

Pelvic alveolar rhabdomyosarcoma in a young adult

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Rhabdomyosarcomas are soft-tissue tumors, rare in adults. Accounting for nearly 5% of childhood cancers, they represent less than 0.03% of adult malignancies (1, 2). Three different subtypes of rhabdomyosarcoma have been described (embryonal, alveolar and pleomorphic), making up approximately 50%, 30%, and 20% of the cases, respectively (3). Although the definitive diagnosis is made pathologically, some distinguishing features among these subtypes, and between rhabdomyosarcomas and other soft-tissue tumors, can be suggested on MRI and CT. We present an interesting case of a 20-year-old female with a locally aggressive pelvic alveolar rhabdomyosarcoma. While the prognosis has improved with newer treatment techniques, overall survival rates remain poor. Our case study presents typical features of a rare disease, which can often present a diagnostic dilemma for clinicians.

Case report

A 20-year-old female presented for treatment of a rapidly enlarging mass in her right groin. The mass was palpable and somewhat painful on physical exam, and the patient was sent for a pelvic MRI for further evaluation. Contrast-enhanced MRI of the pelvis (Fig. 1) showed a 9.7 x 5 x 5.5 cm heterogeneous mass in the perineum on the right, involving the ischiorectal fossa and abutting the labia. The mass was heterogeneous but hypointense on T1-weighted sequences, hyperintense on STIR, and demonstrated heterogeneous contrast enhancement (Figs. 2, 3, and 4).

Contrast-enhanced CT of the abdomen and pelvis on the same date showed a heterogeneous mass in the right ischiorectal fossa with associated infiltration around the right pelvic sidewall, and a few normal-sized pelvic and inguinal lymph nodes. The working differential diagnosis included carcinoma of the vulva or vagina, soft-tissue sarcoma, and neuroendocrine carcinoma. Following the MRI,

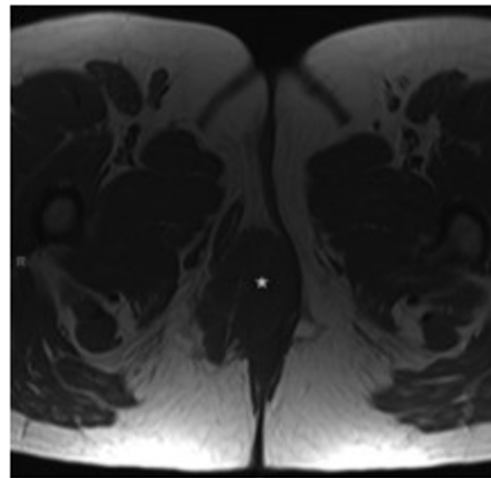


Figure 1. Axial, T1-weighted, precontrast image shows the 9.7 x 5 cm mass (*) in the right ischiorectal fossa, abutting the labia, to be isointense to skeletal muscle.

the patient was sent for biopsy. Results demonstrated markedly atypical small cells in a background of fibroconnective tissue and skeletal muscle, with a leading differential diagnosis of high-grade neuroendocrine carcinoma based on histologic appearance and immunohistochemistry.

Following the biopsy results, the patient underwent radical excision of the pelvic tumor, including radical hemivulvovaginectomy and right inguinofemoral lymphadenec-

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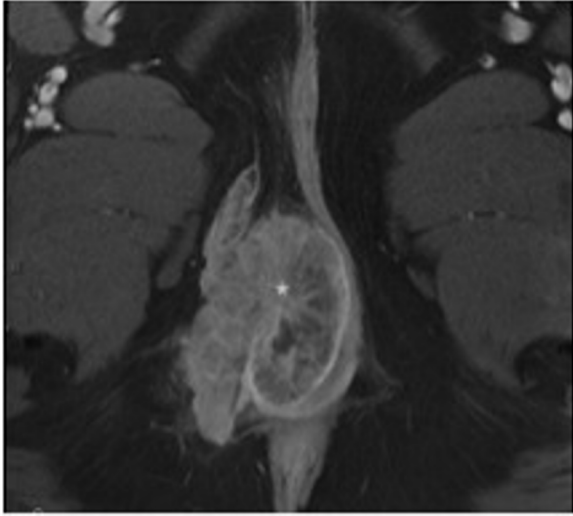


Figure 2. Axial, T1, postcontrast image shows the mass (*) to enhance heterogeneously.

