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Short Report

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Primary Malignant Schwannoma of the Small Bowel

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Schwannomas arise from the Schwann cells that cover the peripheral nerves. Most of them are benign. Malignant schwannomas are most commonly seen in the proximal parts of the upper and lower extremities and trunk (1,2). A malignant schwannoma of the small intestine is an extremely rare disease. Only 24 cases have been reported in the English language medical literature (3).

We report a case of a malignant schwannoma of the small intestine in a 53-year-old woman.

Case Report

A 53-year-old woman was admitted to the hospital in June 1999 because of abdominal pain. On clinical examination, an abdominal mass was found. Ultrasound scanning confirmed the mass. A contrast enhanced CT scan of the abdomen revealed a round semisolid heterogeneous well-defined mass (Figure 1). The presumed diagnosis was intestinal carcinoma. No evidence of metastasis was found. The patient did not have neurofibromatosis.

At surgery, the tumor was found in the distal portion of the ileum. Total excision of the tumor with 100 cm resection of the distal ileum was performed. Exploration of the abdomen revealed no carcinomatosis. The liver was unremarkable, and no peritoneal lesions were identified.

Macroscopic pathological examination showed a 100 cm resected ileum, and a tan-gray solid tumor measuring 14x13x8 cm encircling the distal ileum. The tumor mass was adherent to the serosal surface of the ileum and

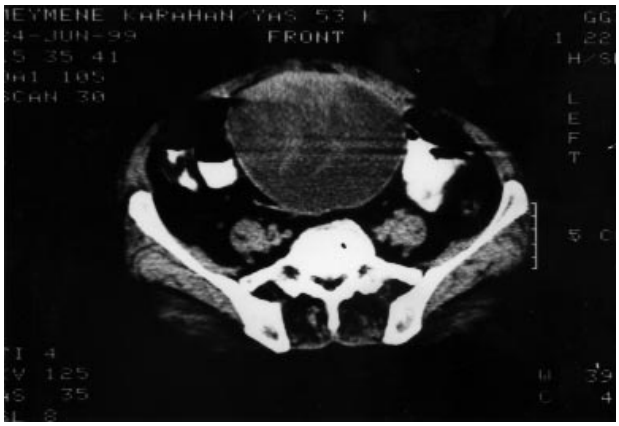

Figure 1. A contrast enhanced CT scan of the abdomen demonstrated a round semisolid heterogeneous well-defined mass.

penetrated the other layers of the distal ileum. The cut surface of the tumor had a gray-white appearance, small cysts, and areas of hemorrhage and necrosis. The surgical margins were negative for the tumor.

Histologically, the tumor showed malignant spindle cells and grew invasively through the intestinal wall. In some areas the tumor had a loose background (Figure 2). The tumor cells had nuclear irregularity, and mitotic activity was easily found. Nucleoli were seen in many tumor cells. In some areas, the tumor contained cartilage and glands made up of well-differentiated, non-ciliated cuboidal cells with clear cytoplasm (Figure 3). Histochemically, the van Gieson stain was negative. Several immunohistochemical stains were used.

For immunohistochemical staining, tissues were fixed in 10% formaldehyde overnight and embedded in

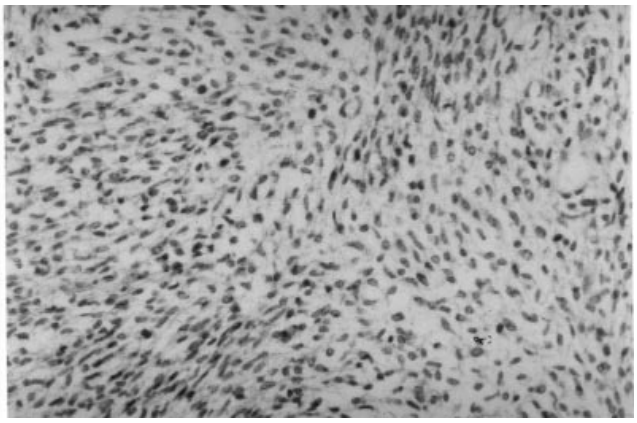

Figure 2. The tumor showing malignant spindle cell within loose background (HE, x200).

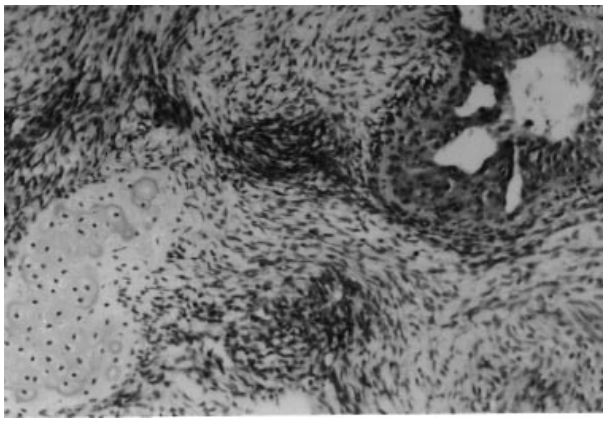

Figure 3. The tumor having a gland made up of non-ciliated cuboidal cells and cartilage (HE, x100).

paraffin. Sections were cut 4 µm thick, dewaxed in xylene, and incubated for 20 minutes in 0.3% H_2O_2 to block endogenous peroxidase activity. The sections were then microwaved for four minutes in PBS (phosphate buffer saline), and incubated with the primary antibody overnight at room temperature. The primary antibody was visualized with diaminobenzidine as chromogen. The sections were counterstained in hematoxylin and eosin, cleared with xylene and coverslipped.

