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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, 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.

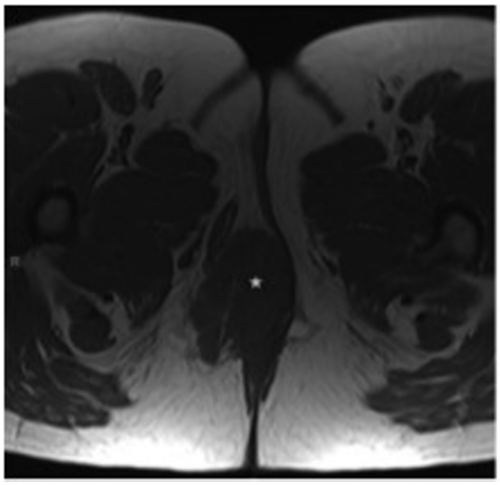

Figure 1. Axial, T1-weighted, precontrast image shows the 9.7 x 5 cm mass (*) in the right ischiorectal fossa, abutting the labia, to be isointense to skeletal muscle.

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

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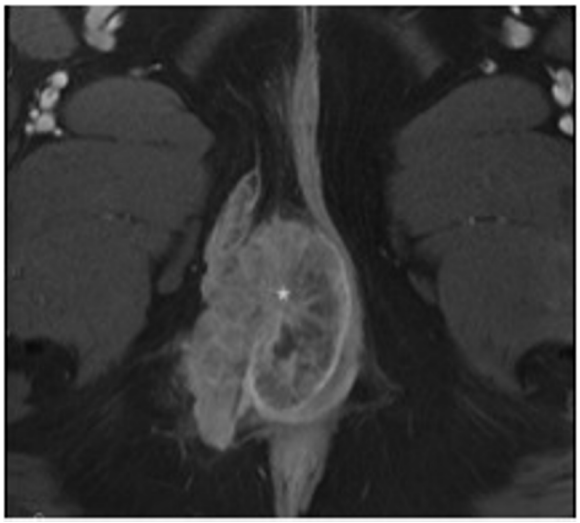

Figure 2. Axial, T1, postcontrast image shows the mass (*) to enhance heterogeneously.

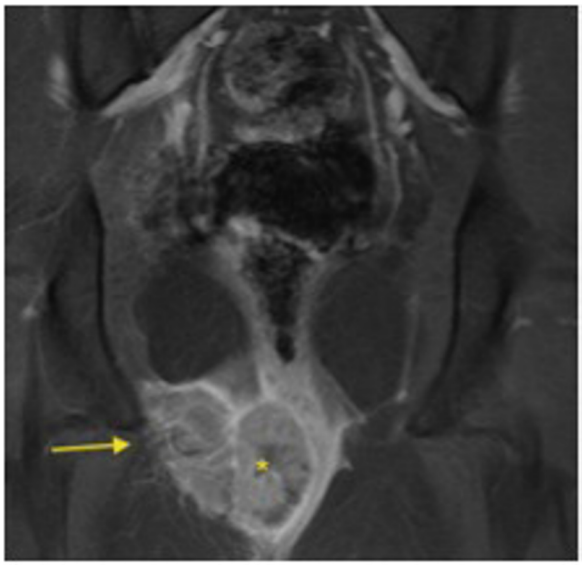

Figure 3. Coronal, T1, postcontrast image shows the mass (*) to infiltrate the adjacent musculature, and have a poorly defined lateral margin (arrow).

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

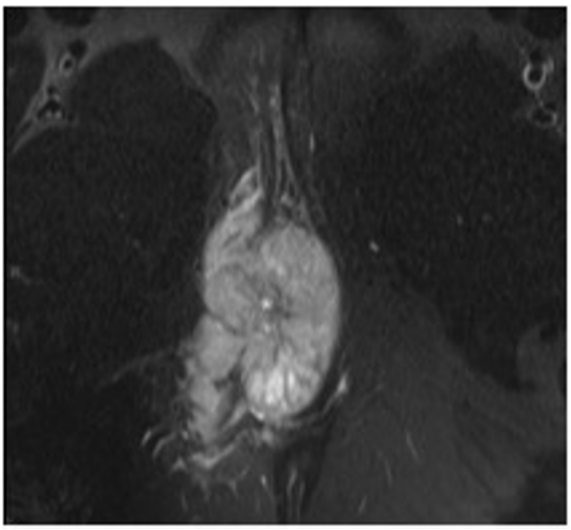

Figure 4. Axial STIR image shows the mass to be relatively hyperintense to skeletal muscle.

