis, hemorrhage, and a mitotic rate of 18 mitoses per 10 high power fields (Figure 2d).

The sarcoma involved the full thickness of the bowel wall, extensively infiltrated mesenteric fat and infiltrated submucosa and mucosa causing broad ulceration, the growth pattern of the angiosarcoma resembled slit-like blood-filled spaces in some areas, larger vascular spaces with papillary intraluminal...
protrusions in other areas (Figure 2a–c). Tumor was focally present at the mesenteric line of resection. No tumor was seen at the proximal and distal lines of resection of the bowel. Five mesenteric lymph nodes were negative for metastasis. No pathological change was seen in random sections of ileum.

Immunohistochemical stains of tumor tissue (Figure 2e,f) showed a strong positive staining for the endothelial markers CD31 and CD34, confirming the angiosarcomatous nature of this sarcoma. There is no staining for the muscle marker desmin, S-100, cytokeratin or CD117.

A follow-up scan on 9 September 2003 showed a 1.5 × 1.2-cm spiculated lesion in the mesentery anterior to the bifurcation of the right iliac vessel.

A PET scan showed increased activity in the right side of the abdomen consistent with recurrent tumor.
There was a focal area of increased PET activity anterior and medial to the right iliac vessels thought to represent spread to celiac nodes.

He was readmitted in October 2003 for rapidly increasing abdominal distention and ascites, necessitating urgent exploratory laparoscopy. At that time there was massive loculated ascites, which could not be drained, and considerable peritoneal studding. Frozen sections showed a poorly differentiated neoplasm. The tumor cells grew in sheets and epithelioid nests, and invaded diffusely into the fat of the abdominal wall. In some foci there were cleft-like spaces within nests of tumor cells; the lesion appeared more epithelioid with fewer well-formed vascular spaces. The tumor cells were strongly positive for vimentin and CD31.

After surgery he continued to be distended and uncomfortable, but was passing small amounts of gas. Chemotherapy with doxorubicin and ifosfamide was suggested. However, the patient developed severe respiratory distress, and expired before starting on chemotherapy.

Discussion

Angiosarcomas are rare neoplasms characterized by proliferation of tumor cells with vascular endothelial features, accounting for only 1–2% of all soft tissue sarcomas [1,2]. They occur most commonly in the scalp, skin and soft tissues of the head and neck region in elderly men.

Intra-abdominal angiosarcomas are very rare neoplasms, which usually arise in the liver or spleen and extremely rarely in the gastrointestinal tract. From 1965 till 2003, only 13 cases of small intestine angiosarcomas have been reported [1,3–13] in the English literature.

Although the precise predisposing factors of this tumor remain unclear, exposure to vinyl chloride (VC), thorotrast, arsenic chemotherapy, trauma, long-standing lymphoedema, and radiotherapy have been implicated in its pathogenesis [1,2,14]

VC is a known chemical carcinogen known to cause angiosarcoma of the liver (ASL). Because ASL has a latency of approximately 20 years, mortality from ASL caused by VC exposure is still to be expected for some time, even if VC is no longer used industrially in the United States.

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In summary, even though angiosarcomas of the small intestine are rare, such a diagnosis might be a consideration in elderly patients with epithelioid angiosarcoma of the small intestine...
significant vinyl chloride exposure and characteristic symptoms. As in this case, capsule endoscopy and PET scan can contribute to the diagnosis, which can be made definitively by pathological and Immunohistochemical examination.

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