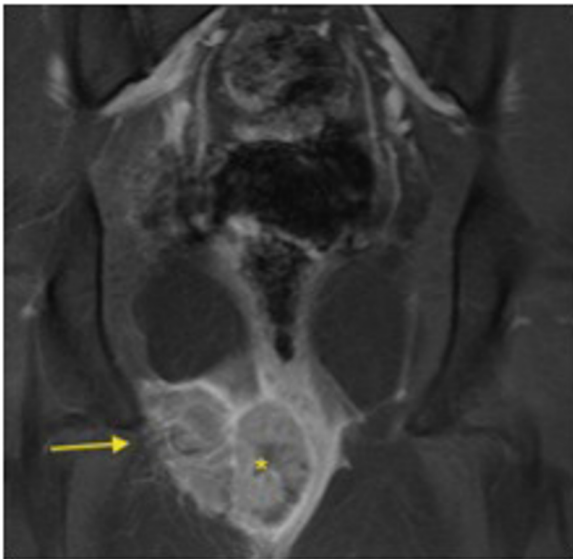


Figure 3. Coronal, T1, postcontrast image shows the mass (*) to infiltrate the adjacent musculature, and have a poorly defined lateral margin (arrow).

tomy, as well as placement of fiducial markers for postoperative radiation therapy. At surgery, the mass was found to be intimately involved with the right inferior pubic ramus, and grossly metastatic lymph nodes were visualized (Figs. 5A, B). These surgical findings confirmed the need for further postsurgical treatment with chemotherapy and radiation.

Microscopically, the resection specimen showed a highly cellular tumor with marked necrosis and fibrous stroma. Numerous fibrovascular septae separated the discohesive tumor cells into discrete nests, where the tumor cells appear to float in alveolar spaces (Figs. 6A, B). The

malignant neoplasm was composed of monomorphous round nuclei (Fig. 7) with deep eosinophilic cytoplasm. A few multinucleated giant cells (Fig. 8) were also present. Immunohistochemically, the tumor cells showed strong positivity for desmin and nuclear positivity for myogenin (Fig. 9) yet negativity for cytokeratin, synaptophysin, and

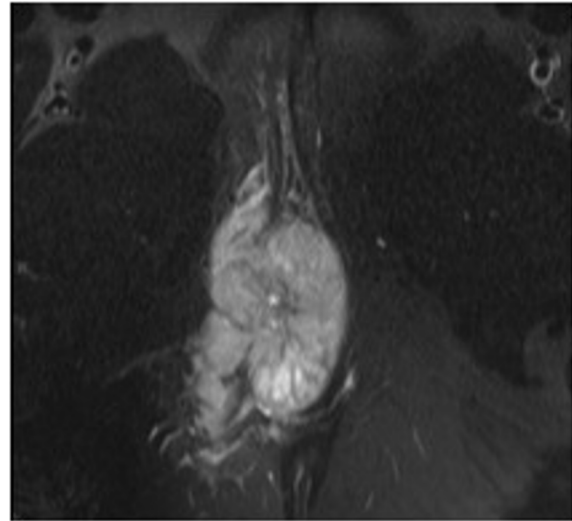


Figure 4. Axial STIR image shows the mass to be relatively hyperintense to skeletal muscle.

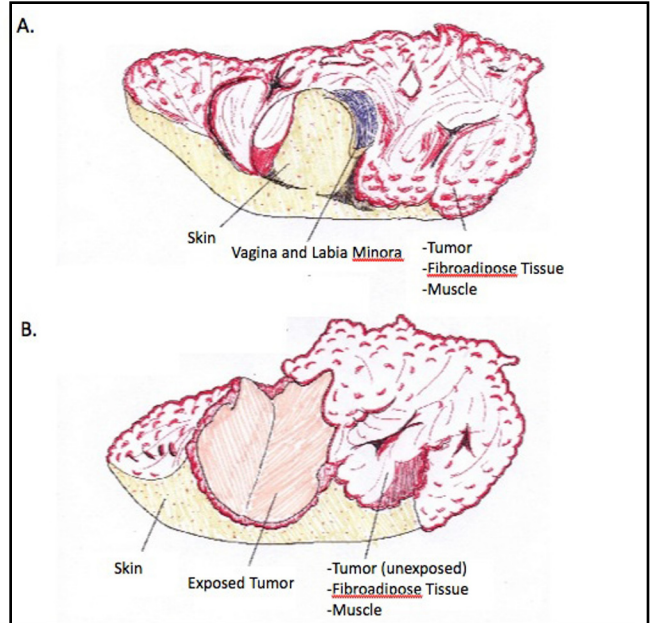


Figure 5. A: Large soft-tissue mass with surrounding skin and fibroadipose tissue, 17 x 10 x 8 cm. Attached skin is 14 x 11.5 cm. Portion of the vagina and labia minora is 4 x 3.5 cm. B: Sectioning of the specimen reveals a large multi-lobulated tumor, 8 x 6 x 5.5 cm. Note tan to slightly yellow gelatinous cut surface, with large areas of caseous necrosis. Tumor is well circumscribed, with a thin capsule.

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