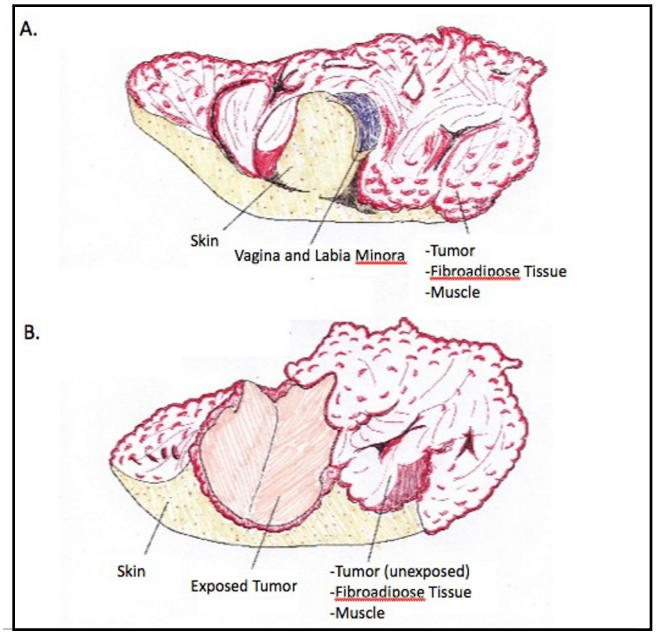

Figure 5. A: Large soft-tissue mass with surrounding skin and fibroadipose tissue, 17 x 10 x 8 cm. Attached skin is 14 x 11.5 cm. Portion of the vagina and labia minora is 4 x 3.5 cm. B: Sectioning of the specimen reveals a large multilobulated tumor, 8 x 6 x 5.5 cm. Note tan to slightly yellow gelatinous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

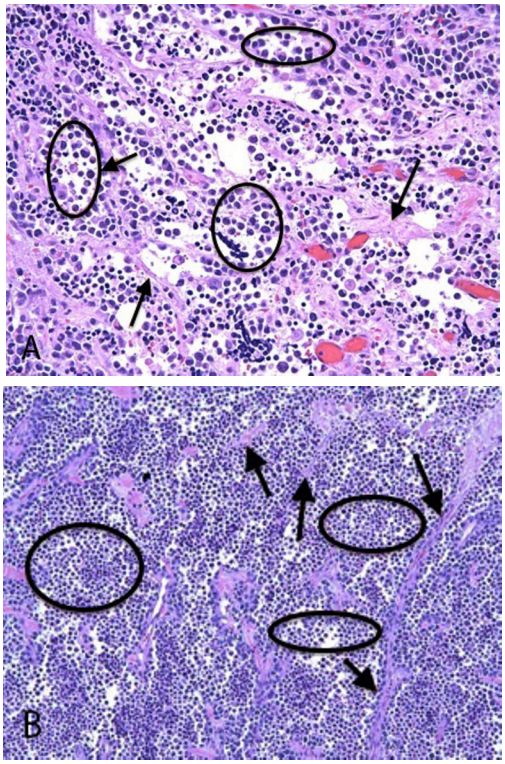

Figure 6. Nests of discohesive tumor cells (ovals) separated by thin fibrovascular septae (arrows), imparting alveolar appearance (A, 10X; B, 4X).

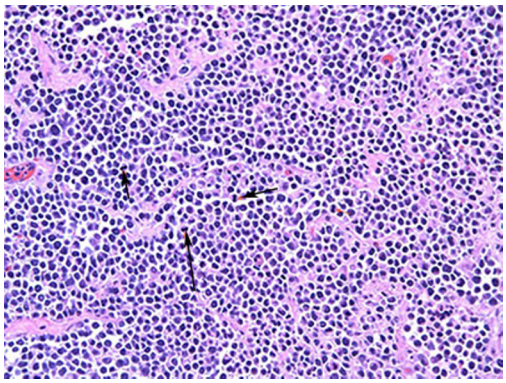

Figure 7. Discohesive monomorphous small round tumor cells with numerous mitoses (arrows) (10X).

chromogranin, establishing the diagnosis of alveolar rhabdomyosarcoma. Positivity for keratin and neuroendocrine markers have been reported in alveolar rhabdomyosarcoma, especially in the sinonasal tract, and may cause a serious diagnostic confusion. This tumor showed extensive positivity for desmin and diffuse nuclear positivity for myf-4 (myogenin) in virtually 100% of the viable lesional cells, excluding the possibility of small-cell carcinoma. The expression of myogenin is limited to skeletal muscle, so nuclear positivity to the antibodies directed against them helps establish the myoid lineage and a diagnosis of rhabdomyosarcoma.

