Case of the Month

A 44 year-old female with a sacral mass

Contributed by Hany Osman, M.D.

Clinical History:

The patient is a 44 year old female with presented with lower back pain and cramps in her lower legs and buttocks. Physical exam shows freckling of the lips, which she says she also notes on her daughter. She has a history of atrial myxoma that was removed and invasive ductal carcinoma of the breast. MRI with contrast of the pelvis revealed an avidly enhancing mass of the sacrum with soft tissue extension. Her uterus was surgically absent on the MRI consistent with a previous hysterectomy. Excisional biopsy was performed and is seen below.

Microscopic Examinations:

See Figures 1-4

Fig. 1

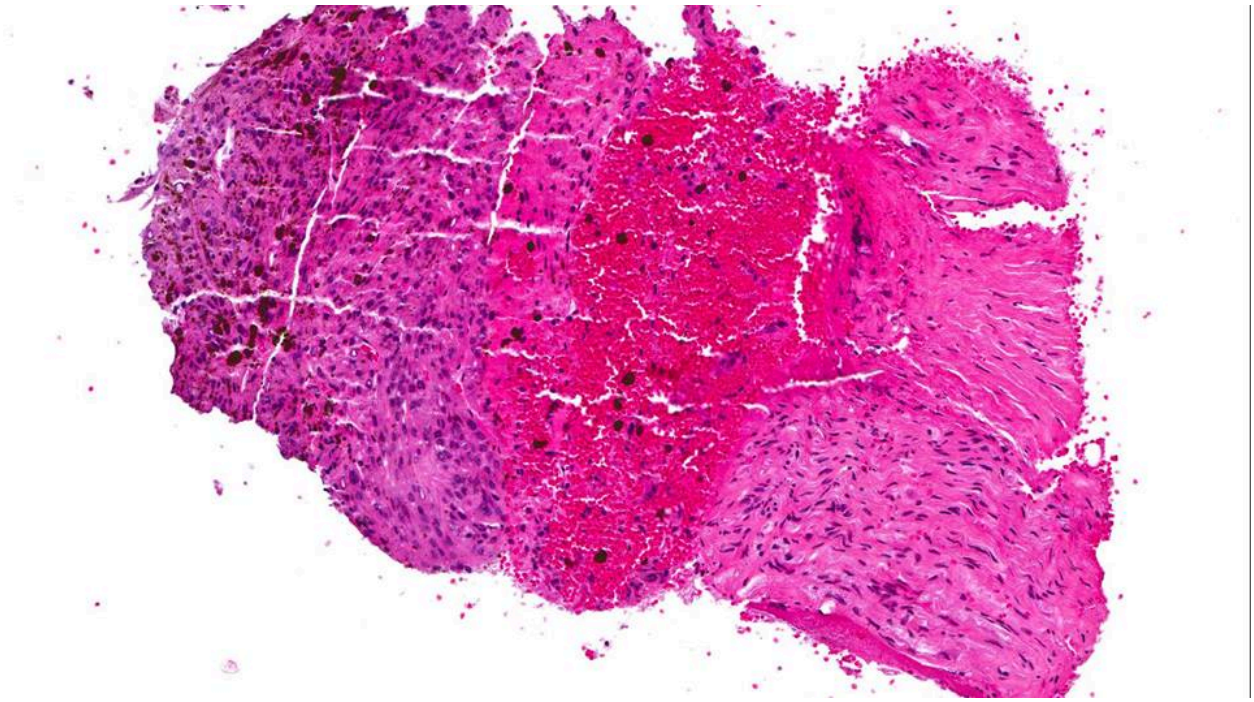

Fig. 2

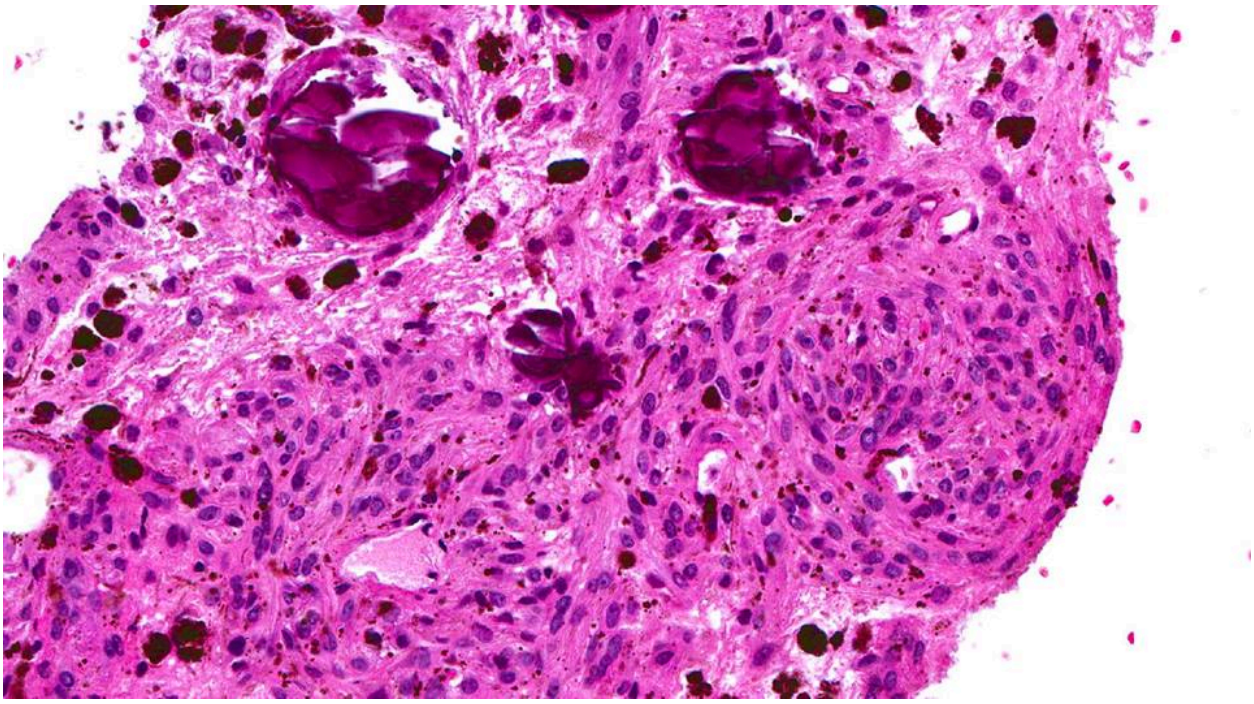

Fig. 3

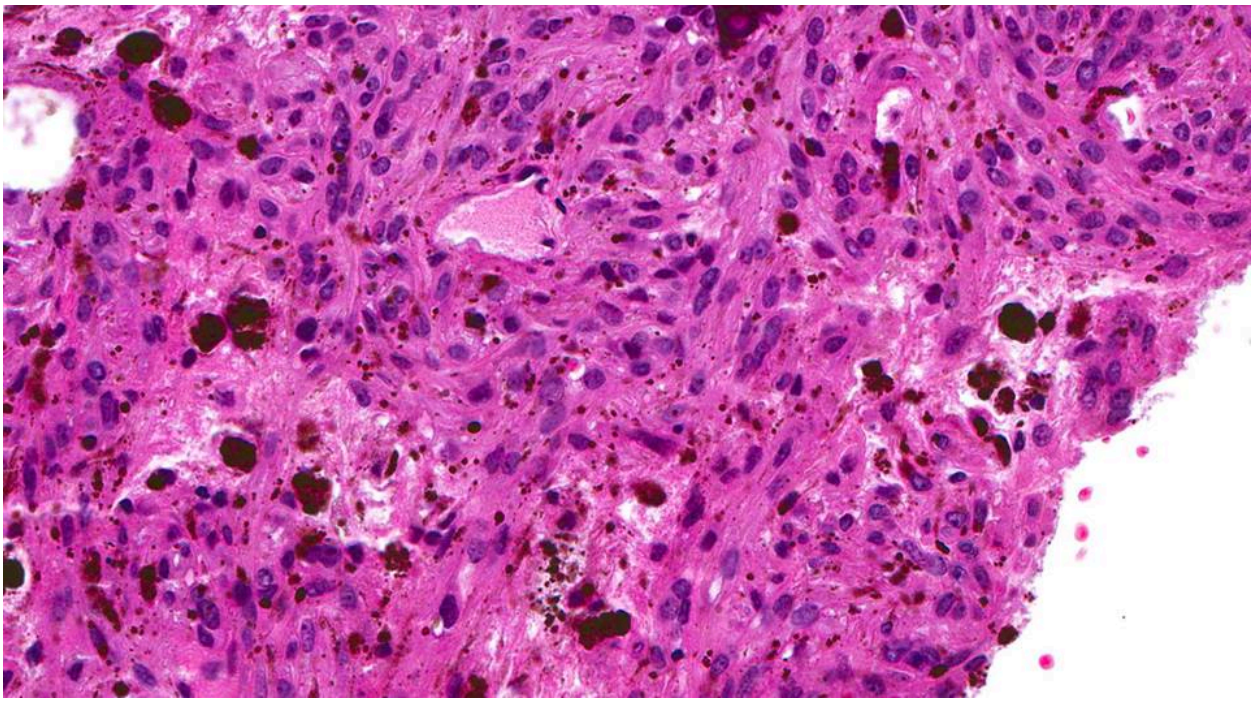

Fig. 4

Figure legend:

1. MRI saggital view showing enhancing lesion involving S1 and S2.
2. Low power view of histological section
3. High power view of histological section
4. Additional high power view of histological section

Immunostaining revealed the cells to be positive for S100, HMB-45, Melan A and negative for CK AE1/3, EMA, CD34, actin, GFAP and estrogen receptor. **What is the diagnosis?**

A. Metastatic ovarian serous carcinoma
B. Pigmented meningioma
C. Malignany peripheral nerve sheath tumor
D. Psammomatous melanotic schwannoma
E. Melanoma
F. Clear cell sarcoma

Discussion:

Psammomatous melanotic schwannoma is a distinctive tumor that is associated with Carney complex. Carney complex is a rare heritable disease associated with multiple neoplasia. Presence of pigmented skin and mucosal lesions are characteristic of this syndrome. In general, melanotic schwannomas consist

of melanin containing nerve sheath tumors and may be psammomatous or non-psammomatous. About half of the psammomatous melanotic schwannomas are associated with Carney complex. The behavior of these lesions is hard to predict, however they do have a tendency to recur locally with incomplete resection and 10% of the tumors do metastasize. The pathologic features are characteristic. They are grossly brown to black and usually have a dumbbell appearance. Unlike a classic schwannoma, psammomatous melanotic schwannomas do not have Antoni A and Antoni B areas, instead they demonstrate spindle and epitheloid cell proliferation with plump cells containing melanin pigment. The cells are arranged in fascicles with occasional whorling and palisading. Psammoma bodies are readily seen in psammomatous melanotic schwannomas. The tumors are typically positive for S100, Melan A and HMB-45. Despite the immunohistochemical resemblance, they are genetically distinct from both melanomas and schwannomas.