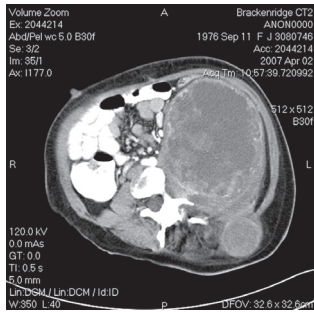
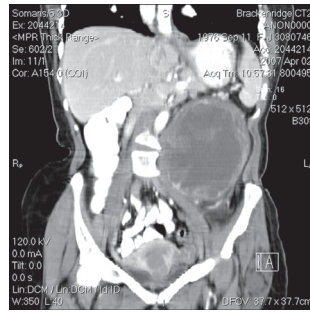


# Abdomen Tumor:

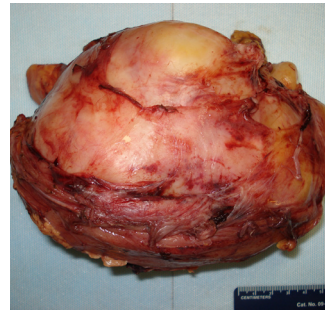
## Malignant Nerve Sheath Tumor in Patient with Neurofibromatosis



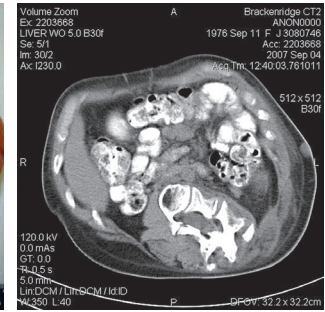
▲ **IMAGE A: CT scan**  
(axial view) of abdomen revealing large retroperitoneal mass with satellite lesion.



▲ **IMAGE B: CT scan**  
(coronal view) of abdomen demonstrating large left sided retroperitoneal mass.



▲ **IMAGE C: Tumor**  
Photo of entire malignant nerve sheath tumor after surgical excision.



▲ **IMAGE D: Follow-up**  
4 month follow up CT scan showing no evidence of recurrence of tumor.

### PATIENT HISTORY:

A 30-year-old Latin American female presented with an enlarging left flank mass. This had increased in size over the 2 months prior to presentation to the surgery clinic.

Her medical history included severe scoliosis as well as hydrocephalus. Her surgical history is significant for placement of a ventriculo-peritoneal shunt as a young child.

A review of her symptoms confirmed left flank discomfort and left-side abdominal discomfort and fullness. On physical exam, she is a pleasant Hispanic female with severe scoliosis. If standing straight as possible she is about 4 feet 6 inches tall. She had a large palpable fullness in the left side of her abdomen and a baseball sized subcutaneous mass in her left flank.

Diagnostic studies included a CT scan of her abdomen and pelvis which revealed a large left-sided retroperitoneal mass with what appeared to be satellite lesions in the left flank and small bowel. CT guided biopsy of the retroperitoneal mass was suggestive of a schwannoma.

### TREATMENT:

She underwent exploratory laparotomy and resection of the retroperitoneal mass. Branches of the left femoral nerve were incorporated into the mass and had to be sacrificed in order to completely resect the mass. At the same time,

the left flank mass was excised as well as two small bowel masses. Additionally, three small skin lesions overlying her left flank mass were removed simultaneously. Her post-operative course was uneventful with the exception of anticipated left lower extremity weakness.

The pathology revealed that the retroperitoneal mass was a malignant peripheral nerve sheath tumor. The small bowel masses were gastrointestinal stromal tumors (GIST). The left flank mass was an atypical neurofibroma and the overlying skin lesions were dermal neurofibromas.

Her case was discussed at the multi-disciplinary adult cancer conference and it was decided that she would receive adjuvant therapy.

### DISCUSSION:

Malignant nerve sheath tumors are uncommon and most are associated with neurofibromatosis. They are treated with radical surgical excision with wide radial margins when possible. If unable to completely excise, subtotal excision is performed followed by radiation therapy. Unfortunately these tumors are not very responsive to radiation therapy or chemotherapy.

Neurofibromatosis is a rare genetic disorder characterized by multiple benign tumors (neurofibromas) in the skin and soft tissues. There are different types of neurofibromatosis

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and this patient meets the criteria for Neurofibromatosis Type 1. Rarely, they can develop malignant nerve sheath tumors as our patient did. Additionally, patients can have a variety of intracranial abnormalities including stenosis of the aqueduct of Sylvius leading to hydrocephalus as this patient has. Other associated conditions include skeletal malformations such as progressive curvature of the spine (scoliosis).

#### **OUTCOME AND FOLLOW-UP:**

At four months post op, the patient is doing well with increasing strength in her left leg with the help of physical therapy. Thus far, there is no evidence of recurrence of any of her tumors. She will continue to get close follow up and routine imaging studies to detect any evidence of recurrence of her disease.