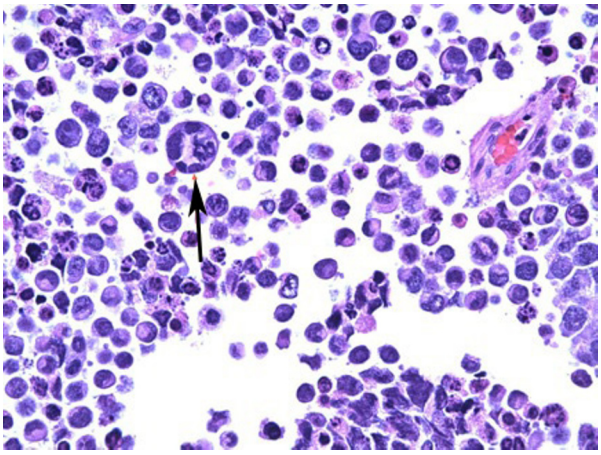

Figure 8. Occasional multinucleate giant cell (arrow) (40X).

A PET scan (Fig. 10) showed residual tumor deposits in the postoperative bed, consistent with high-grade malignancy by quantitative analysis. There were also focal areas of hypermetabolism along the right pelvic sidewall and

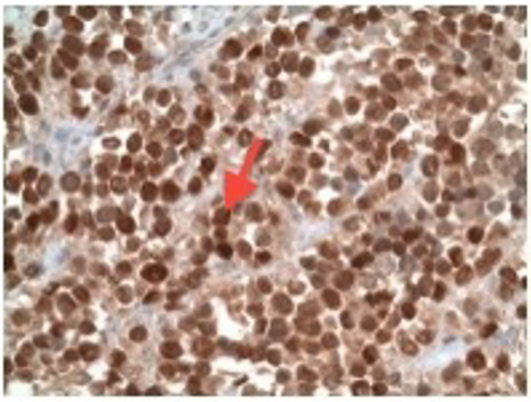

Figure 9. Nuclear myogenin positivity in tumor cells (20X).

ischiorectal fossa, as well as hypermetabolic portacaval lymph nodes, consistent with metastatic disease. The patient was started on Vincristine, Actinomycin, and Cyclophosphamide and is still currently undergoing treatment with chemotherapy.

Discussion

Rhabdomyosarcomas derive from rhabdomyoblasts, primitive mesenchymal precursor cells of skeletal muscle (4, 5). Most commonly, they occur in children under the age of 12. In adults, rhabdomyosarcomas are extremely rare, accounting for approximately 0.03% of solid tumors (1, 2). Overall incidence is 4-6 per 1,000,000.

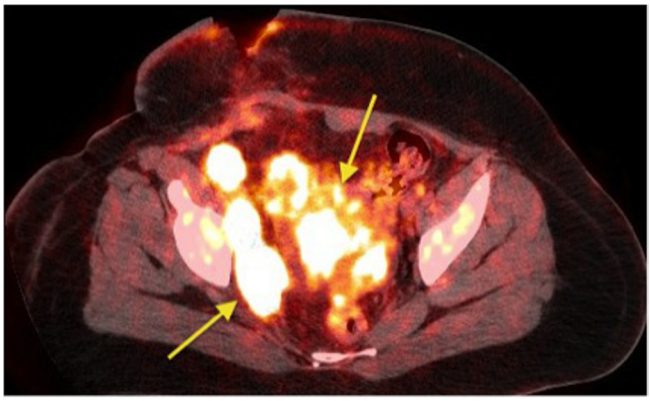

Figure 10. PET/CT image demonstrating foci of hypermetabolism within the postoperative bed and the right pelvic sidewall (arrows).

Of the three known subtypes, alveolar rhabdomyosarcoma is the most common to occur in the young-adult age group (6). The imaging appearance of rhabdomyosarcomas can vary. One study by Allen et al. in 2007 showed that the majority are isointense to skeletal muscle on T1, and hyperintense to skeletal muscle on T2. Following IV gadolinium administration, all tumors showed either moderate or avid heterogeneous enhancement. On unenhanced CT, the tumors ranged from iso- to hyperdense to skeletal muscle (7). Necrosis is a common feature, while the presence of calcification argues against rhabdomyosarcoma (8). Imaging can play a vital role in treatment planning and surgical staging. MRI is the imaging exam of choice for pelvic rhabdomyosarcomas, with complementary CT examinations used to evaluate potential involvement of cortical bone. Differentiating among the different subtypes is primarily in the realm of the pathologist; however, some features may suggest a particular histologic diagnosis. The embryonal subtype tends to be the most homogeneous and isodense to skeletal muscle, while the alveolar subtype is usually more heterogeneous (7).

Common treatment is surgical resection, if possible, followed by radiation and chemotherapy (9-11). Unfortunately, the prognosis remains poor, as many patients have metastases at the time of diagnosis and recurrence is quite common. Lymphadenopathy is common at presentation, and the lungs represent the most common site of metastases (12). The 5-year survival rate is less than 50%. Children overall fare better than adults (13).

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