

Clinical Image

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Extraskelletal Ewings sarcoma of the right inguinal region

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Case presentation

12-year-old Caucasian male presented to the emergency department with a two-day history of vomiting and a new rapidly growing right groin mass. The patient denied pain or urinary symptoms. Physical examination revealed a palpable, non-tender, non-reducible, semi-solid mass in the right inguinal region. Ultrasound examination showed a heterogeneous mass with solid and cystic components. CT imaging confirmed the mass is separate from the spermatic cord and the absence of lymphadenopathy.

An ultrasound-guided core biopsy was performed. Microscopic examination, immunohistochemical staining and FISH analysis confirmed a diagnosis of Extraskelletal Ewing Sarcoma (EES). Staging fluorodeoxyglucose F-18 PET/CT showed increased avidity of the mass without metastatic disease. Chemotherapy with vincristine, doxorubicin, and cyclophosphamide followed by ifosfamide and etoposide was initiated. Three months later, the patient underwent surgical resection with no recurrence or metastasis on follow-up imaging.

Discussion

EES is a rare type of Ewing Sarcoma (ES), accounting for about 25% of all cases [1]. Its average age of diagnosis is 20 years with equal gender distribution [2]. Caucasians are predominantly affected, and the most common location is the paravertebral region (30%), followed by the lower extremities (25%) [3]. EES usually presents as a rapidly growing solitary mass, with lungs and bone being metastatic sites [3].

CT is often the first imaging modality used, showing a well-demarcated mass with muscle-like density and heterogeneous enhancement [3]. MRI is the preferred method for characterizing EES, with the mass often containing areas of high T2-weighted signal intensity due to cystic change or necrosis [3]. FDG PET/CT is sensitive for staging and detecting nodal and distant metastases, as EES typically exhibits increased metabolic activity. Definitive diagnosis requires biopsy and pathological examination.

Local disease management starts with neoadjuvant chemotherapy before radiation or surgical resection. The 5-year survival rate for localized disease is approximately 75% with appropriate chemotherapy regimens, but drops to 15-32% for metastatic disease [3].

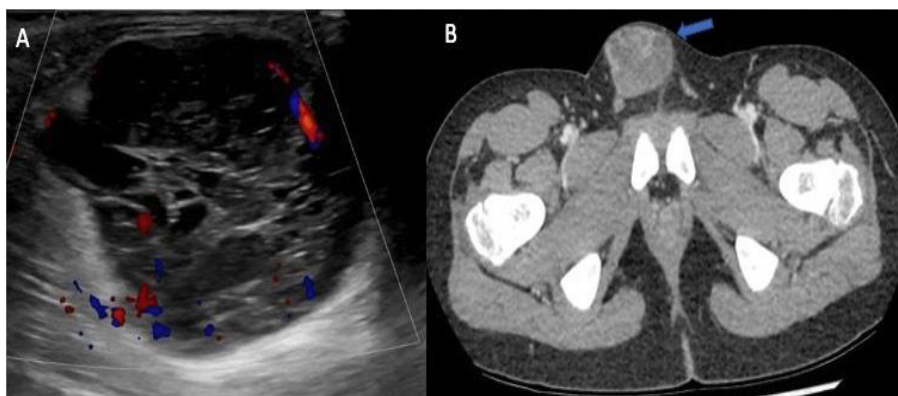


Figure 1: **A)** Ultrasound of the right inguinal region of a 12-year-old male showing heterogenous solid and cystic mass with some internal vascularity on Doppler sequences. **B)** Axial computed tomography scan of the pelvis showing a 3.9 x 3.7 x 4 cm heterogenous well circumscribed mass in the right inguinal region (arrow) anteriorly and separate from the spermatic cord.

References

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