S-100 protein and vimentin were diffusely positive, whereas neuron specific enolase was focally positive (Figure 4). In immunohistochemical staining, keratin, smooth muscle actin and desmin were negative. The pathological diagnosis was malignant schwannoma.

The patient was followed up regularly for eleven month at our hospital. We have not detected any evidence of recurrence. The patient was well 11 months after surgery.

Malignant schwannomas most commonly arise from the proximal parts of the upper and lower extremities and trunk. A few cases have been reported within the head and neck regions, whereas malignant schwannoma of the small intestine is extremely rare (1,2). The tumor is typically a disease of adult life. Most of them occur in patients between 20 and 50 years of age (3,4).

The clinical symptoms of an intestinal malignant schwannoma include fatigue, weight loss and abdominal discomfort. Intestinal obstruction due to compression may cause pain and later ileus may develop. Because these symptoms are usually non-specific and vague, the diagnosis is often late. The preoperative diagnosis of malignant schwannoma of the small intestine may be

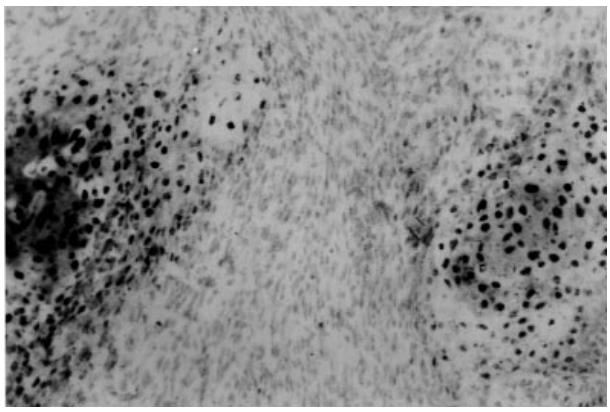

Figure 4-A. Neuron specific enolase focal immunoreactivity in spindle tumor cells (labelled streptavidin biotin method, x200).

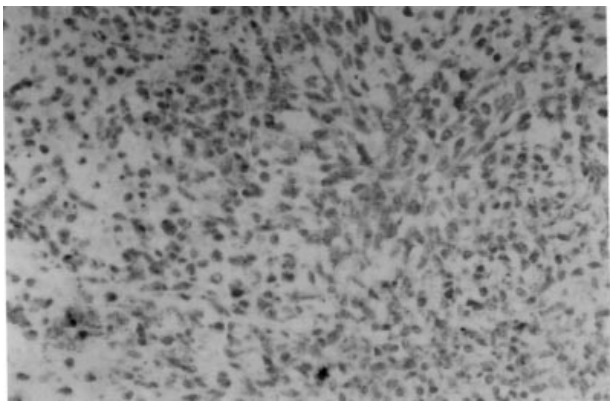

Figure 4-B. S-100 weakly focal immunoreactivity in spindle tumor cells (labelled streptavidin biotin method, x200).

difficult. A palpable mass may be found in the abdomen, as in our patient (3).

There seems to be no characteristic macroscopic picture of malignant schwannoma. The gross appearance is similar to that of other soft tissue sarcomas. The tumor is large and areas of hemorrhage and necrosis are seen. Most malignant schwannomas are quite easily diagnosed as malignancies (5).

It is important to distinguish them from other sarcomas. One of the best ways to differentiate leiomyosarcomas from malignant schwannomas is by light microscopy with van Gieson staining. Also, the immunoreaction for actin is positive in leiomyosarcomas (3,5).

Malignant schwannomas are diagnosed based on the mitotic activity of the tumor. Areas of hemorrhage and necrosis also suggest malignancy. Both benign and malignant Schwann cell neoplasms may contain areas of nuclear atypia, epithelioid areas, melanocytic differentiation, and other mesenchymal elements. Areas of chondrosarcoma, osteosarcoma and angiosarcoma can also be seen (1). Classical schwannomas and malignant schwannomas are immunoreactive for S100, leu7, laminin and glial fibrillary acidic protein, whereas the immunoreaction for desmin is negative in malignant schwannomas (3,1).

On rare occasions, the glands in these tumors may be difficult to distinguish from biphasic synovial sarcomas because the glandular elements may be virtually identical (1). Malignant schwannomas arise from the peripheral nerve sheath, whereas synovial sarcomas have an anatomic relationship to a joint. In our patient, the tumor's location and the gross and microscopic features of the tumor helped us to distinguish between synovial sarcomas and malignant schwannomas. Also, the negative immunostaining for keratin favors malignant schwannoma (6).

Although the glandular elements set this tumor apart as a peculiar histological variant, they essentially serve no role in predicting its biological behavior. The source of the epithelial elements of the glands within these tumors has remained controversial. Some authors have suggested that they arise from heterotopic ependymal cells located within the peripheral nerve, and some authors claim to have documented ependymal features (1).

The primary treatment is surgical. Remission after chemotherapy has been reported. Among the 24 reported patients with a malignant schwannoma of the small intestine, only two patients survived for more than five years. Because of the great risk of recurrence, patients with a malignant schwannoma must be followed up by abdominal CT or ultrasonography after surgery (3).

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